Abstract:

Congenital cervical teratomas are rare tumors. Although essentially benign, obstruction of the airway is the major challenge in the neonatal period. Prognosis is good provided that the airway is quickly stabilized and resection is not delayed. Prenatal diagnosis allows planning of resuscitative efforts in advance. We report a case of immature cervical teratoma in a near-term male infant born to a 32 year-old woman. The tumor was removed surgically with no recurrence after three and a half years.

Introduction:

Cervical teratomas are quite rare tumors. They are usually reported as isolated cases or as reviews in the surgical or pathology literature. Maternal ultrasound can detect various fetal anomalies that may be amenable to prenatal or neonatal intervention. Cervical teratoma is one of these anomalies that has the potential for upper airway obstruction and severe respiratory distress secondary to tracheal compression and is usually diagnosed in newborn or stillborn infants presenting with upper airway obstruction. Early diagnosis and planned management are of paramount importance for the survival of these infants. We describe a case of cervical teratoma diagnosed antenatally as a huge neck swelling with upper airway obstruction.

Case report:

A 32 year-old, gravida 8, para 4, pregnant lady developed severe polyhydramnios at midterm. Ultrasound examination performed at 24 weeks of gestation demonstrated a huge neck mass, diagnosed provisionally as a cystic hygroma. An elective caesarian section was planned for week 36 and a healthy male child was delivered. Shortly afterwards he developed a life-threatening airway obstruction. The vocal cords were not easily visualized and the larynx was twisted markedly in an oblique direction. A difficult endotracheal intubation was carried out successfully and the baby was transferred to the Neonatal Intensive Care Unit (NICU) for mechanical ventilation.

The mass, measuring approximately 13x11x5 cm., covered most of the anterior neck and lower facial areas. It had a heterogeneous consistency with well-defined edges. Ultrasound (US) examination confirmed the finding of a mixed cystic and solid lesion. Figures 1 & 2 show the patient preoperatively.

Plain X-ray showed calcification and a CT Scan showed significant tracheal compression (Figure 3). The clinical impression, as well as the radiological findings, excluded the possibility of cystic hygroma and a differential diagnosis of congenital goiter or tumor was considered. Thyroid function tests were all within normal limits. A Tc.99 scan at day two was inconclusive in detecting thyroid gland elements amongst the tumor tissues. A diagnosis of cervical teratoma was then made and the patient was booked for surgical exploration at day five. Preoperative assessment of alpha feto-protein (AFP) was normal.

Upon exploration the mass appeared well encapsulated. There were no definite thyroid gland that could be detected and separated from the tumor and so total excision of the mass was carried out. Grossly the mass weighed 123 grams and measured approximately 13x11x5 cm., covered most of the anterior neck and lower facial areas. It had a heterogeneous consistency with well-defined edges. Ultrasound (US) examination confirmed the finding of a mixed cystic and solid lesion. Figures 1 & 2 show the patient preoperatively.

Figure 1: The patient preoperatively.

Figure 2: Patient preoperatively, “close up view”, showing the large mass and the irregular nodular surface.

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Neonatal Cervical Teratoma


Figure 3: CT scans of the patient preoperatively showing the tumor extension, tracheal compression, heterogeneous consistency and classification.

9 x 6 x 4 cm. The outer surface was intact and bosselated, the cut surface was partially solid and cystic with variable size cysts. Histopathological examination revealed an immature teratoma with thyroid tissue stretched between the capsule and the tumor tissue. The teratoma was composed predominately of mature tissues representing the three germ cell layers, including connective tissue bands, cartilage, adipose tissue, skeletal and smooth muscle, choroid plexus, differentiated neuroglia ganglion cells and various epithelial-lined cysts and skin. These tissues were haphazardly mixed with immature fetal tissues exhibiting neuroblastic differentiation and rosette formation, comprising approximately 15-20% of the solid tissue. No areas of malignant germ cell tumor or carcinomatous differentiation could be seen (Figures 4-7).

Figure 4: Cut surface of the mass showing partially solid and cystic areas with variable sized cysts. Thyroid tissues are seen stretched out under the capsule.

Figure 5: Residual thyroid tissue stretched at the periphery of the tumor.

Figure 6: High power section showing variable sizes epithelia-lined cysts.

Figure 7: Immature neuro-epithelial component forming sheets of neuroblasts and immature neural structures forming rosettes.

The patient developed hypothyroidism postoperatively and was maintained on L-thyroxin. A follow up CT scan revealed no residual tumor tissues and since the mass was an immature teratoma the progress was monitored strictly by the pediatric surgeon, oncologist and endocrinologist. Serial examination and serum samples of AFP were assessed every three months in the first year after surgery, then every six months for the rest of the follow up period. Apart from his endocrinological defect, the patient is currently thriving and leading a normal life for his age and stage of development (Figures 8 & 9).

Figure 8: Postoperative follow up after 6 months.

Figure 9: Postoperative follow up at the age of 3½ years.
Neonatal Cervical Teratoma


Discussion:

Teratomas are rare congenital neoplasms occurring with an incidence of about 1 in 40,000 live births(6). In most series of teratomas, cervical lesions account for only 1.4 to 5.5% of all the tumor sites encountered(7-9). Diagnosis using ultrasound was first reported in 1978(10) and then later in 1982(11). Today the prenatal ultrasound examination is considered an essential and reliable tool for detecting cervico-facial teratomas in utero(6,9,12-14).

A high proportion of these tumors (approximately 33%) is associated with maternal polyhydramnios, especially tumors larger than 10cm in diameter. In such cases the neonate can be delivered electively taking special precautions and with experienced personnel attending(5). Calcifications and tracheal deviation are seen in a plain X-ray in 50% of cases. A mixed solid and cystic lesion in ultrasound is highly suggestive of teratoma and this differentiates it from cystic hygroma and congenital goiter. Whenever such a lesion is suspected, thyroid function studies are required pre- and post-operatively in addition to AFP monitoring.

Cervical teratomas are uniformly benign in infancy but, as in the case of sacrococcygeal teratomas, they have a potential for malignancy and occasional malignant transformation and consequent death have been reported(15,16). On the other hand, disease-free survival following tumor excision has been reported on a number of occasions(17-19). Early surgical intervention is important to relieve the obstruction of the airway and to safeguard against later malignant transformation. The standard technique is similar to that of thyroid lobectomy as the lesions are usually unilateral and encapsulated(18). In our patient, however, the teratoma arose from within the thyroid gland and was intimately associated with the thyroid tissues in such a way that identification of the thyroid gland tissue was impossible intraoperatively. To ensure complete extirpation of the tumor mass, total thyroidectomy had to be carried out as well. Post-operatively the patient required thyroid replacement therapy and prolonged follow-up.

In the literature cervical teratoma has been long divided into thyroid teratoma and cervical teratoma depending upon the presence of thyroid tissue as a tumor element. This classification was found to be impractical by many authors, as both types behave clinically in the same way. Consequently this classification has been largely abandoned in favor of the more general term "cervical teratoma"(18).

The most important variable affecting long-term survival has been the successful and complete resection of the tumor allied to the histopathological pattern of the teratoma where the immature type has a higher potential for malignancy(6). An immature teratoma variant was detected in our patient who has been followed meticulously for three and a half years. Apart from his endocrinical deficit, he is doing well and both the family and the caring physicians are satisfied with this long-term result.

In conclusion, congenital cervical teratomas, though rare, carry a potentially lethal prognosis not only because of their possible future malignancy but also because of the immediate effects of airway obstruction. Prenatal diagnosis and planned postnatal management can significantly alter the outcome of such a lesion.

References: