Ciro Esposito
Giovanni Esposito
Editors

Pediatric Surgical Diseases

A Radiologic Surgical Case Study Approach

Springer

C. Esposito · G. Esposito (Eds.)

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C.T. Albanese · M. Fujioka · G. MacKinlay · N. Rollins
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To my mentors, Ettore Ruggieri, Giuseppe Zannini and François D'Allaines, who showed me the way to be a part of the medical world.
With gratitude,
Giovanni Esposito

To my father, the best pediatric surgeon I have ever known,
and the man who instilled in me the thrill, challenge and satisfaction of lifelong learning.
I thank him for his continuous teaching, and for transferring to me his love for the children we care for every day.
With gratitude,
Ciro Esposito

Preface

The idea to write this book was born about 3 years ago when we were changing the seat at the Unit of Pediatric Surgery of the University of Naples "Federico II."

We realized that during the 30-year activity of our unit we had collected a large number of rare and interesting cases of pediatric surgical diseases.

The difficulties met in the diagnosis of many of these cases gave us the idea to publish some of the most important ones, each with a collection of images accompanied by text providing a practical guide to reach the final diagnosis.

Since our idea was welcomed by numerous colleagues and friends, we proposed to Springer to publish this book.

All the authors who accepted to participate in our project (pediatricians, pediatric surgeons, pediatric and adult radiologists) are considered leading world experts in the diagnosis and treatment of pediatric surgical diseases, and we are extremely grateful to them for their contribution and for devoting their time to producing such outstanding reviews.

Thus, with their precious help, we have created an educational text, focusing on more than 200 case reports of pediatric surgical diseases, which will certainly be very useful to all professionals working in pediatrics who need to prepare themselves when approaching a variety of diagnostic and therapeutic problems in conditions affecting infants and children.

We would like to thank in particular our associate editors, Craig T. Albanese, Masayuki Fujioka, Gordon MacKinlay, Nancy Rollins, and Felix Schier, whose great competence and extensive experience in this field helped us in writing, collecting, and organizing this material.

Ciro Esposito, Giovanni Esposito
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Abbreviations

AP	Anteroposterior
ALT	Alanine aminotransferase
AST	Aspartate aminotransferase
CE	Contrast enhanced
CRP	C-reactive protein
CT	Computed tomography
DMSA	Dimercaptosuccinic acid
ECG	Electrocardiogram
ERCP	Endoscopic retrograde cholangiopancreatography
EXIT	Ex utero intrapartum treatment
FDG	[(18)F]-fluorodeoxyglucose
FLAIR	Fluid attenuated inversion recovery
FNAB	Fine-needle aspiration biopsy
Gd	Gadolinium
Gd-DTPA	Gadolinium-diethylenetriamine pentaacetic acid
GER	Gastroesophageal reflux
GI	Gastrointestinal
HCG	Human chorionic gonadotropin
ICU	Intensive care unit
IV	Intravenous
IVU	Intravenous urogram
MCDK	Multicystic dysplastic kidney
MIBG	Metaiodobenzylguanidine
MR	Magnetic resonance
NMR	Nuclear magnetic resonance
NPO	Nil per os
PET	Positron emission tomography
SIOP	International Society of Pediatric Oncology
STIR	Short tau inversion recovery
US	Ultrasonography
UTI	Urinary tract infection
VATS	Video-assisted thoracoscopic surgery
VCE	Video capsule endoscopy
VUR	Vesicoureteral reflux

Introduction

We think that a quick introduction is necessary to explain how this book is organized and how it should be read.

The book is divided into eight sections, six of them focusing on different parts of the human body and two sections dedicated to emergency, trauma, and tumors.

The book is easy to read and includes more than 200 case reports of different pathological conditions. Each radiological surgical case comprises no more than two printed pages.

The question page (Q) is a right-hand page; the answer page (A) is found overleaf.

The Q page contains radiological images (radiography, CT, MRI, scintigraphy, ultrasonography etc.), focusing on a specific case. Accompanying the figures is the clinical history of the patient along with some questions on the interpretation of the images for the diagnosis and management of the case presented.

The A page contains the interpretation of the images shown on the Q page, and possibly other figures of diagnostic procedures performed in the same case in order to obtain a clear diagnosis. The A page also includes information about the particular condition affecting the patient and the management of the case shown, including therapy and follow-up. Most case reports end with a short "Suggested Reading" list.

1

Head and Neck

Introduction

The patient of pediatric neurosurgery ranges from the preterm infant to the adolescent. The diseases addressed in pediatric neurosurgery include congenital disorders such as hydrocephalus, spina bifida, and craniosynostosis, particular types of tumors of the central nervous system, cerebrovascular diseases including vascular malformations and moyamoya disease, intractable epilepsy, and traumatic brain injury or acquired disorders. Pediatric neurosurgery requires team work that involves pediatric specialists from various fields including pediatric anesthesia, neurology, neuro-oncology, plastic surgery, psychology, and neuroradiology.

In this section, 13 pediatric cases of head or neck diseases are presented. These include brain tumors and cerebrovascular diseases. The imaging studies performed for the diagnosis of these pathological conditions are computed tomography, magnetic resonance imaging, cerebral angiography, and single photon emission computed tomography. The brain tumors discussed in this section are craniopharyngioma, ganglioglioma, medulloblastoma, choroid plexus papilloma, pilocytic astrocytoma, dysembryoplastic neuroepithelial tumor, anaplastic astrocytoma, and atypical meningioma. Cerebral arterial aneurysm, moyamoya disease, and moyamoya syndrome are also presented.

All patients underwent neurosurgical operations and had a good recovery after the surgery. We hope the readers will enjoy learning the characteristics of diagnostic imaging in these surgical cases.

Q 1

Nancy Rollins

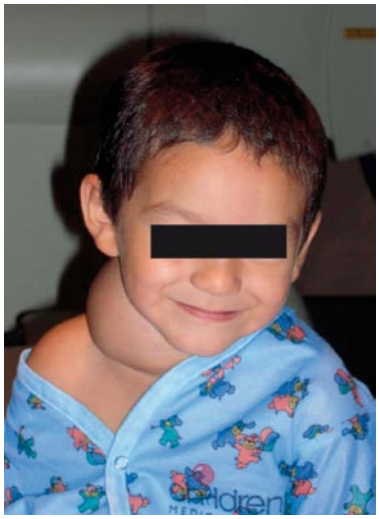

Fig. 1

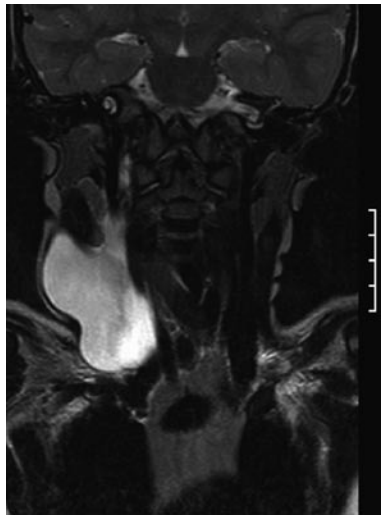

Fig. 2

A 4-year-old boy presented with a soft, slowly enlarging, right cervical soft-tissue mass (Fig. 1).

- What is the differential diagnosis?
- What is the best imaging strategy?
- What does the MR image show (Fig. 2)?
- Should the lesion be biopsied or resected?
- Is there a nonsurgical alternative for treatment?

A 1

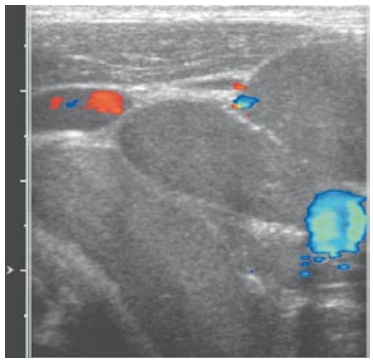

Fig. 3

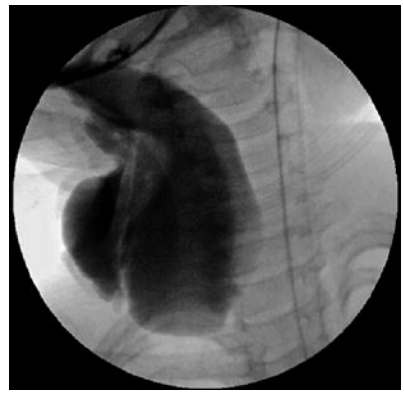

Fig. 4

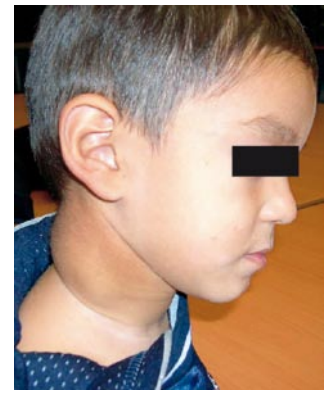

Fig. 5

The differential diagnosis includes a low-flow vascular malformation, neuroblastoma or nerve-sheath tumor, or possibly a rhabdomyosarcoma. Contrast-enhanced MR imaging is used to define the internal architecture of the lesion, e.g., solid vs. cystic, slow flow vs. high flow, and the extent of the lesion proximity to vital structures such as the carotid sheath. Lymphatic malformations are usually low signal on T1 images; however, the signal intensity on the T1 images may be similar to or higher than that of regional muscle if bleeding has occurred into the lymphatic malformation or if the fluid has a high protein content. The presence of fluid-fluid levels is almost pathognomonic of a lymphatic malformation. On T2 images and STIR sequences (Fig. 2), the fluid shows very bright signal although blood products may decrease in signal intensity. Ultrasonography may show a cyst(s) with absence of echoes or medium levels of echoes in high proteinaceous or hemorrhagic fluid (Fig. 3). The MR image shows a large, fluid-filled, unilocular cyst extending from the skull base to the supraclavicular region anterior to the sternocleidomastoid muscle. The imaging findings are classic for a macrocystic lymphatic malformation and biopsy is not indicated. The lesion is best treated with sclerotherapy.

Lymphatic malformations are composed of dysplastic vesicles or pouches filled with lymphatic fluid. The pouches of fluid may be large (macrocystic) or microcystic. Lymphatic malformations are often admixed with a venous malformation, e. g., venolymphatic malformations. Macrocystic lymphatic malformations and mixed venolymphatic malformations are amenable to sclerotherapy, whereas microcystic lesions and microcystic components of lymphatic malformations are usually not. Sclerotherapy is usually performed under fluoroscopic guidance, although ultrasonography is useful in puncturing nonpalpable lesions. The cyst should be emptied of fluid as much as possible. Contrast medium is injected to document correct intralesional positioning of the needle and lack of extravasation of contrast and of the sclerosing agent (Fig. 4). Effective sclerosing agents include OK-432, absolute alcohol, and doxycycline. OK 432 (picibinal) is a lyophilized mixture of a low-virulence strain of *Streptococcus pyogenes* mixed with benzypenicillin. Intralesional hemorrhage may complicate sclerotherapy, which is seen as an abrupt increase in the size of the lymphatic malformation and change from a soft spongy lesion to a tense slightly painful one. Intralesional hemorrhage does not, as a rule, require drainage since the hemorrhage will slowly resolve. Figure 5 shows the patient 2 weeks after sclerotherapy.

Q 2

Nancy Rollins

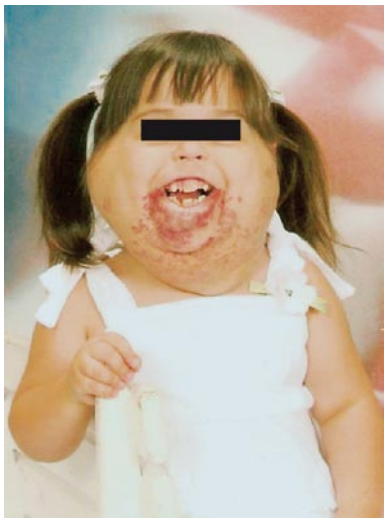

Fig. 1

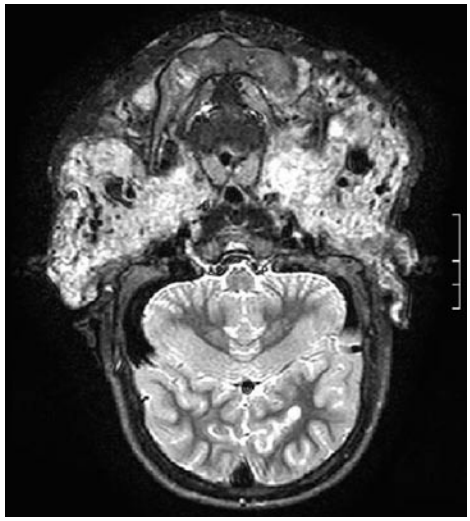

Fig. 2

A 3-year-old girl presented with a large disfiguring facial mass that failed to involute with high-dose pulsed steroids and alpha interferon. Figure 1 shows the patient at presentation. Figure 2 is a cross-sectional image of the face.

- What is the differential diagnosis?
- Should this lesion be biopsied?
- What are the options for medical management?
- What does the MR imaging show?

A 2

MR imaging (Fig. 2) shows a large nonlipomatous mass which enhances and which has extensive involvement of the parotid glands and muscles of mastication as well as the infratemporal fossa. Branches of the external carotid arteries are dilated as are the internal jugular veins indicating a high-flow lesion.

The patient underwent sequential arterial embolizations using particles and coils with considerable decrease in the size of the lesion. Figure 3 shows the patient 1 year later. There is residual facial deformity due to residual fibro-adipose tissue that will be corrected surgically. Laser therapy will be used to treat the remaining cutaneous component.

Hemangiomas usually appear within 2 weeks after birth as a small red blemish or bump, which grows rapidly. The lesion may spontaneously regress, usually between 12–18 months of age. Complete regression results in the lesion being inapparent by age 3–5 years of age, with no or only minor residual scaring. In other patients, involution may take longer; 50% will involute by age 5, 70% by age 7, and 90% by the age of 9. Lesions which regress slowly are often associated with scaring, atrophoderma, stria, and cutaneous discoloration. Hemangiomas that require early aggressive treatment include those that are cosmetically deforming, growing rapidly, or obstructing vision, hearing, breathing, eating or, any other body function.

Systemic corticosteroids 2–3 mg/kg, given for 4-8 weeks comprise the first-line therapy for complicated hemangiomas; regression rates of up to 90% have been reported. Intralesional corticosteroid injections may be used for lesions that are smaller than 3 cm in diameter and well-defined and for lesions that show ulceration. Three to five intralesional injections are usually given at 6-week intervals; each dose should not exceed 3 mg/kg.

Hemangioma not responsive to corticosteroid therapy may be treated with both alpha and the 2a form of alpha interferon. However, treatment with interferon is associated with the development of irreversible spastic diplegia in about 20% of children. Vincristine is now recommended for hemangiomas with airway, eyelid, and orbital involvement, disseminated neonatal hemangiomatosis of the skin, liver, kidney, and cardiac failure. A weekly dosage of 1 mg/m(2) is injected intravenously. The dose is tapered depending on the clinical response. The reported range of injections is 5–25 with a length of treatment of 1.5–8 months. Dramatic response may be observed within 1 month of treatment, although a slow protracted response may also occur.

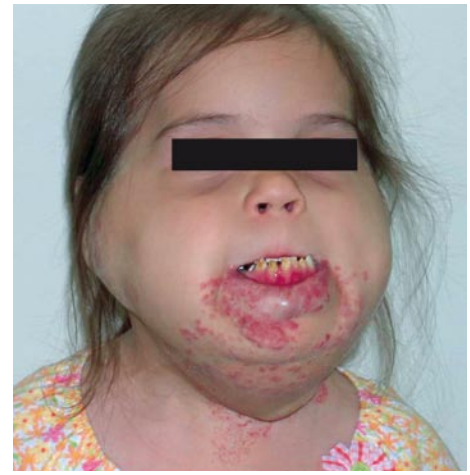

Fig. 4

Superficial hemangiomas may be treated with pulsed dye laser, but deeper lesions are not treatable with this modality as the depth of laser penetration is only 1–2 mm. For large multicompartmental facial lesions, arterial embolization is usually effective at accelerating the regression of the hemangiomas. The procedure involves superselective catherization of branches of the external carotid arteries and occlusion of arteries supplying the hemangiomas using particulate material and small endovascular coils. The internal carotid arteries should also be studied to assess what, if any, contributions to the hemangiomas arise from the internal carotid arteries and to exclude carotid stenosis in patients with PHACE syndrome (posterior fossa brain malformations, hemangiomas, arterial anomalies, coarctation of the aorta and cardiac defects, and eye abnormalities).

Sequential embolizations are needed to devascularize the lesions because arterial collaterals form rapidly. Potential complications of embolization include inadvertent embolization of the central retinal artery causing blindness as well as stroke and damage to the femoral arteries resulting in leg length discrepancy. If surgical removal or reconstruction is needed, preoperative superselective embolization is recommended to minimize intraoperative blood loss.

Q 3

François Luks

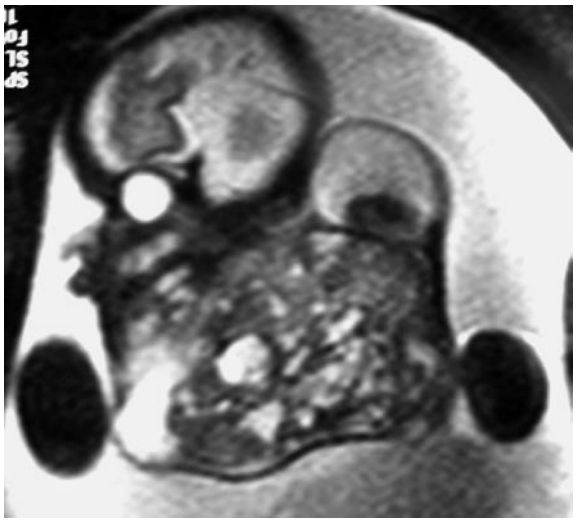

Fig. 1

On routine prenatal ultrasound at 22 weeks, a complex cystic mass was found in the cervical region of an otherwise normal-appearing fetus.

On subsequent examinations at 24 and 26 weeks, the mass was seen to increase dramatically in size. At 26 weeks, moderate polyhydramnios was noted. The remainder of the examination was normal.

MR imaging was performed to better characterize the mass (Fig. 1). At that time, the total size of the mass was larger than the fetal head. Again, polyhydramnios was noted.

- What is the most likely diagnosis, and what is the differential diagnosis?
- How should the expecting couple be counseled?
- Is intervention before birth indicated?
- How should the pregnancy be further monitored, and what might prompt early intervention? Should time, place, and/or mode of delivery be altered?
- Is neonatal intervention required? If so, how soon after delivery?
- What is the prognosis for a fetus with this condition?

A 3

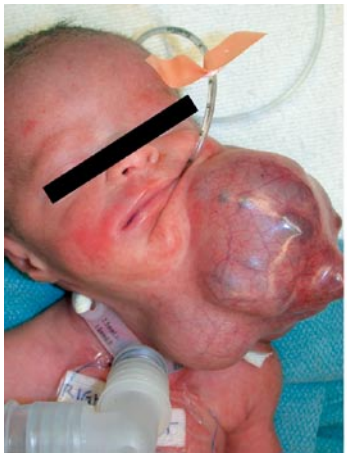

Fig. 2

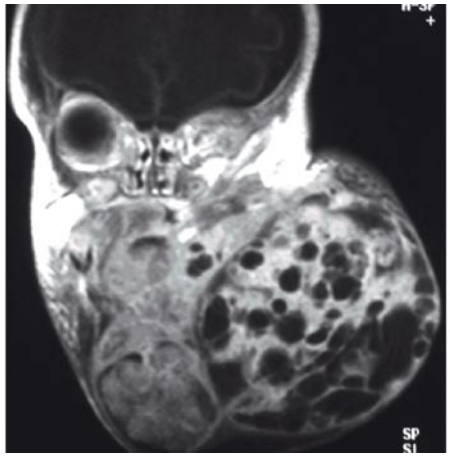

Fig. 3

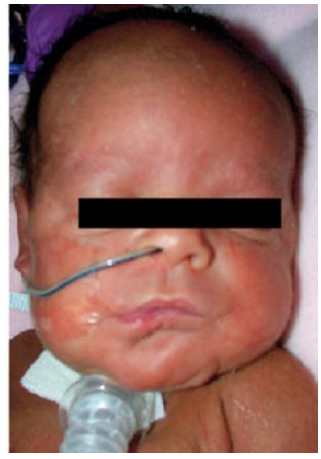

Fig. 4

The size of the lesion and its complex, cystic/solid and heterogeneous appearance are typical of a cervical teratoma. If predominantly cystic, the only other possible diagnosis would be a cystic hygroma.

Head and neck teratomas are far less frequent than pelvic and sacrococcygeal ones, and are less likely than sacrococcygeal teratomas to cause significant vascular steal, fetal hydrops, or mirror syndrome (concomitant maternal preeclampsia). However, the size and location of this lesion are likely to cause some degree of respiratory obstruction at birth. The presence of polyhydramnios suggests that fetal swallowing is impaired, causing further concern about neonatal respiratory distress.

In the past, the mortality rate of large cervical teratomas exceeded 50% because of airway obstruction at birth. In addition, the presence of polyhydramnios increases the risk of premature rupture of membranes and preterm delivery.

Because of the rapid growth of the lesion, several multidisciplinary meetings were held to plan an EXIT procedure: ex-utero, intrapartum treatment of the upper airway obstruction. This approach requires a planned and controlled C-section whereby uterine contractions are suppressed, preventing separation of the placenta. Only the head and neck of the fetus are delivered, leaving the umbilical cord in utero. Thus, an airway can be obtained while the infant remains on placental support. Once the airway is secured, the cord can be clamped and the infant delivered. This approach requires a very high level of control and collaboration between maternal-fetal medicine specialists (perinatologists), maternal anesthesiologists, pediatric surgical specialists, and neonatologists. Obtaining an airway can range from simple orotracheal intubation to rigid bronchoscopy as a temporary airway and tracheostomy or even (partial) resection of the obstructing mass. EXIT procedures of up to 60–90 min have been described, although the average duration of this procedure is about 20 min.

Because of the polyhydramnios and the risk of preterm labor, it is important to choose the time of delivery by EXIT carefully: in the present case, the mother experienced some contractions at 29 weeks, and an EXIT was performed at 32 weeks. Several days before the planned procedure, glucocorticoids were administered to the mother to accelerate lung maturation.

At delivery, the diagnosis of cervical teratoma was confirmed (Fig. 2). Intubation proved impossible, and a tracheostomy was performed.

MR imaging was performed in the ensuing days (Fig. 3), and semi-elective resection of the entire mass was performed at 8 days of life. Despite the massive distortion of normal structures, these lesions are not invasive, and symmetry is usually restored postoperatively (Fig. 4).

Q 4

Giovanni Esposito and Ciro Esposito

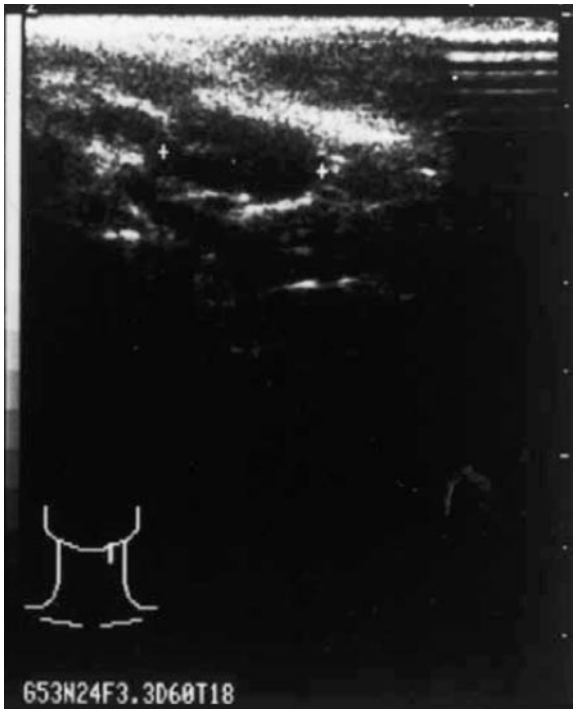

Fig. 1

A 4-year-old child without previous medical problems, apart from an episode of bilateral parotitis at the age of 3 months, was admitted to hospital after the mother noticed a swelling on the child's left cheek. After objective examination a painless mass was found with indistinct margins in the left parotid region. An ultrasound study (Fig. 1) and other examinations were performed, indicating the need for surgical intervention.

- What does the ultrasound demonstrate?
- What other examinations were performed?
- What was the suspected diagnosis?
- What was the surgical treatment?
- What was the definitive diagnosis?
- What was the follow-up?

A 4

The ultrasound shows a small hypoechogenic lesion in the left parotid region.

A fine-needle aspiration biopsy (FNAB) was carried out, which demonstrated the presence of adipocytes mixed with acinic cells at histologic examination.

The diagnosis was a lipoma of the parotid gland. The surgical treatment comprised superficial parotidectomy with conservation of the facial nerve. On the basis of a histologic examination of the removed specimen (Fig. 2), demonstrating the presence of mature adipocytes with abnormal mature and multilobular adipose tissue combined with inflammatory cells (Fig. 3), the definitive diagnosis was that of parotid lipomatosis.

The postoperative course was uneventful and the child was discharged from hospital 6 days after the operation. At the 5-year follow-up, the child had recovered completely.

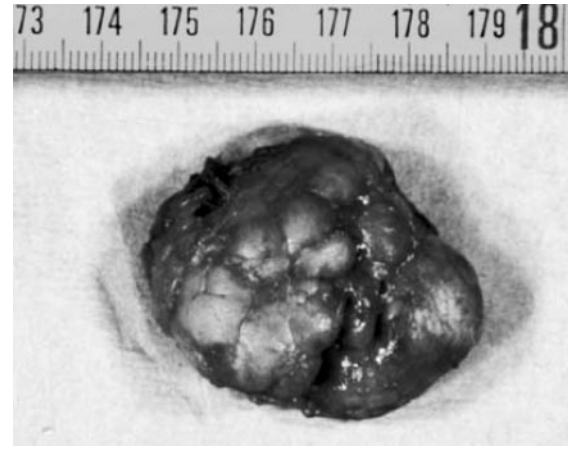

Fig. 2

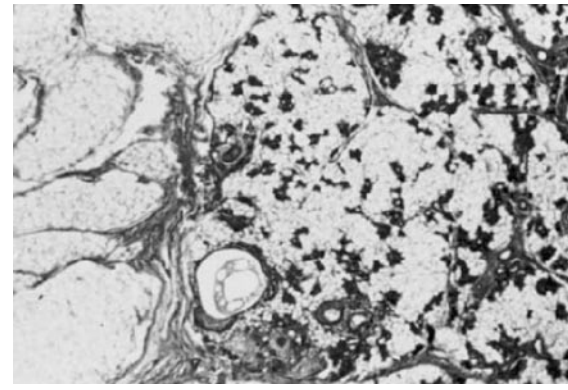

Fig. 3

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Q 5

Luigi Califano and Francesco Longo

A 13-year-old girl presented with a history of parotid swelling from 15 months, which was not related to eating. During this period she did not experience fever or pain.

When the girl presented to our department, she had already undergone many cycles of antibiotic therapy, without any result. The results of blood examinations were normal.

The surgeon asked for a CT scan with iodine medium.

- What does Fig. 1 show?
- Why did the surgeon perform this examination?
- Are there further examinations to be performed in this case?
- What pathological condition is affecting this girl?
- What is the best way to manage this condition?

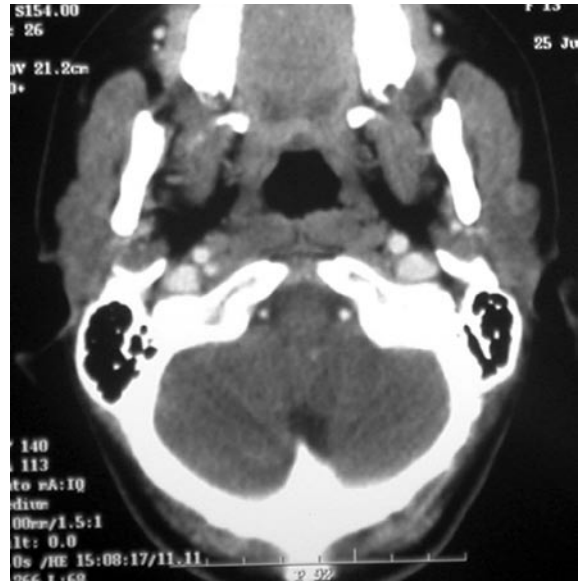

Fig. 1

A 5

This girl was affected by a parotid acinic cell neoplasm. The history and clinical examination already pointed to a parotid tumor. A CT scan with iodine medium allows one to gain more information about the precise location of the neoplasm and to define its borders and its relationships with closer structures.

Figure 1 shows a roundish, inhomogeneous lesion of 10-mm diameter, without peripheral anomalies compatible with pleomorphic adenoma or noninfiltrating neoplasms, localized in the superficial part of the parotid gland. There was no involvement of the neck lymph nodes.

For a preoperative diagnosis to be made, it is necessary to perform fine-needle aspiration biopsy (FNAB), which has very good accuracy with very little discomfort for the patient. Obtaining an almost-certain diagnosis allows one to plan the intervention with better accuracy and to prepare the patient for the surgical procedure. The FNAB in this case showed a solid proliferation of acinic cells arranged in solid blocks, suggesting an acinic cell tumor.

The patient underwent total parotidectomy under general anesthesia. A face lift approach was used on this young patient to minimize the effect of scars (Fig. 2). Moreover, in an attempt to avoid the risk of Frey syndrome, a temporalis fascia flap was used to cover the nerve and to reduce the gap left by the parotid gland excision (Figs. 3–4). No sign of facial nerve damage was evident after intervention. Histological examination of the surgical specimen confirmed the preoperative diagnosis of acinic cell tumor.

The patient was followed up every 3 months with ultrasonography and twice a year with CT in the first year, and then with ultrasonography twice a year and CT once a year. Four years after the intervention, the patient was free of disease.

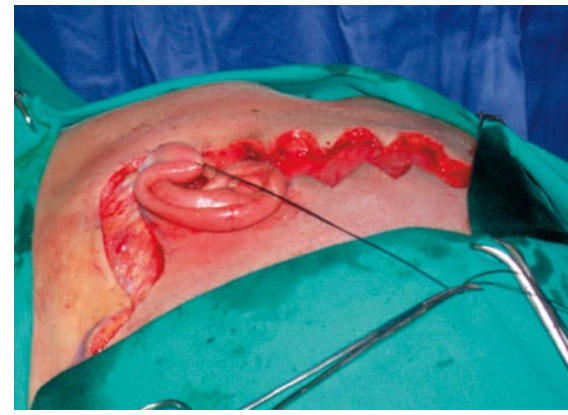

Fig. 2

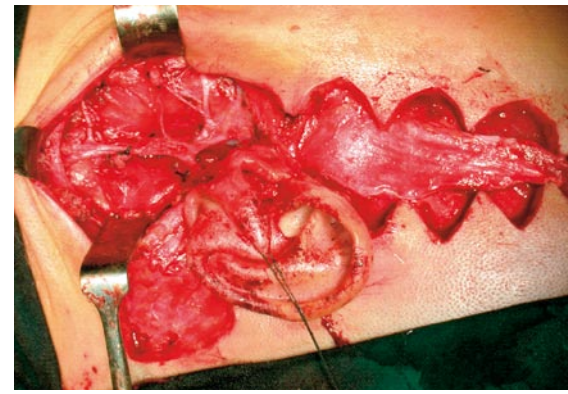

Fig. 3

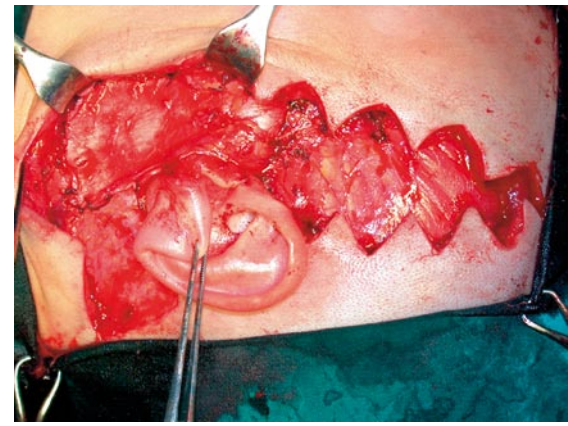

Fig. 4

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Q 6

Luigi Califano, Paola Esposito, and Francesco Longo

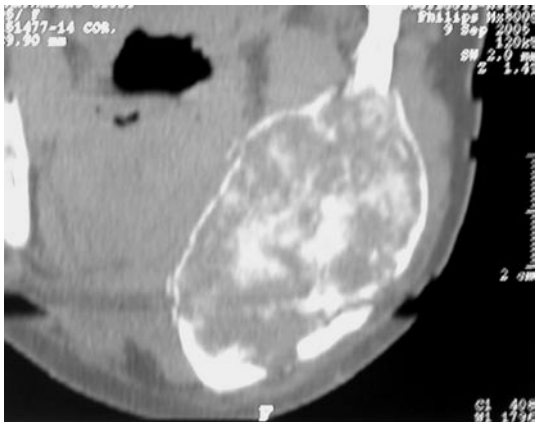

Fig. 1

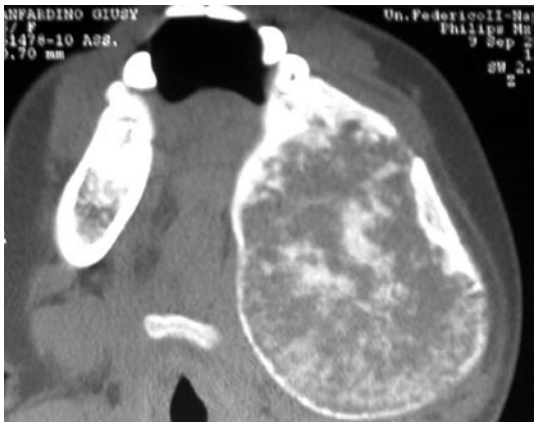

Fig. 2

A 3-year-old girl presented with mandibular swelling that had appeared 6 months earlier.

The girl had growth hormone deficit and psychomotor retardation, facial dysmorphism (telecantus), and an arachnoid cyst in the adenohypophyseal region. The child had the clinical picture of an atypical facial-cardio-cutaneous syndrome.

After the girl's mother noted swelling of the mandible, a pediatrician performed CT (Figs. 1, 2) and referred the child to a maxillofacial surgeon.

- What do Figs. 1 and 2 show?
- What other diagnostic examinations can be performed?
- What is the diagnosis?
- What is the appropriate treatment?
- What is the prognosis for this young patient?

A 6

This child is affected by fibrous dysplasia of the mandible, a condition in which normal medullary bone is gradually replaced by abnormal fibrous connective proliferation. This disease is localized in the head in less than 10% of cases and produces monostotic or polyostotic lesions, isolated or in association with McCune-Albright syndrome.

This disease most commonly presents as an asymptomatic, slow enlargement of the involved bone. The monostotic form is the most common (80%) and more frequently affects the ribs, long bones, pelvis, jaws, and skull. Maxillary lesions occur more frequently than mandibular ones.

Fibrous dysplasia characteristically occurs during the first and second decade of life and becomes stable after puberty.

In the classic presentation, as shown in Figs. 1 and 2, the radiographic appearance is described as homogeneous radio-opacity with numerous trabeculae of woven bone imparting a ground glass appearance. Lesions of fibrous dysplasia may also present as unilocular or multilocular radiolucencies. There are no alterations in laboratory values.

A three-dimensional CT scan can be very useful in showing the complete extension of the disease and in planning preoperatively the margins of surgical intervention.

Typical histological features are represented by proliferation of fibrous connective tissue with trabeculae of immature bone tissue. A histological differential diagnosis with osteoma, fibroma, or Paget's disease can be difficult to make without the correct interpretation of clinical and radiological findings.

Once a jaw lesion is diagnosed, the extent of skeletal involvement should be investigated, with plain radiographs, total-body CT, or scintigraphy. In our case, there was only one lesion.

Because of the stabilization of growth after puberty, the treatment consists only of biopsy for confirmation of the diagnosis and periodic follow-up for small lesions and bone recontouring via a transoral approach to treat functional or cosmetic disability (Figs. 3, 4). En bloc resection is unnecessary considering the lesion's slow-growing and non-neoplastic nature.

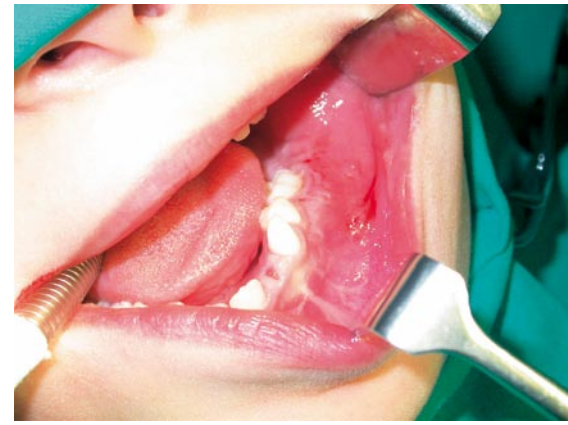

Fig. 3

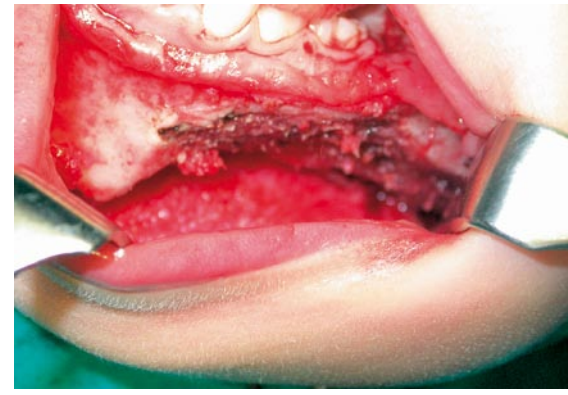

Fig. 4

The incidence of malignant transformation is very rare, less than 1%, and is related to cases in which patients were treated with radiotherapy, which should always be avoided considering the benign nature and limited growth of the lesion.

Q 7

Luigi Califano and Francesco Longo

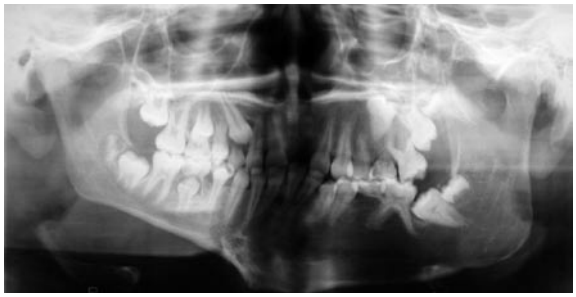

Fig. 1

An 11-year-old girl presented with a history of facial asymmetry that had started 18 months previously with dental crowding.

The patient was in general good health and had menarche when she was 10 years old. She did not complain of any pain or discomfort.

When the girl presented to our department, she had a deviation of the chin and a cross-bite and had already started orthodontic treatment without any result.

The surgeon asked for a panoramic plain radiograph followed by CT with three-dimensional reconstruction.

- What do Figs. 1–3 show?
- Should further examinations be performed in this case? If so, why?
- What condition is affecting this girl?
- What is the best way to manage this pathological condition?

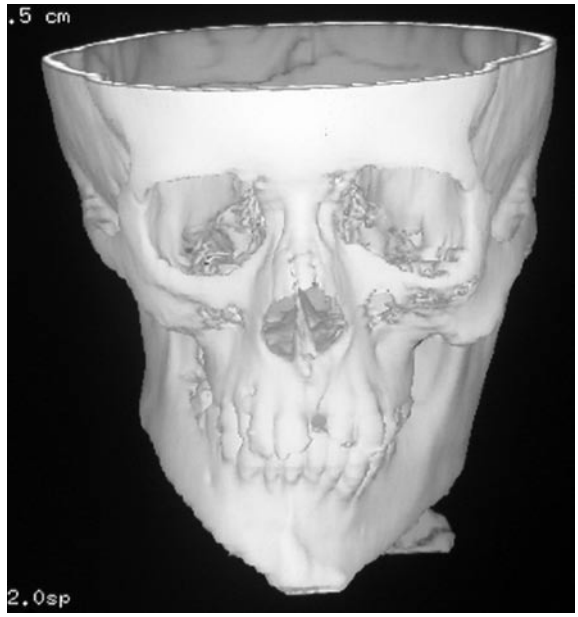

Fig. 2

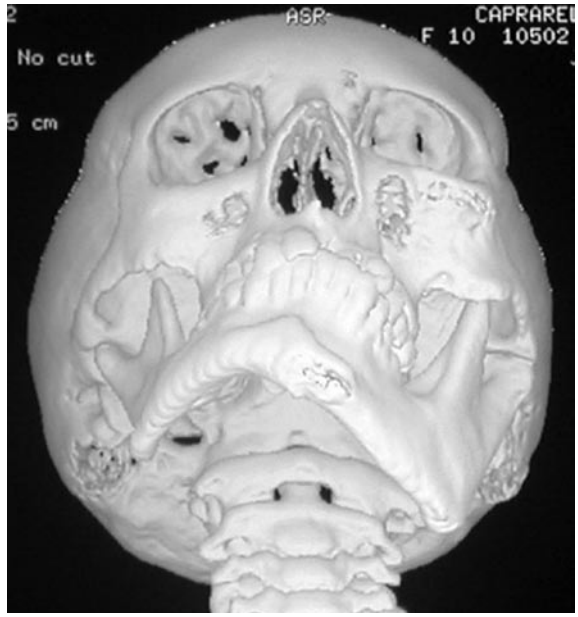

Fig. 3

A 7

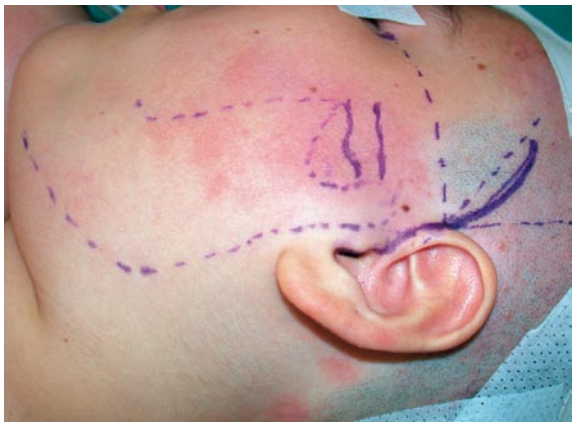

Fig. 4

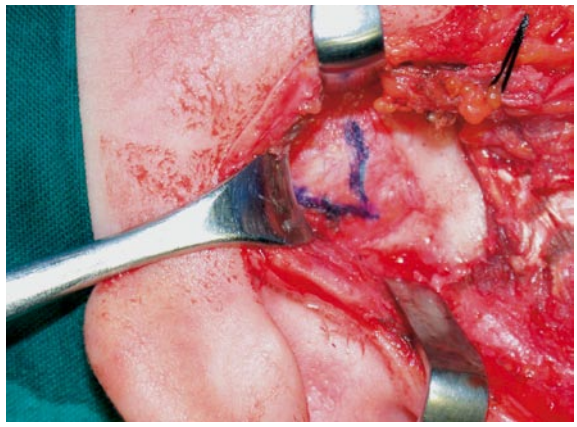

Fig. 5

This girl was affected by hemimandibular hyperplasia. Hemimandibular hyperplasia is a developing anomaly of the mandible which appears at puberty and often continues in the following years. Its features are asymmetry of the lower third of the face, deviation of the mandible to the opposite side of the affected hemimandible, and secondary growth anomaly of the maxilla.

A panoramic plain radiograph (Fig. 1) shows an elongation and enlargement of one side of the mandible, i.e., enlargement of the condyle, condylar neck and ascending and horizontal rami; the head of the condyle can be normal in cases of slow growth or enlarged in cases of rapid evolution. A cephalometric study shows variable dental compensation in the sagittal and vertical planes. CT with three-dimensional reconstruction (Figs. 2, 3) clearly shows the hemimandibular enlargement.

To manage correctly these patients, it is very important to collect all clinical data. In fact, a history of rapid changes suggests an active growth, while a history showing an old presentation and slow changes indicates inactivity or reduced activity of the condylar process. Bone scans are very useful for confirming an active growth, but there is a high risk of false-positive findings.

In cases of active growth, as in our case, condylectomy is the treatment of choice. The patient needs close follow-up and orthodontic treatment to prepare for a two-jaw surgery if the maxilla is also affected.

Our patient underwent condylectomy via a rhitidectomy approach to minimize scars and avoid any risk of facial nerve damage (Fig. 4). An intraoperative view of the condyle is presented in Fig. 5.

The patient was followed up for 3 years, with progressive improvement of the occlusion and facial symmetry.

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Q 8

Luigi Califano and Francesco Longo

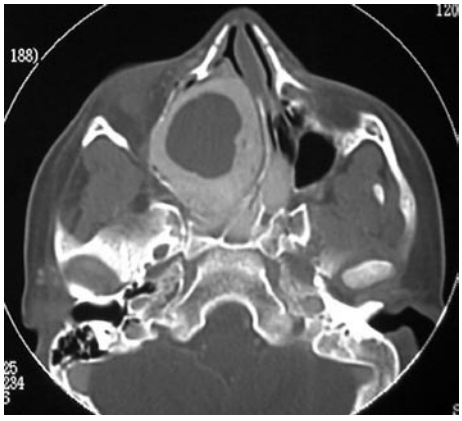

Fig. 1

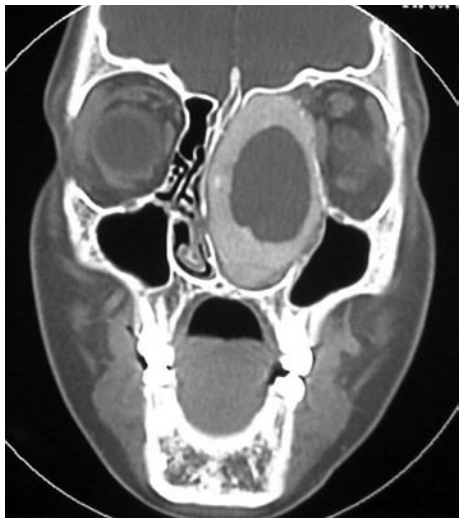

Fig. 2

A 14-year-old boy presented with facial asymmetry, nasal obstruction, and exophthalmus that had started 1 year previously.

The patient was in general good health. He did not complain of any pain, but reported diplopia.

To evaluate the airway status, the surgeon asked for a CT scan.

- What do Figs. 1 and 2 show?
- What is the diagnosis?
- What treatment plan should be followed?

A 8

The patient had juvenile aggressive ossifying fibroma. Juvenile aggressive ossifying fibroma occurs in young patients generally before the age of 15 years. The most common sites of occurrence include the maxilla, frontal bones, ethmoid bones, and paranasal sinus.

At clinical presentation, facial asymmetry, nasal obstruction, and exophthalmus with visual disturbances are generally present. The tumor presents a progressive and often rapid enlargement with thinning and erosion of adjacent bone.

The CT studies show an expansile, destructive, radiopaque, or hyperintense nonhomogeneous lesion (Figs. 1, 2).

The approach to juvenile aggressive ossifying fibroma is surgical. Depending on the size, location, and extension of the tumor, the goal of treatment should be complete excision, which in many cases requires an aggressive surgical procedure. En bloc resection should be evaluated in cases of large or recurrent lesions and can cause significant deformities especially in the case of orbital or cranial involvement.

Close follow-up is mandatory, since the reported recurrence rate for this tumor is between 30% and 60%; however, no case of metastasis has been reported.

Our patient underwent submental intubation to avoid the presence of an anesthesia tube in the operative field. The lesion was reached and removed via a bipartite Le Fort 1 fracture (Fig. 3). The fracture lines were fixed with biodegradable plates to avoid any alteration of the residual growth of the maxilla (Fig. 4).

The patient was followed-up for 3 years with clinical examinations and CT every 4 months, and at the time of writing was free of disease (Fig. 5).

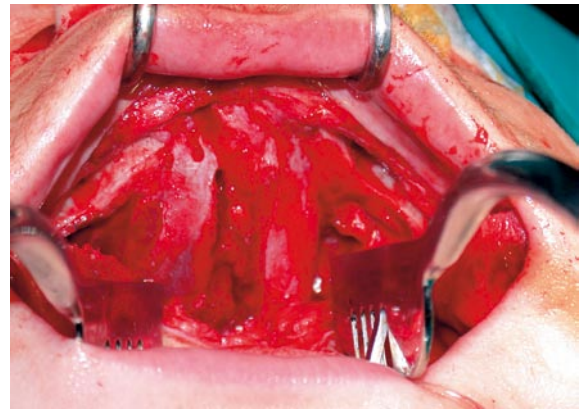

Fig. 3

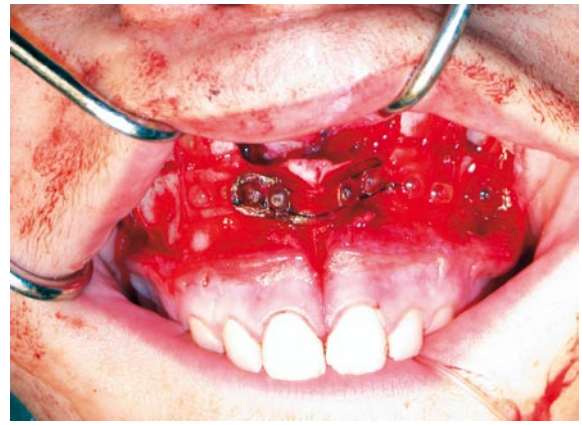

Fig. 4

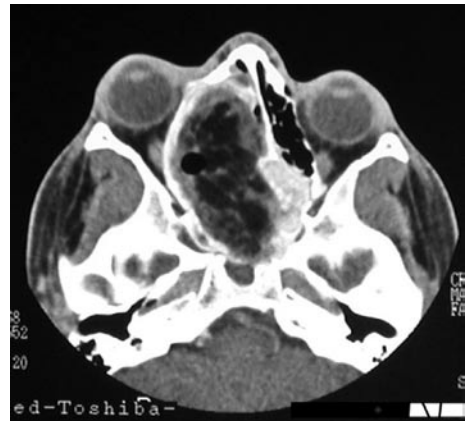

Fig. 5

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Q 9

Craig T. Albanese

A routine screening ultrasound at 20 weeks' gestation was performed (Fig. 1) revealing a large mass (*arrows*) protruding from the fetal oral cavity (*open arrow*). The exact origin could not be delineated by this examination. There were calcifications (*arrowhead*) in the mass.

MR imaging (Fig. 2) was performed, which confirmed the diagnosis of an oral cavity-based mass (*arrow*). There was polyhydramnios (*asterisk*) secondary to the obstructing mass in the oral cavity.

- What is the diagnosis?

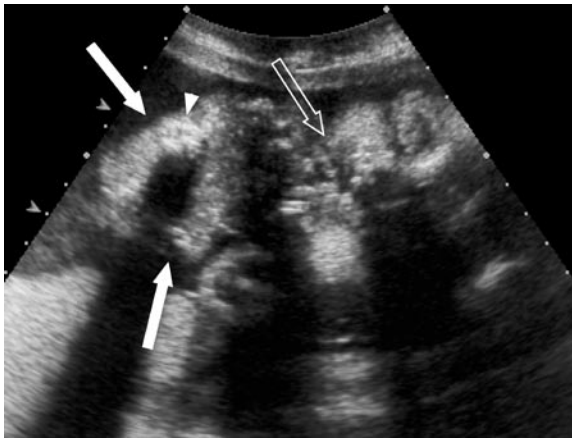

Fig. 1

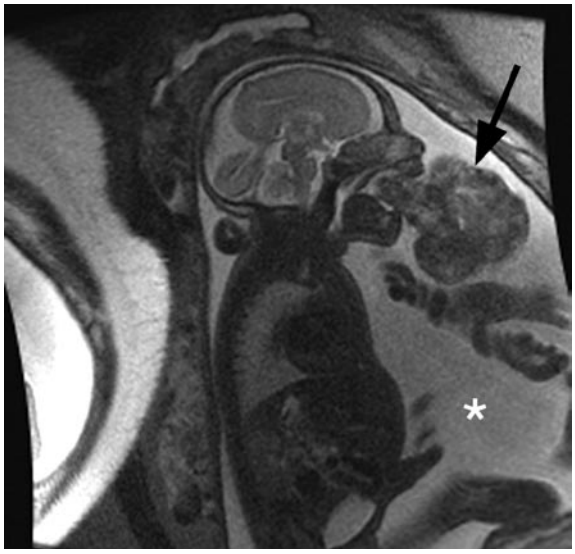

Fig. 2

A 9

This large mass with calcifications is most consistent with an oral teratoma (epignathus).

Because of the risk of airway compromise, the fetus was delivered using the EXIT (ex utero intrapartum treatment) strategy. During the procedure, the uterus is opened and the mass is removed while the fetus is still connected to the placenta. After resecting the mass (Fig. 2) that was emanating from the area of the hard palate, orotracheal intubation was performed, the umbilical cord cut, and the baby delivered. The child recovered uneventfully.

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Q 10

Masayuki Fujioka, Carl Muroi, Nadia Khan, and Yasuhiro Yonekawa

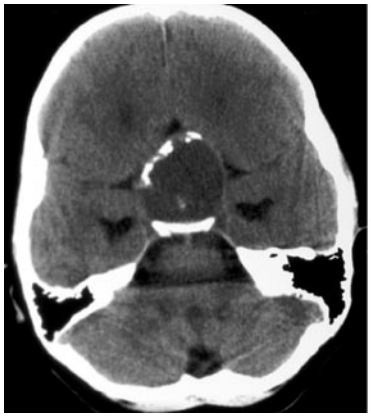

Fig. 1

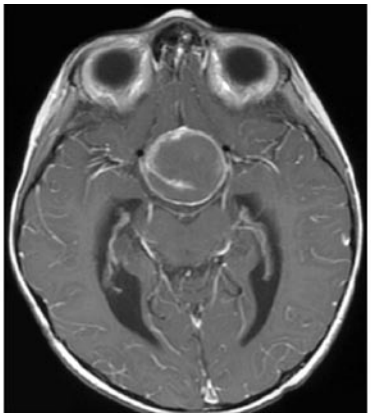

Fig. 2

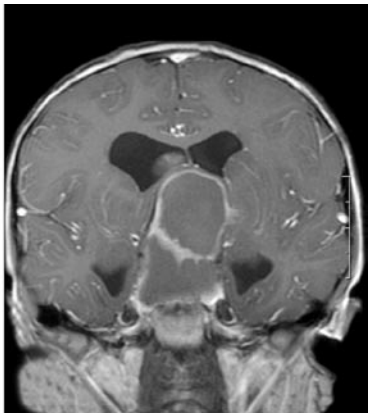

Fig. 3

A 6-year-old girl suffered from headaches, poor appetite, general fatigue, and projectile vomiting for 3 days.

According to her father, the girl had recently been saying that it was dark even during the day. The presence of increased intracranial pressure and visual disturbance was suspected.

On admission to hospital, the bilateral light reflex (direct and indirect) was weak. CT scanning of the head was performed (Fig. 1).

She had decreased skin turgor suggestive of dehydration associated with polyuria (low specific gravity). Her blood glucose level was normal and the plasma natrium level was moderately increased. She also presented with growth failure and short statue.

- What does Fig. 1 show?
- What pathophysiology should be suspected?
- What does the MR imaging study (Gd-T1-weighted) of the head show (Figs. 2–4)?

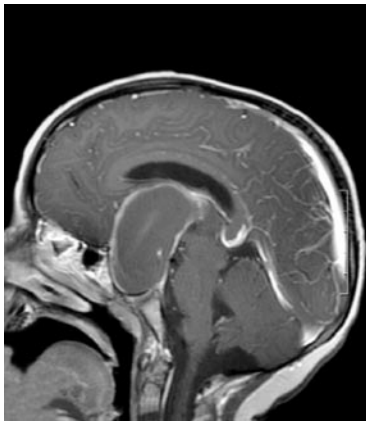

Fig. 4

- What diagnosis can be considered?
- What treatment should be performed?

A 10

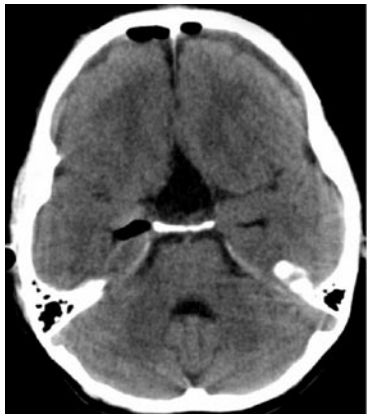

Fig. 5

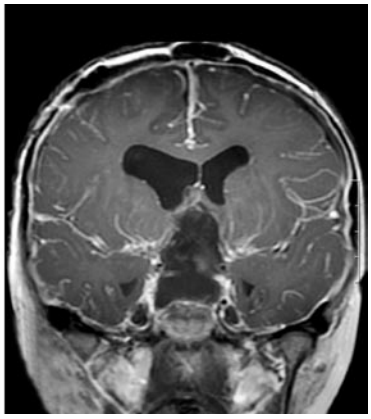

Fig. 6

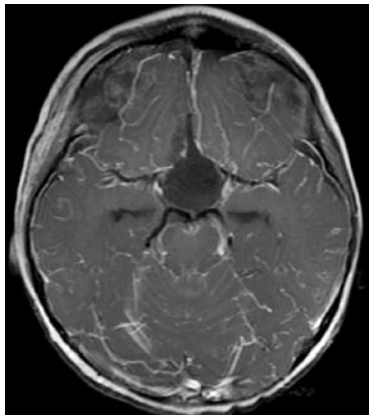

Fig. 7

The CT scan (Fig. 1) and T1-weighted MR images with administration of gadolinium-diethylenetriamine pentaacetic acid (Gd-DTPA; Figs. 2–4) reveal a cystic tumor extending from the sella turcica through the suprasellar cistern to the third ventricle. The CT scan shows calcification in some portions of the cyst wall. The tumor cyst wall is enhanced on the Gd-T1-weighted MR image. The MR images show dilatation of the bilateral lateral ventricle due to the obstruction of the foramen of Monro (obstructive hydrocephalus).

The differential diagnosis for brain tumor in this region includes craniopharyngioma, germ cell tumor, pituitary adenoma, glioma, and meningioma.

The simultaneous presence of dehydration and excessive urination of low specific gravity suggests diabetes insipidus. In addition, the patient had growth failure suggestive of pituitary dwarfism. The endocrinological study showed hypopituitarism with decreased blood levels of growth hormone, insulin-like growth factor-1 (somatomedin-C), and thyroid-stimulating hormone.

Craniopharyngioma is the most probable diagnosis for a suprasellar cystic tumor with partially calcified wall, leading to hypopituitarism in this 6-year-old girl. The tumor occupying the third ventricle was totally removed, without additional deficits, via an interhemispheric transrostrum corporis callosi and lamina terminals approach with the patient in the supine position.

The postoperative CT scan and MR images (Figs. 5–7) demonstrate the total removal of the tumor. The histological diagnosis is adamantinomatous craniopharyngioma. The characteristic features include peripheral palisading of nuclei, loose arrangements of squamous cells, and nodules of keratin and calcification.

In general, the features of craniopharyngioma on CT images include cyst formation with calcification and enhancement effects of the cyst wall and solid portion. The signal behavior of craniopharyngioma on T1-weighted images varies according to its cystic contents (hypo-, iso-, and hyper-intensity). The increased intensity on T1-weighted images seems to result partly from the high concentration of liquid cholesterol. On T2-weighted images, craniopharyngiomas are commonly hyperintense. The solid portions are heterogeneously enhanced and the cyst wall is strongly enhanced on Gd-T1-weighted images.

Postoperative hormonal replacement therapy was initiated with administration of adrenal corticosteroid, thyroid hormone, and antidiuretic hormone. After 6 months, the patient had improved visual acuity (right 0.2 and left 0.6) and no apparent recurrence of craniopharyngioma.

Q 11

Giovanni Esposito, Aurelie Chiappinelli, Gianfranco Vallone, and Ciro Esposito

An infant weighing 3,350 g, born at term after a normal pregnancy and delivery, had a mass in the submandibular left region, the size of a walnut, in front of the sternocleidomastoid muscle. On US the mass appeared to be a characteristic lymphangioma, and therefore it was decided not to treat the lesion but to follow its evolution.

Because the mass increased in size, hospitalization was decided when the child was 3 months old.

On admission, the child was in good condition, without any local or general anomalies except for the presence of the mass.

The left submandibular region was deformed by an irregular ovoidal swelling with a maximum diameter of 8 cm, showing partly cystic and partly solid consistency.

Laboratory test results were normal.

After another US study, an intervention was decided on (Fig. 1).

- What does Fig. 1 show?
- What was the diagnosis?
- What was found during the intervention and what was performed?
- What was the definitive diagnosis?
- What was the follow-up?

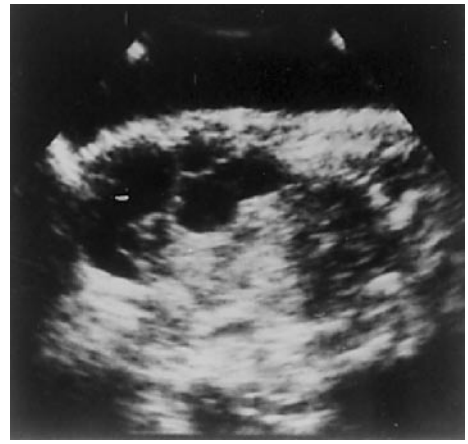

Fig. 1

A 11

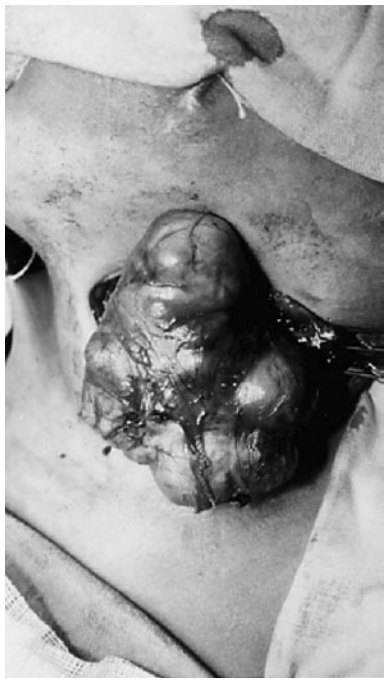

Fig. 2

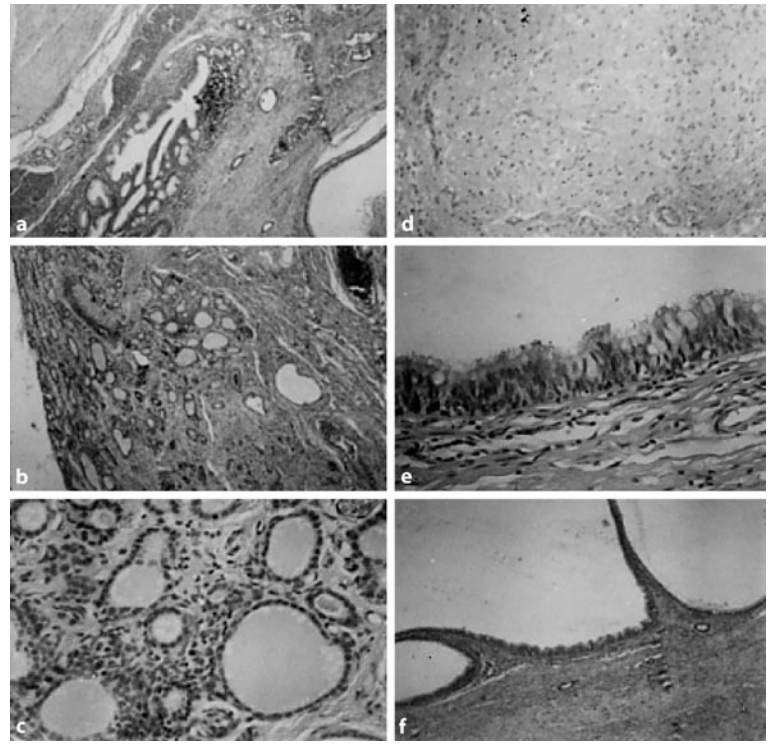

Fig. 3

The US shows a mass with a complex structure characterized by multiple eccentric fluid areas around a central solid irregular area.

The diagnosis was cervical teratoma.

At surgery, the multicystic mass was reached via a transversal cut along its maximum diameter. The mass was excised with some difficulties because it extended to the back of the trachea reaching the paravertebral space. The mass, measuring 6×4×2.5 cm, showed multiple small cysts around a central solid area (Fig. 2).

On the basis of the histologic examination, which showed embryogenic tissues including mainly mature thyroid tissue, the definitive diagnosis was that of thyroid teratoma (Fig. 3).

The postoperative course was uneventful, and at follow-up after 2 years the boy was in good condition without any relapse.

Q 12

Ugo De Luca

A 9-year-old boy had an asymptomatic, midline anterior cervical mass initially interpreted by his parents as the Adam's apple (Figs. 1, 2). The mass was mobile during swallowing, firm, and it had well-defined margins. The growth had been slow and no infections occurred.

The patient underwent ultrasonography (Fig. 3), which revealed a hypoechoic cystic mass very close to the hyoid bone.

- What is the diagnosis?
- What is the differential diagnosis?
- What is the treatment?
- What complications can be expected?

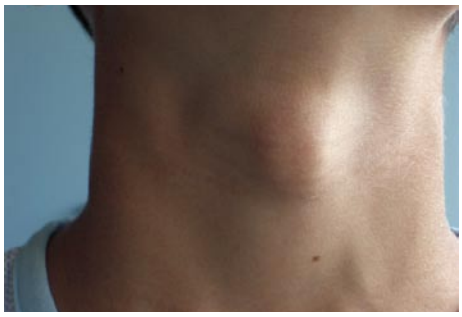

Fig. 1

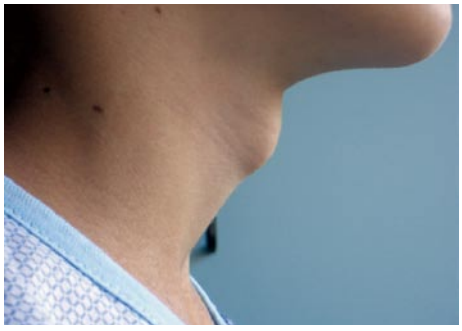

Fig. 2

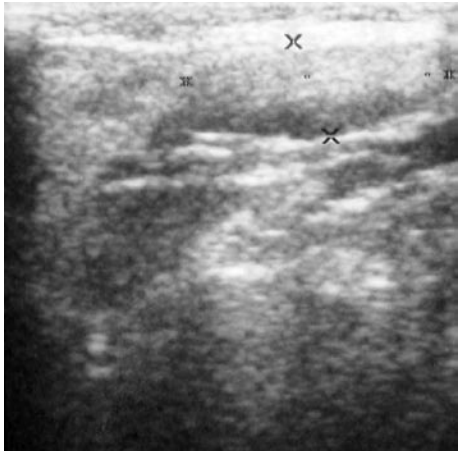

Fig. 3

A 12

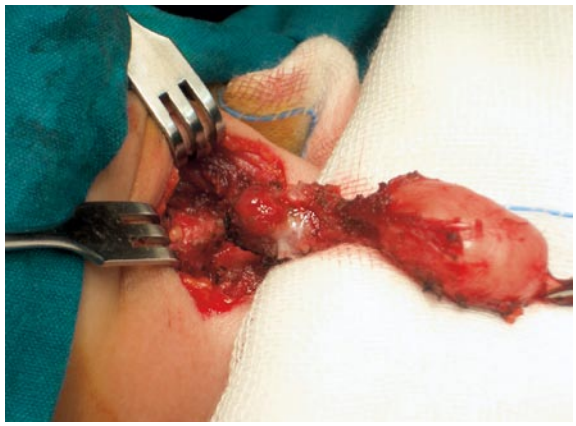

Fig. 4

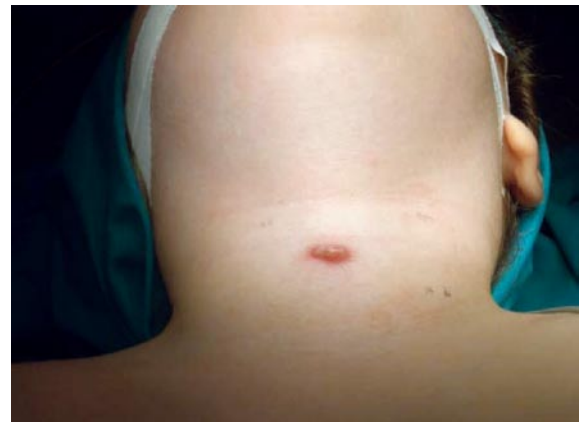

Fig. 5

The midline cervical mass was a large thyroglossal duct cyst (TDC). It did not become infected and slowly increased in volume.

The differential diagnosis must include lymph node hyperplasia and dermoid cyst.

The treatment of choice for TDC is surgery via the Sistrunk procedure. Once the surgeon has isolated the cyst through a transverse cervical incision, the hyoid bone body (1–2 cm) must be resected together with the cyst, and the dissection should be continued until the foramen cecum at the base of the tongue (Fig. 4).

Accurate hemostasis must be achieved and closure in layers realized. No drainage is necessary and the child can be discharged on the same day of surgery.

Complications of unoperated TDC are recurrent infections, fistulization (Figs. 5, 6) to the skin with consequent retracting scar, and possible, although rare, cancerization.

Complications of operated cases are postoperative infection and recurrence of the cyst, which is very high if the Sistrunk procedure is not radical; the recurrence rate is 4% in complete Sistrunk operations and rises to 20% if several cyst infections precede the radical operation.

Moreover, attention must be paid to the distal portion (toward the pyramidal lobe of the thyroid gland) of the thyroglossal duct, if present; if this is ignored, it could be responsible for relapse.

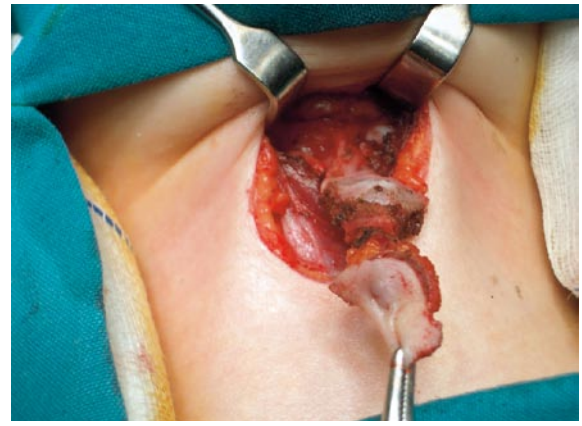

Fig. 6

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Q 13

Nancy Rollins

A 13-month-old girl presented with a slow-growing, soft, purplish mass involving the left buccal surface and lip (Fig. 1). The lesion was seen at birth as a small bluish region. The parents were told the lesion represented a small hemangioma which would involute with time.

- What is the differential diagnosis?
- What is the best imaging strategy?
- Should the lesion be biopsied or resected?
- Is there a nonsurgical alternative for treatment?

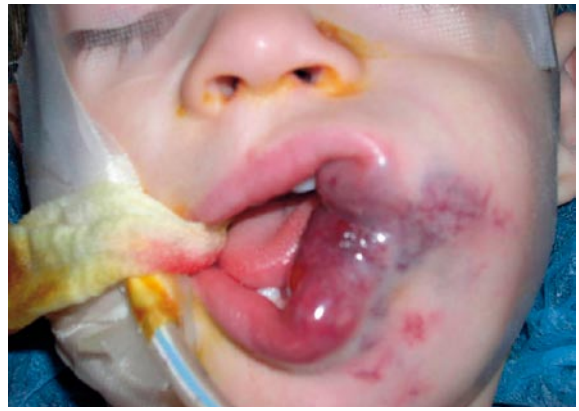

Fig. 1

A 13

The purplish lesion is a venous malformation. The patient underwent MR imaging using gadolinium-DTPA. An axial short tau inversion recovery (STIR) sequence (Fig. 2) and a post-contrast T1 spin echo (Fig. 3) sequence through the lesion were performed. The lesion is well-circumscribed, nonlipomatous, with moderately intense enhancement and no dilated feeding arteries. The lesion represents a congenital venous malformation. The patient underwent sclerotherapy with absolute alcohol repeated four times 4–6 weeks apart (Figs. 4, 5). Figure 6 shows the final result. Residual cutaneous discoloration was treated with laser.

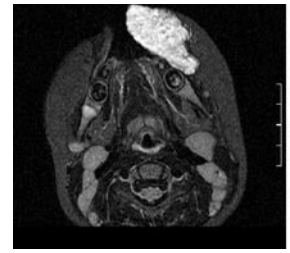

Fig. 2

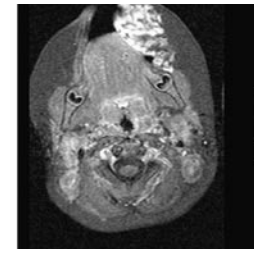

Fig. 3

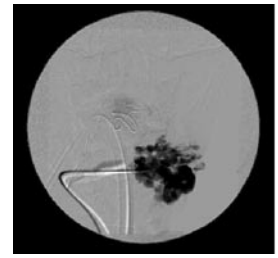

Fig. 4

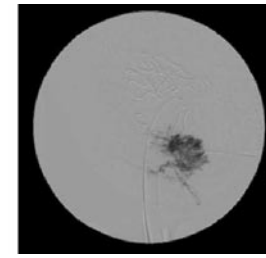

Fig. 5

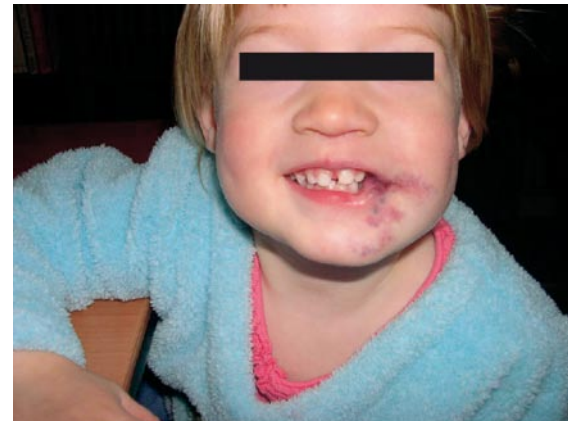

Fig. 6

The venous malformation should not be confused with a hemangioma. Attempts at surgical resection would be disfiguring and unlikely to affect a complete cure. Biopsy is contraindicated. The patient was therefore referred for sclerotherapy. Prior to sclerotherapy, cross-sectional imaging is mandatory and is best performed with MR imaging. MR with contrast is needed to define the composition of the lesion; i.e., venous, lymphatic, or mixed, as well as to assess the extent of the lesion and the proximity to neurovascular structures. Lesions that have a sharp abrupt transition from the surrounding tissue respond better to sclerotherapy than do lesions with ill-defined margins that cross tissue and fascial planes. Sclerotherapy is the injection of aqueous or oleic solutions into abnormally dilated vascular or lymphatic channels inducing damage to the endothelial lining and resulting in thrombosis, fibrosis, stenosis, and local scarring. Residual cosmetic deformity due to formation of scar tissue within the venous malformation is treated with surgery. Blood loss is minimized by the sclerotherapy and complete resection of the lesion is more readily accomplished. For deeper venous malformations that present with pain or functional impairment in the absence of cosmetic deformity, subsequent surgery is not usually needed. Large multi-compartmental venous malformations of the face and extremities often have phleboliths, calcified blood clots that are diagnostic of venous malformations and which are not seen in hemangiomas. These large lesions are usually not curable and treatment in the form of sequential sclerotherapy is designed to decrease pain and promote full functionality. Venous malformations usually demonstrate significant growth with the onset of puberty, especially in female subjects.

There are multiple agents used for sclerotherapy including bleomycin, sodium tetradecyl, ethibloc (ethanolamine oleate), absolute alcohol, and doxycycline. Bleomycin is a chemotherapeutic agent known to cause pulmonary fibrosis and should be avoided for nonmalignant disease in children. Absolute alcohol denudes the endothelial cells upon direct contact with the vessel wall. Permanent damage to the vessel wall results in complete obliteration of the vessel lumen, which prevents recanalization. Sclerosing agents are usually mixed with nonionic

contrast and the installation of the sclerosant should be performed under fluoroscopic guidance to limit complications resulting from extravasation of the sclerosing agent into the regional soft tissue or draining veins.

Complications of sclerotherapy using absolute alcohol include skin necrosis, neuropathy, muscle atrophy, and cardiovascular collapse. Absolute alcohol is particularly dangerous, and installation of absolute alcohol into venous structures should be performed with careful monitoring of the patient's vital signs by the anesthesiologist. No more than 1.0 cc/kg ethanol should be given during sclerotherapy. Reported complications include respiratory depression, cardiac arrhythmias, seizures, rhabdomyolysis, hypoglycemia, and death. The treated site undergoes marked often very painful swelling.

Doxycycline is a tetracycline derivative commonly used for malignant pleural effusions via direct intrapleural injection. Animal studies have shown that doxycycline can induce a marked decrement in neural function when applied to the subepineural layers of the sciatic nerve in the rat, and phrenic nerve paralysis after intrapleural installation of doxycycline has been reported. Caution should be used therefore when a facial venous malformation involves the distribution of the facial nerve.

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2

Thorax

Introduction

Pediatric surgical pathology in the chest can occur in the heart, the lung, the mediastinum, or the chest wall. Mechanisms of disease can range from congenital anomalies, infectious or inflammatory conditions, trauma, neoplasia, or disorders of organ function. This section addresses a wide variety of thoracic problems encountered by the pediatric surgeon, and gives examples of how these problems can be diagnosed and managed effectively.

The diagnosis of thoracic surgical problems starts with a careful history and physical examination. For many pulmonary, tracheal, and mediastinal problems, attention to the subtleties of breathing patterns, stridor, chest wall movement and symmetry, and auscultation will permit the surgeon to narrow down the diagnostic alternatives before any imaging is done. Similarly, it is on the basis of the clinical evaluation of the patient that the clinician will be able to prioritize interventions. The "ABCs" (rapid evaluation of ***a***irway, ***b***reathing, and ***c***irculation) are more important in thoracic diseases than in any other part of the body.

There are several levels of imaging commonly used for the evaluation of thoracic disease. Chest radiography is one of the oldest and most commonly used modalities, and will often reveal everything the surgeon needs to know. In many cases the chest radiograph may be more sensitive than physical examination for identifying lung pathology, air or fluid in the pleural space, and mediastinal masses. More recently, the use of computerized tomography has provided additional information and sensitivity over plain chest radiography, but at the expense of significantly higher radiation dose, which must be kept in mind when using this modality. Ultrasound has had limited utility in the chest, because the air in the lungs interferes with the sound waves. However, for evaluation of the heart, blood flow in vessels, and pleural fluid collections, ultrasound offers excellent imaging without radiation. Magnetic resonance imaging also has the advantage of providing excellent images without radiation, but it has been underutilized in the chest. This is an area for future development.

More invasive diagnostic modalities include bronchoscopy, with or without bronchoalveolar lavage, image-guided needle and core biopsies, and video-assisted thoracoscopic surgery, which of course is increasingly being used for therapeutic purposes as well.

Finally, the widespread use of prenatal diagnostic techniques, ranging from the maternal alpha-fetal protein test to sophisticated ultrasound, has resulted in identification of many thoracic anomalies before birth. This has provided the opportunity for counseling, altering the location or mode of delivery, and in some cases performing fetal interventions for life-threatening conditions.

Q 14

Jürgen Schleef

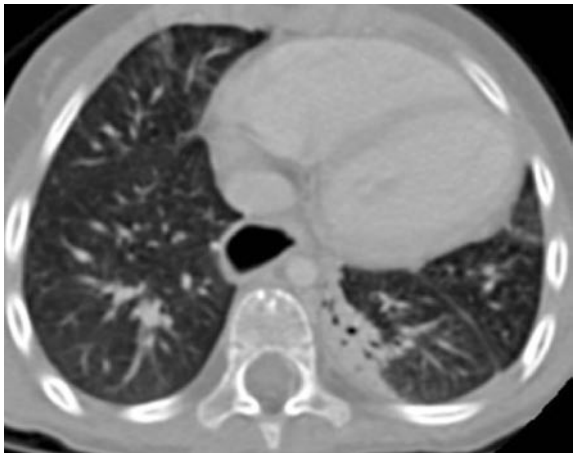

Fig. 1

A 4-year-old girl had a history of recurrent pulmonary infections from the age of 20 months, which were usually treated with antibiotics.

The mother informed the pediatrician that the girl had a prenatal diagnosis of a small lung malformation, but that a plain radiograph of the chest at the age of 1 year was normal. No further investigations were performed.

The girl was brought to our hospital again with fever and a cough. The white blood cell count was pathologic at 22,000; the CRP value was 8.2.

The doctor in the emergency department decided, on the basis of the history provided by the mother, to ask for a CT scan.

- What does Fig. 1 show?
- Why did the surgeon in the emergency department decide to perform a CT scan?
- What condition is affecting this child?
- How should this condition be treated?

A 14

Figure 1 shows a CT scan of the thorax. In the inferior part, a pulmonary malformation (congenital cystic adenomatoid malformation; CCAM) is visible. The radiologist described this malformation and ruled out extrapulmonary sequestration, since no separate blood supply from the inferior aorta could be identified.

The history of the child is typical of a connatal condition (pulmonary malformation). In most of these cases, a chest radiograph is not indicated to rule out a postnatal persisting malformation. A CT scan (or MR imaging) is mandatory in every case.

CCAM is a condition that is frequently found at prenatal ultrasound examination. If this pathological condition is not treated, persistent pulmonary infections resistant to antibiotic treatment are very frequent. In rare cases, an associated malignancy has been described in the literature.

The therapy is usually surgical. In extrapulmonary sequestration, an embolization of the supplying vessel is proposed by some authors as an alternative treatment.

In this case an atypical resection of the malformation was performed by thoracoscopy. Figure 2 shows the thoracoscopic view, while Fig. 3 shows the postoperative CT scan.

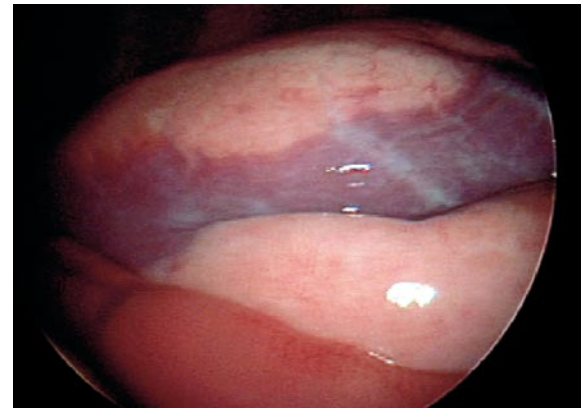

Fig. 2

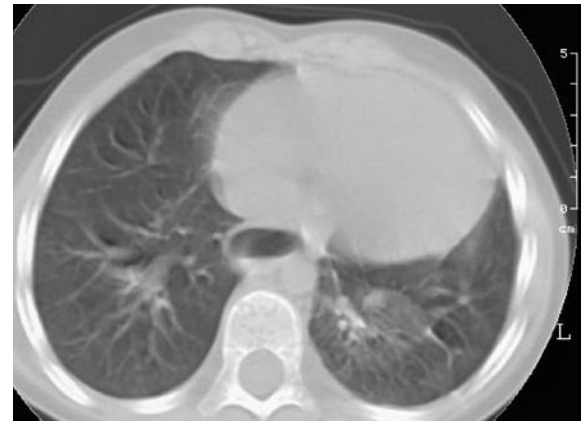

Fig. 3

Suggested Reading

1. Davenport M, Warne SA, Cacciaguerra S, Patel S, Greenough A, Nicolaides K. Current outcome of antenatally diagnosed cystic lung disease. J Pediatr Surg 2004; 39(4):549–56
2. Gornall AS, Budd JL, Draper ES, Konje JC, Kurinczuk JJ. Congenital cystic adenomatoid malformation: accuracy of prenatal diagnosis, prevalence and outcome in a general population. Prenat Diagn 2003; 15; 23(12):997–1002
3. Illanes S, Hunter A, Evans M, Cusick E, Soothill P. Prenatal diagnosis of echogenic lung: evolution and outcome. Ultrasound Obstet Gynecol 2005; 26(2):145–9
4. Pai S, Eng HL, Lee SY, Hsiao CC, Huang WT, Huang SC Rhabdomyosarcoma arising within congenital cystic adenomatoid malformation. Pediatr Blood Cancer 2005; 45(6): 841–5
5. Shanmugam G, MacArthur K, Pollock JC. Congenital lung malformations–antenatal and postnatal evaluation and management. Eur J Cardiothorac Surg 2005; 27(1):45–52

Q 15

Jürgen Schleef

An 11-year-old girl was brought to our hospital with a chest wall deformity. The mother reported that the girl was growing normally, but the deformity, which had been present since birth, was getting worse and the pectus had become "deeper" in the last year.

The girl was very shy, avoided sport activities, and told the surgeon that she sometimes felt short of breath.

The surgeon decided, on the basis of the history supplied by the mother, to ask for a CT scan of the thorax, an electrocardiogram (ECG), echocardiography, and a pulmonary function test.

- What does Fig. 1 show?
- Why did the surgeon ask for these examinations?
- What pathological condition is affecting this child?
- How should this condition be treated?

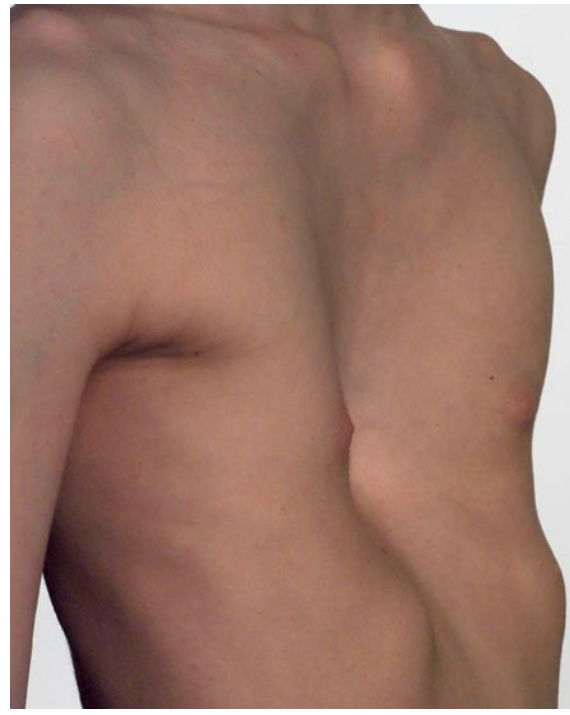

Fig. 1

A 15

Figure 1 is a picture of the chest of an 11-year-old girl. The deformity is a typical pectus excavatum. Note the retraction of the sternum.

Figure 2 is a CT slice of the thorax as requested by the surgeon. It shows the typical configuration of a pectus excavatum with a left side shift of the heart. The anterior thoracic wall is retracted.

The surgeon asked for the examination to rule out further associated diseases. In most cases, the heart or the lungs are not affected.

The treatment of this condition can be surgical. Different techniques are described in the literature. The most popular technique is a thoracoscopic-assisted approach with retrosternal bar implantation, as described by Donald Nuss. Figure 3 shows the clinical result 2 years after surgery on this patient.

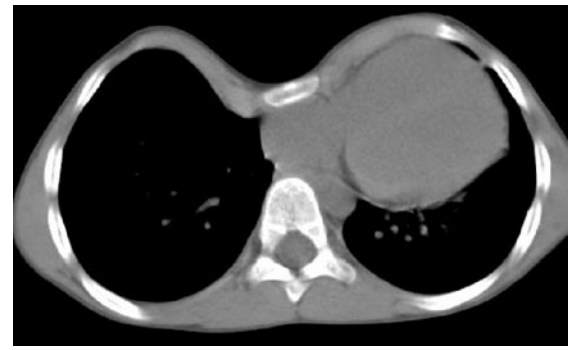

Fig. 2

Fig. 3

Suggested Reading

1. Croitoru DP, Kelly RE Jr, Goretsky MJ, Gustin T, Keever R, Nuss D. The minimally invasive Nuss technique for recurrent or failed pectus excavatum repair in 50 patients. J Pediatr Surg 2005; 40(1):181–6; discussion 186–7
2. Goretsky MJ, Kelly RE Jr, Croitoru D, Nuss D. Chest wall anomalies: pectus excavatum and pectus carinatum. Adolesc Med Clin 2004; 15(3):455–71
3. Krasopoulos G, Dusmet M, Ladas G, Goldstraw P. Nuss procedure improves the quality of life in young male adults with pectus excavatum deformity. Eur J Cardiothorac Surg 2006 Jan; 29(1):1–5. Epub 2005 Dec 5. PMID: 16337131 [PubMed – indexed for MEDLINE]
4. Lawson ML, Mellins RB, Tabangin M, Kelly RE Jr, Croitoru DP, Goretsky MJ, Nuss D. Impact of pectus excavatum on pulmonary function before and after repair with the Nuss procedure. J Pediatr Surg 2005; 40(1):174–80

Q 16

Nancy Rollins and Korgun Koral

A 2-day-old infant presented with respiratory distress.

- What does the chest radiograph show (Fig. 1)?
- What are the CT findings (Figs. 2, 3)?
- What is the differential diagnosis?
- What is the diagnosis?
- What are the types and prognostic implications?
- How is this condition treated?

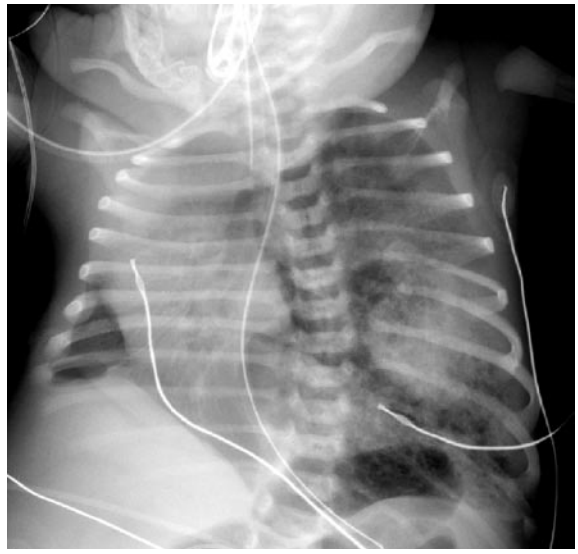

Fig. 1

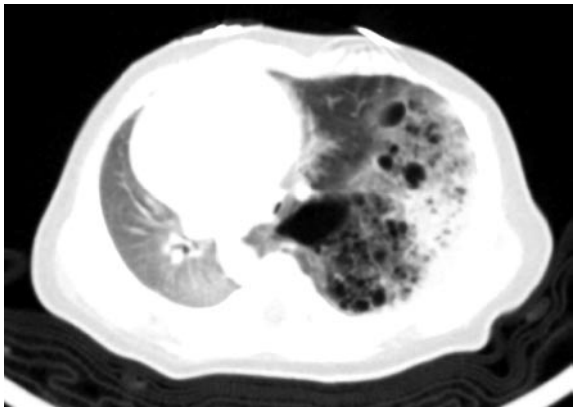

Fig. 2

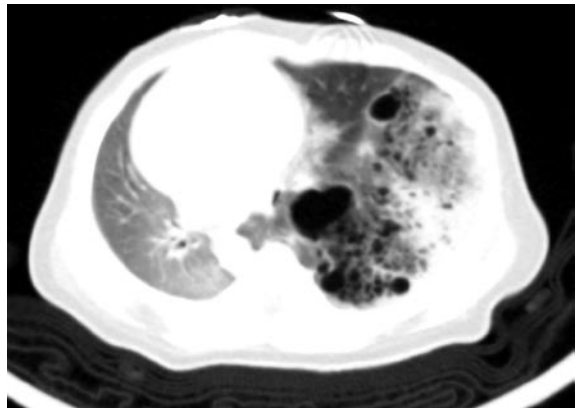

Fig. 3

A 16

There is a rightward shift of the mediastinal structures with expansion of the left hemithorax. A small air cyst can be appreciated in the left lung.

A shift of the mediastinum to the right is seen with a small right lung. There are multiple small cysts in the left lung with areas of solid tissue.

The differential diagnosis includes cystic congenital adenomatoid malformation (CCAM), pulmonary sequestration, congenital diaphragmatic hernia, and cavitary necrosis complicating pneumonia.

The diagnosis is CCAM, type 2. Type 1 consists of one or more large cysts. In type 2, numerous small cysts of uniform size are present. Type 3 appears solid on imaging, but has microcysts. Different types of CCAM do not have different clinical implications.

Treatment is surgical resection.

Suggested Reading

1. Donnelly LF. Pocket Radiologist: Pediatrics. Amirsys Ed. 2002.
2. Kuhn JP, Slovis TL, Haller JO. Caffey's Pediatric Diagnostic Imaging. Elsevier Ed. 2004.

Q 17

Nancy Rollins and Korgun Koral

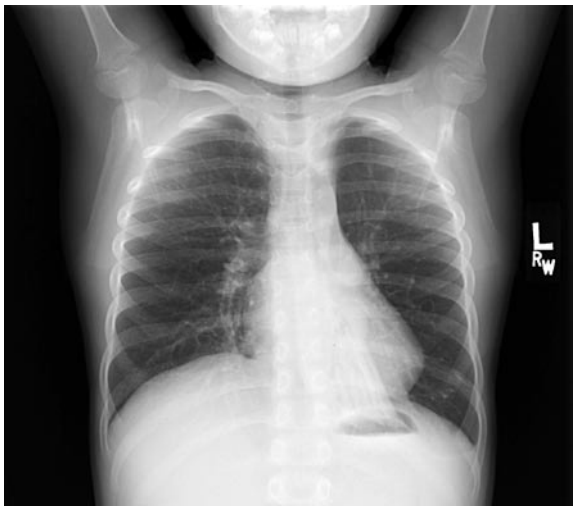

Fig. 1

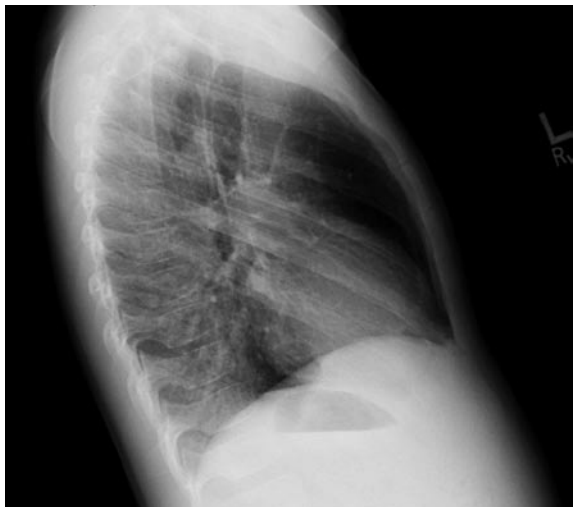

Fig. 2

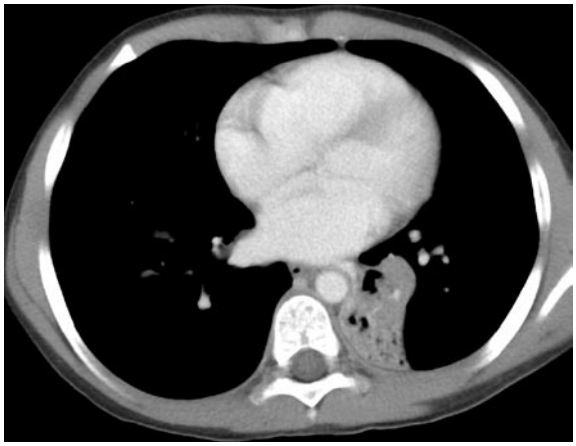

Fig. 3

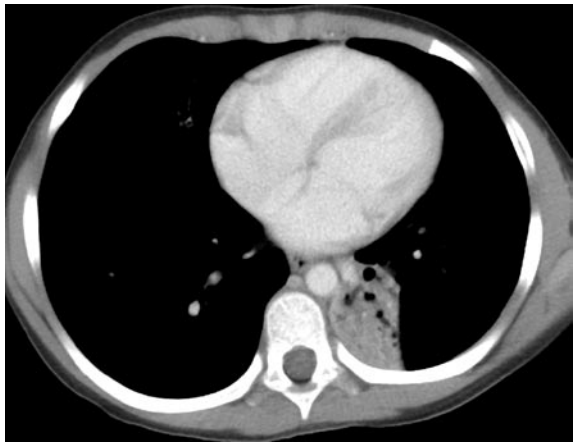

Fig. 4

A 6-year-old boy presented with multiple episodes of left lower lobe pneumonia.

- What are the findings on the posteroanterior and lateral chest radiographs (Figs. 1, 2)?
- What do the CT scans of the chest show (Figs. 3, 4)?
- What is the differential diagnosis?
- What are the types of this abnormality?
- How is it treated?