SECTION EDITOR: WALTER W. TUNNESSEN, JR, MD

Picture of the Month

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AN ANTENATAL ultrasound examination of a 39-year-old woman with a pregnancy of 21 weeks' gestation demonstrated a large, complex, multicystic cervical mass measuring 10×12 cm on the fetus. The neck mass was felt to be compressing the upper airway and, potentially, the esophagus. The prenatal course was complicated by maternal polyhydramnios. The infant was delivered at term via an EXIT (ex utero intrapartum treatment) procedure. At delivery, a huge mass was noted surrounding the infant's neck (**Figure 1**). The infant was immediately intubated.

The mass was excised on the third day of life (**Figure 2**).

From the Department of Pediatric Surgery, Children's Hospital of Pittsburgh, Pittsburgh, Pa.

Figure 1.

Figure 2.

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Denouement and Discussion

Cervical Hygroma and Ex Utero Intrapartum Treatment (EXIT)

Figure 1. A huge cystic hygroma distorts the entire anterior cervical area.

Figure 2. Appearance of the infant following neck mass resection and tracheostomy.

Cystic hygroma is one of the most common benign cervical anomalies, often presenting as a large, airway-obstructing lesion. The majority of these cysts diagnosed postnatally are detected in the neonatal period, with 80% present by age 2 years.[1] Although cystic hygromas may be associated with Turner syndrome, various chromosomal trisomies, and other anomalies, they mainly occur in children with normal chromosomal karyotypes.[2]

PATHOGENESIS

Cystic hygromas are congenital malformations of the lymphatic system; developmental lymphangiomas derived from the primitive embryonic jugular venolymphatic sacs.[1] The cysts occur most often in the neck; however, 10% are found in the axilla and mediastinum. The tumor has indiscrete margins and is composed of dilated cystic spaces lined by endothelial cells. The lack of encapsulation results in difficult surgical removal and incomplete resection, which in turn results in frequent recurrences. Cystic hygromas typically grow gradually, with infrequent spontaneous regression.

COMPLICATIONS

The most common and life-threatening complication of cystic hygroma is acute airway obstruction. Hemorrhage into or infection of the cystic hygroma may lead to rapid increase in size, with subsequent airway and esophageal compression. The mass is often too large to allow vaginal delivery without encountering airway complications and consequent anoxic brain injury to the infant. Prenatal diagnosis affords the opportunity to plan a surgical approach and to evaluate and secure an airway.

The initial manifestations of cystic hygroma are intrauterine and include polyhydramnios, fetal hydrops, hyperechogenic lung fields, and inverted diaphragms.[3] The polyhydramnios is secondary to compression of the upper esophagus. Alveoli distal to the upper airway obstruction become dilated.

EX UTERO INTRAPARTUM TREATMENT

Ex utero intrapartum treatment (EXIT) is defined as a partial delivery of the fetus through a lower transverse hysterotomy by ultrasound guidance, performed while maintaining maternal-fetal blood flow. This surgical technique was first described in the delivery of fetuses with congenital diaphragmatic hernias. The technique was necessary to provide continuous oxygenation to the fetus while tracheal clips that were placed in utero to stimulate prenatal lung growth were removed.[4] Since this original description, EXIT use has been extended as a method of securing an airway in fetuses with airway obstruction while maintaining oxygenation via uteroplacental gas exchange.[5]

TREATMENT AND PROGNOSIS

The treatment of choice of a large cervical cystic hygroma is excision. Care must be taken to carefully indentify vital structures adjacent to the cyst, including cranial nerves, major vascular structures, or soft tissues such as the hypopharynx, parotid gland, or trachea.[6] Postoperative complications, including recurrence, nerve damage, and infection, occur in 30% or more of cases.[6] Multiple operations may be necessary to remove complex lesions, particularly those with extension into the mediastinum. Recurrence rates depend on the complexity of the lesion and degree of excision. Incompletely excised lesions may recur in 50% to 100% of cases.[6]

EXIT is a promising technique for infants with prenatally diagnosed airway-obstructing lesions, including large cervical hygromas.

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Submissions

The Editor is seeking submissions for a new feature, *Clinical Problem Solving*, which will combine *Picture of the Month*, *Radiological Case of the Month*, and *Pathological Case of the Month*. Our aim is to demonstrate the thinking process of a master clinician involved in approaching a patient with an unknown disease. The discussion of such cases should place the clinician's expertise into the context of the prevailing medical literature on the topic. Manuscripts should be between 3000 and 4000 words and may include photographs and radiographs.

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