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CASE REPORT

Huge Neck Masses Causing Respiratory Distress in Neonates: Two Cases of Congenital Cervical Teratoma

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Congenital cervical teratomas are rare and usually large enough to cause respiratory distress in the neonatal period. We present two cases of congenital huge cystic neck masses in which distinguishing cervical cystic hygroma and congenital cystic teratoma was not possible through radiologic imaging techniques. Experience with the first case, which was initially diagnosed and treated as cystic hygroma by injection sclerotherapy, led to early suspicion and surgery in the second case. The masses were excised completely and histopathologic diagnoses were congenital teratoma in both patients. Our aim is to review congenital huge neck masses causing respiratory distress in early neonatal life to highlight this dilemma briefly with these interesting cases.

1. Introduction

Fetal neck masses are rare and may be encountered during fetal anomaly screening scan during the second trimester. It is essential to distinguish the different pathologies as they influence prenatal counseling, antenatal, and postnatal management. In the neonatal period, differentiating the various neck masses and reaching an accurate diagnosis are important for the prognosis and appropriate timing of surgical treatment.[1] The anatomic location of the mass can yield a clue about probable origin of the mass; anterior neck masses include teratoma, epignathus, goiter, bronchogenic cyst, and hemangioma; and posterior masses include cystic hygroma (cystic lymphangiomas located in the head and neck), cervical meningocele, occipital encephalocele, and hemangioma.[1] Lymphangiomas and teratomas may be seen as giant fetal neck masses.[2] Complete and meticulous surgical excision is the recommendation for each of them; however, several medical treatments have been preferred in the treatment of lymphangioma to avoid surgical complications.[3] Herein, we report two cases of congenital cervical

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teratoma, one of which was initially treated as lymphangioma according to radiological diagnosis.

2. Case Reports

2.1. Case 1

A 1-day-old newborn was referred to our unit for a huge neck mass. The mass was first detected at 12 weeks' gestation as a small cyst, but the subsequent ultrasonography (US) scan was normal. Physical examination revealed a huge neck mass (Figure 1). The chest radiography, abdominal US, and echocardiography were normal. The tumor involving both sides of the neck extended from the auricle to the clavicle. US revealed a 13 cm $\times$ 9.5 cm $\times$ 7 cm multiloculated, multiseptate mass. Blood flow in the tumor was fairly unremarkable, and pulsating flows were not detected. Computed tomography (CT) revealed an 8 cm $\times$ 13 cm $\times$ 7 cm multiloculated cystic mass, which extended from the left maxilla to bilateral submandibular area, and to the anterior chest surrounding the trachea at both sides with scattered microcalcifications (Figure 2A).

Based on the US findings, we considered the tumor to be lymphangioma. We planned injection of OK-432 (Picibanil, Chugaiv Pharmaceutical Co., Tokyo, Japan). During the 3rd postnatal week, the baby developed respiratory distress and was intubated. On postnatal Day 27, OK-432 was injected into the largest cysts under ultrasonographic guidance in the interventional radiology unit. After fluid aspiration from the lesion, the same amount of OK-432 was introduced up to a maximum of 20 mL with a concentration of 0.01 mg/dL. Cervical magnetic resonance imaging (MRI) obtained 10 days after repeated sclerosing therapy revealed multiloculated cystic lesions with septae of varying thickness and no reduction in the size of the mass (Figures 2B and C). Surgical intervention was undertaken through a left-sided incision over the tumor. In the midline, the mass tightly adhered to the tracheal cartilages. The tracheal cartilage was injured during excision in vertical plane in a 1.5 cm length and primarily repaired. The huge mass was totally excised. Respiratory distress required tracheotomy on postoperative Day 29, and feeding was maintained via a gastrostomy tube. The patient did not experience any recurrent mass during 1-year follow-up. The postoperative MRI showed normal anatomic structures without recurrent disease (Figure 2D).

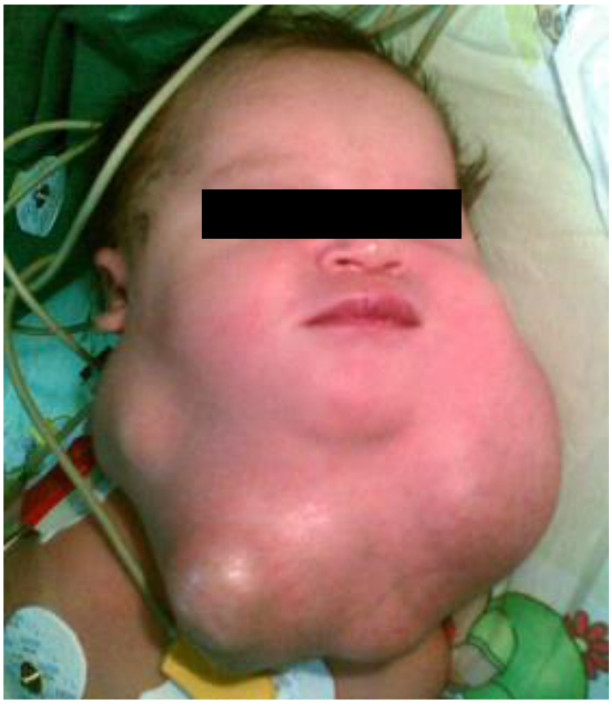

Figure 1 Huge lobulated irregular neck mass.

Microscopic examination of the specimen revealed skin adnexal structures, mature glial tissue, cartilage, smooth muscle, fibrous and fatty tissue, minor salivary glands, respiratory and gastrointestinal structures, and thyroid tissue. These findings were consistent with mature teratoma that developed from all three germ layers (Figure 3A).

2.2. Case 2

A 2-day-old male child was referred to our hospital for a large cervical mass. The child developed respiratory distress soon after birth and, therefore, he was intubated. On the first antenatal US, which was performed at 12 weeks' gestation, no abnormality was detected. However, polihydroamnios was detected on US in the 3rd trimester.

The tumor was located predominantly on the left side of the neck from the level of mandible to the supraclavicular space extended to the right side. The α-fetoprotein (AFP) level was 226.689 ng/mL. The chest radiography, abdominal US, and echocardiography did not reveal any malformations except the neck mass. MRI confirmed a 5 cm $\times$ 3.5 cm multiloculated cystic mass, which was hyperintense on the T2-weighted images, and demonstrated peripheral wall enhancement on the T1-weighted image, extending from the left hypopharynx—parapharyngeal space to the caudal anterior midline, submandibular region, and partly to the right sternocleidomastoid muscle (Figure 4). On the 2nd day, surgical intervention was undertaken through a left-sided incision over the tumor. A multiloculated cystic mass, which had a definite capsule, was totally resected with preservation of the significant locoregional structures such as thyroid and parathyroid glands. The AFP level dropped to 63.125 ng/mL on postoperative Day 5. He was extubated on postoperative Day 7.

The mass was composed predominantly of immature neuroepithelial tissue forming rosette-like structures, tubules and immature cartilage (Figure 3B), bone, smooth muscle, and fatty tissue in histopathological examination. The diagnosis was immature teratoma.

The patient was discharged on Day 45. During the 5-month follow-up, the patient remained well, developing normally with no evidence of recurrence.

3. Discussion

Routine antenatal US in populations reduced the unexpected cervical masses presenting soon after delivery. Antenatal diagnosis may also be made following targeted US evaluation

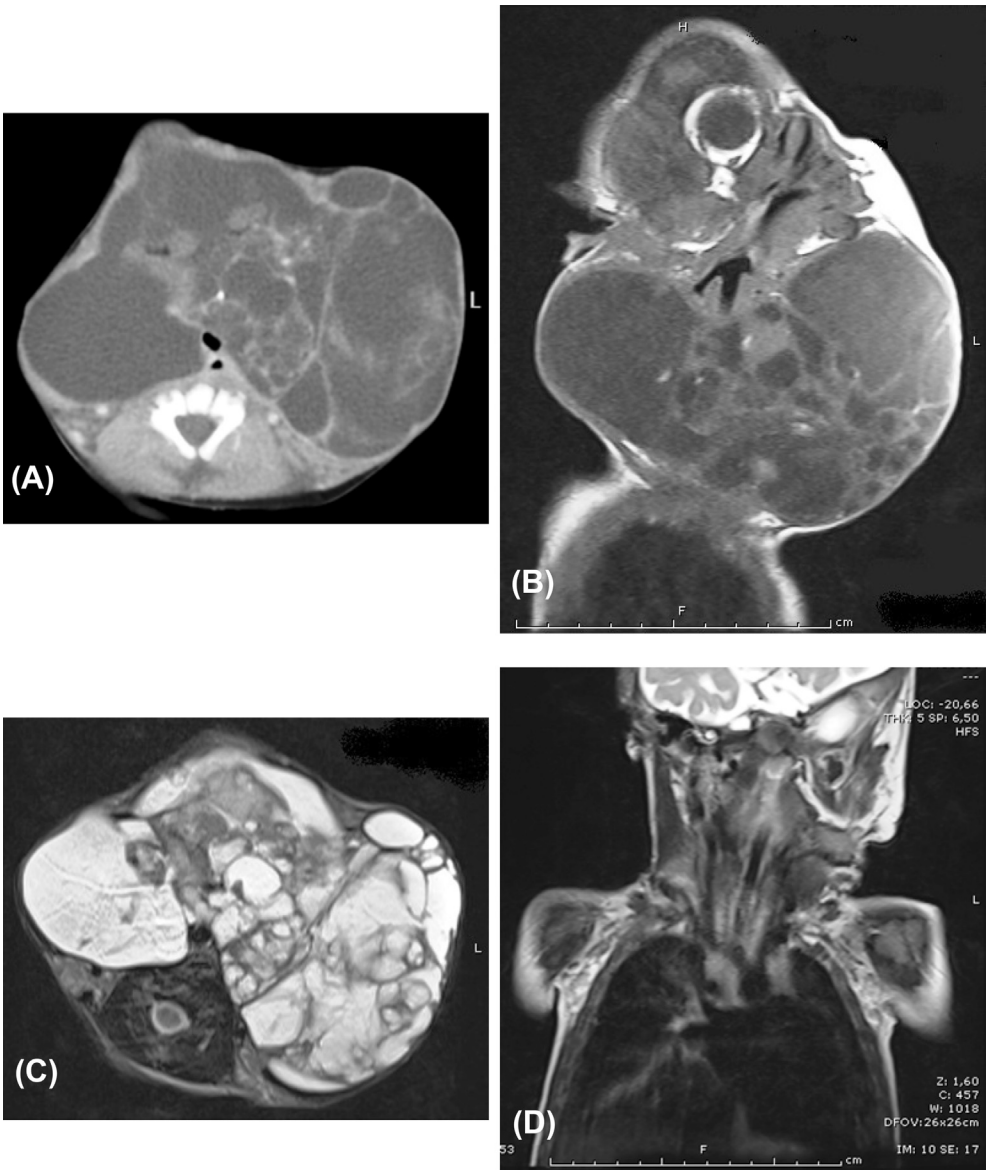

Figure 2 (A) Axial computed tomography revealed an 8 cm × 13 cm × 7 cm diameter cystic mass that extended from the subcutaneous fat tissue on the left maxilla to bilateral submandibular area and to the anterior chest wall through the anterior part of the trachea in the midline surrounding the trachea at both sides with scattered microcalcification and no solid components; submandibular glands and thyroid gland were not visualized. (B) Coronal T1-weighted magnetic resonance imaging (MRI) revealed multiloculated a cystic lesion settled in the anterior part of the neck, extending to both sides, but with a large component observed in the left side with septa of varying thicknesses. (C) Axial T2-weighted MRI revealed high intensity on T2-weighted images with no reduction in the size of the mass and also noted an absence of fat. (D) Postoperative MRI: Coronal T1-weighted images revealed no recurrent mass.

for polyhydramnios, which is presented in 20% of cases.[4] A cystic lesion was observed in the early weeks of gestation in Case 1, and polyhydramnios was detected in the third trimester in Case 2. However, antenatal diagnosis of cervical mass was not made in presented patients.

When faced with a newborn with a big mass in the neck region, it is important to make the diagnosis as soon as possible. The differential diagnoses of a congenital neck mass include cystic hygroma, hemangioma, lipoma, dermoid cyst, congenital goiter, cervical meningocele, occipital encephalocele, bronchogenic cyst, branchial cleft cyst, thyroglossal duct cyst, and teratoma.[5] The clinical differentiation between cystic hygroma and cervical teratoma can be very difficult because of the similarity in the patient's age and sex, the location of the lesion, and clinical presentation. The large size of the tumors often obscures the origin of the lesion.[1]

Head and neck teratomas account for approximately 3—5% of all neonatal teratomas, occurring with an incidence of 1 in 20,000—40,000 live births. They usually have their origins from anterolateral structures with the extension to the midline,[4] as in our cases. Presentation during live birth can vary, depending on the mass effects of the lesion, ranging from an asymptomatic neck mass to an

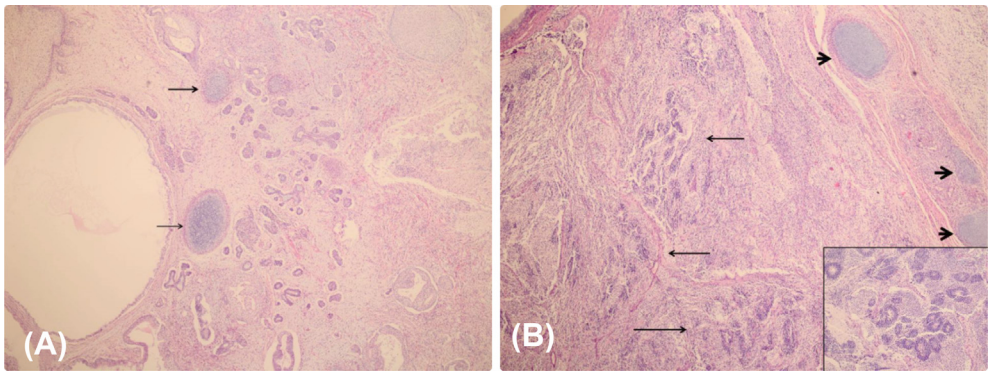

Figure 3 (A) Mature teratoma composed of fetal cartilage (arrows), respiratory epithelium, and minor salivary glands (hematoxylin and eosin ×40). (B) The neoplasm was largely composed of immature neuroepithelial elements (long-thin arrows), and immature cartlilage (short-thick arrows; hematoxylin and eosin ×40). Inset: Neoplastic immature neuroepithelial elements: neuroepithelial rosettes and tubules (hematoxylin and eosin ×100).

obstructed labor, or life threatening airway compromise.[4] In Case 1, respiratory compromise appeared 2 weeks after delivery, and in Case 2, airway security through entubation was required soon after delivery. Patients with cervical teratoma as an isolated anomaly can do well after postnatal resection if the airway is secured prior to surgery. Patients with pulmonary hypoplasia, in addition to the huge cervical mass, are unable to be oxygenated, and they die of respiratory insufficiency despite an adequate airway being maintained.[5] It is reported that pulmonary hypoplasia was not seen in patients with giant cervical lymphangioma. This finding was attributed to the teratoma being more solid and less compressible than cervical lymphangioma.[6] In the presented patients the airway obstruction was present, but there was no lung hypoplasia.

Upon US examination, cervical lymphangiomas appear as fluid-filled cystic spaces, divided by fine septae, and are usually located in the anterior and posterior triangles of the neck.[6] In contrast to lymphangiomas, cervical teratomas are usually asymmetrical and unilateral, with well-defined margins. Most are multiloculated, irregular masses with solid and cystic components. The presence of solid components is useful in distinguishing a cervical teratoma from lymphangioma, and as high as 50% of cervical teratomas will have calcifications. When calcifications are present in combination with a partially cystic and solid neck mass, it is pathognomonic of a cervical teratoma.[7] When the diagnosis is in question, a high-resolution MRI or CT scan has been shown to aid in clarifying the diagnosis.[8] On CT scans, the tumor limits, proximity to the airway, and great vessels can

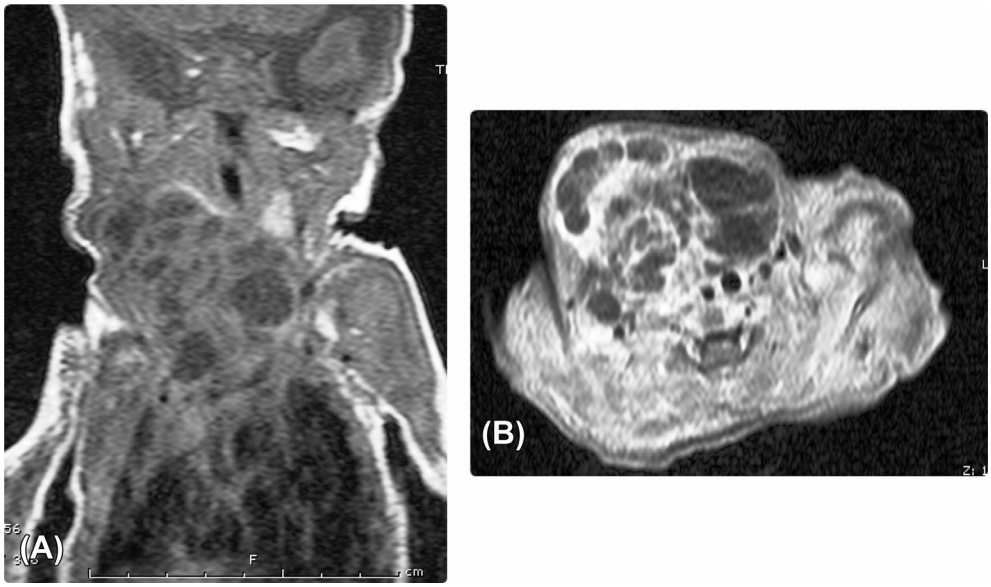

Figure 4 (A) Coronal T1-weighted magnetic resonance imaging revealed 5 cm × 3.5 cm-diameter multiloculated cyst which is hypointense on the T1-weighted image extending from the right hypopharynx—parapharyngeal space to the caudal anterior midline, submandibular region, and partly to the right sternocleidomastoid muscle. The mass was lobulated and composed of predominantly multilocular cystic components. (B) On axial T2-weighted magnetic resonance imaging, the cysts appear hyperintense.

be seen, and calcification within the tumor is clearly defined. An MRI provides similar information, with the benefit of multiplanar imaging without radiation exposure. On an MRI, teratomas are hypo- or isointense on T1-weighted images, and hyperintense on T2-weighted images.[9] Frequently, teratomas show unilocular or multilocular cystic components, and if truly cystic, it can be impossible to distinguish from cystic hygroma via CT or MRI.[4]

Distinguishing hygroma from teratoma by AFP level has a number of pitfalls. Fetal development, gestational age, birth weight, and age since birth all contribute to the normally elevated *postpartum* AFP levels, regardless of the presence or absence of teratoma. A further pitfall is that mature teratomas may not produce high quantities of AFP, and may, therefore, recur or undergo malignant transformation without significant AFP production.[10] In our first case, due to radiological signs, we did not consider teratoma; therefore, an initial AFP level was not obtained. In the second case, the AFP level was high preoperatively and dropped to one-fourth on the 5th postoperative day.

Congenital cervical teratomas must be differentiated from cystic hygromas. Both occur in patients of the same age range, may be very large, and have a similar location and extension, and the same clinical presentation. They are both composed of primarily cystic elements. Although calcifications on imaging work-up may facilitate diagnosis of congenital cervical teratoma, the absence of calcifications do not exclude the diagnosis of cervical teratoma.[11]

The prompt and complete surgical resection of congenital cervical teratomas is the appropriate treatment.[12] In the presented cases, previous experience with Case 1, rather than radiological imaging, facilitated the early operation of Case 2. The histopathologic diagnosis was mature teratoma in Case 1, and immature teratoma in Case 2. In Case 2, early operation facilitated early weaning from the ventilator support and resulted in a better outcome and a better quality of life.

Clinicians should be cautious when faced with a fetus or neonate with a cystic mass in the head and neck. Teratoma, even though extremely rare, should be considered as part of the differential diagnosis and confirmed or excluded prior to planning definitive treatment.

Conflicts of interest

All authors declare no conflicts of interest.

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