Q 1

Nancy Rollins

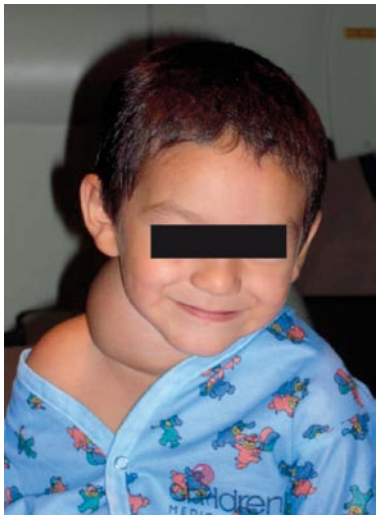

Fig. 1

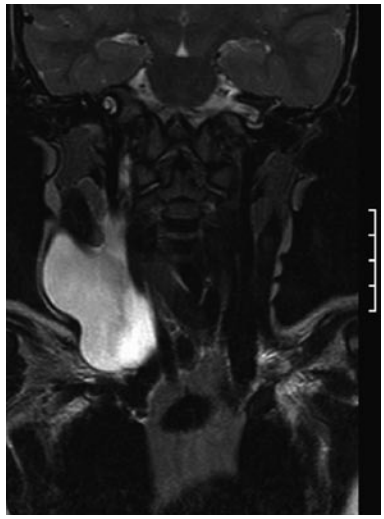

Fig. 2

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

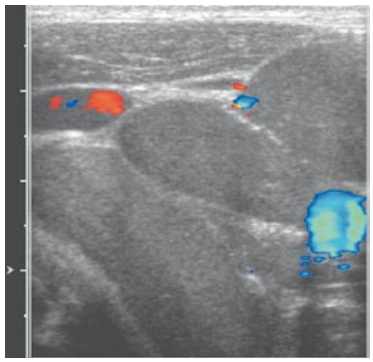

Fig. 3

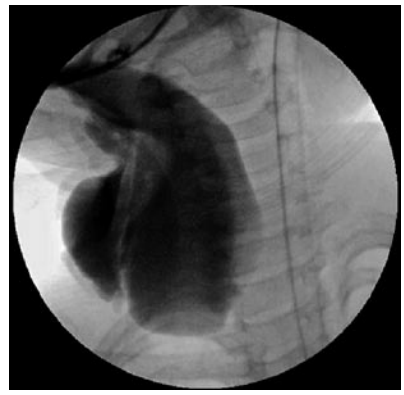

Fig. 4

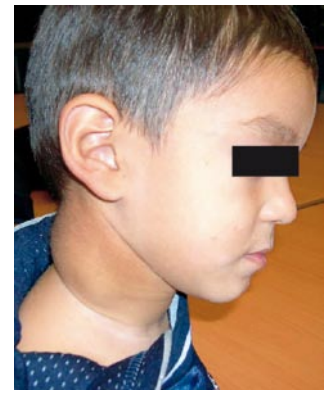

Fig. 5

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

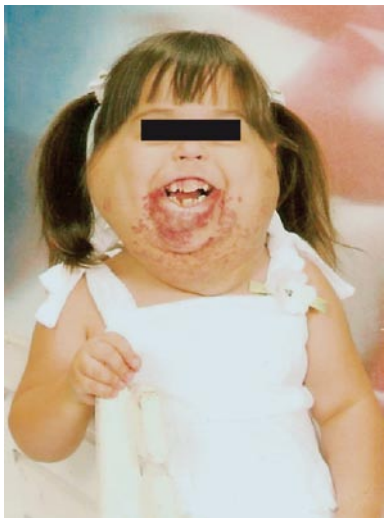

Fig. 1

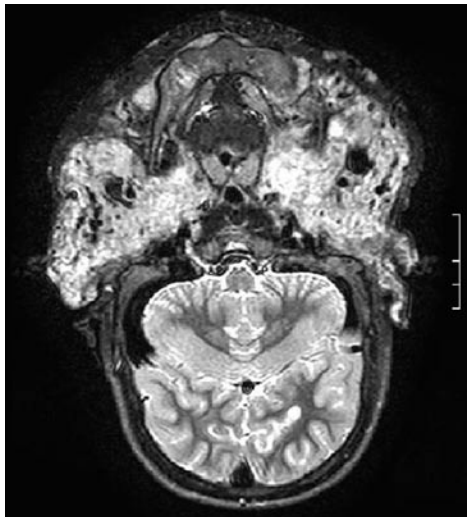

Fig. 2

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.

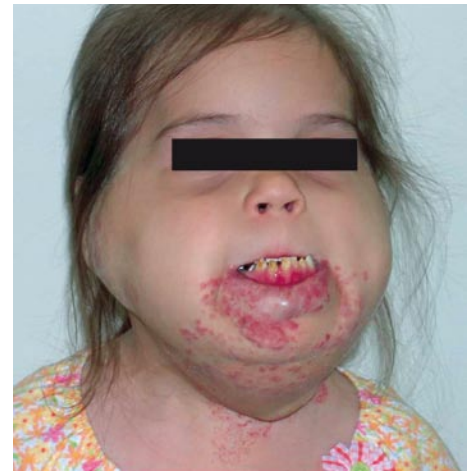

Fig. 4

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

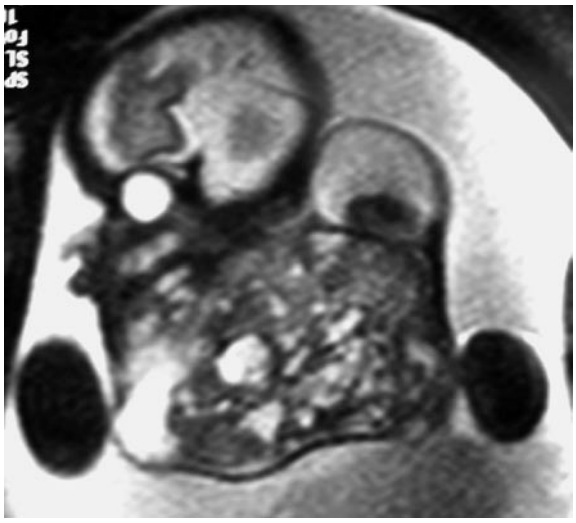

Fig. 1

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

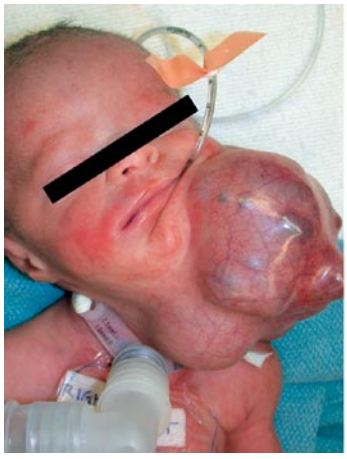

Fig. 2

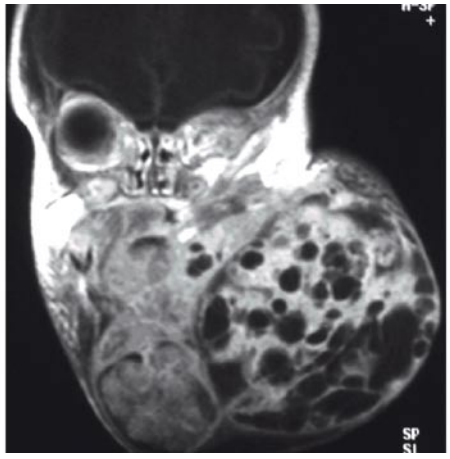

Fig. 3

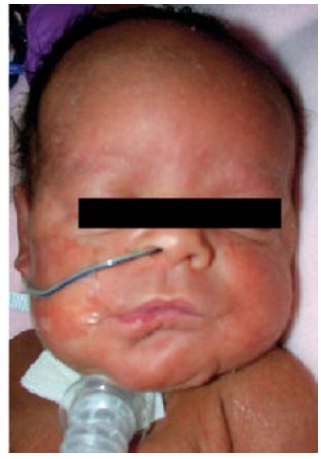

Fig. 4

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).

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