A Huge Immature Cervical Teratoma; Antenatal Diagnosis, and its Management – An Unusual Entity

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ABSTRACT

Cervical teratomas are not reported quite frequently in medical literature, especially from Indian subcontinent. Prenatal diagnosis of this condition is even rarer, making the management protocols difficult. In absence of prompt intervention in the form of airway decompression, morbidity and mortality is very high. Presented here is the case of antenatal diagnosed cervical mass, which was managed successfully in immediate post-natal period with uneventful survival and normalized tumor marker levels.

Key words:
Airway compression, alpha fetoproteins, neck mass, teratoma

INTRODUCTION

Teratomas are tumors derived from all the three germ cell layers namely ectoderm, mesoderm, and endoderm and, therefore, contain the tissues of all the germ cell layers. Location-wise, these tumors are ubiquitous in the body, though the relative frequency varies greatly.[1] Most commonly found in the sacrococcygeal region, these tumors are extremely rare in neck region. The sign and symptoms are mostly attributed to the mass effect of these lesions, making the cervical tumors potentially lethal. The computed tomographic (CT) scan is superior to MRI in this case. In principle, the MRI is the preferred imaging method without radiation but when we need to do the staging of an unknown tumor, CT gives more appropriate information about the vascular involvement. Nowadays, management includes maintenance of the maternal-fetal circulation, allowing continuous fetal oxygenation, and surgery is the preferred treatment. Long-term follow-up is done by estimation of relative levels of pre- and post-operative levels of tumors markers.

CASE REPORT

A 26-years-old second gravida female reported at 37th week with antenatal ultrasound of the fetus showing well-defined and hyperechoic solid mass of size of 5.5 × 6 centimeters in the anterior aspect of neck [Figure 1]. Associated findings included mild polyhydromnios and vertical lie. Guarded prognosis with conservative management till the birth of the fetus was advised. Mode of the delivery was to be decided by obstetric indications. Finally, the baby was born by cesarean section and had huge cervical mass. Baby cried immediately after birth and had respiratory distress of mild nature apparent only in supine position when the compression effect was maximum. The distress was relieved in lateral position, and baby was maintaining saturation with minimal oxygen support.

On initial assessment, the vitals of the baby were stable. Examination of respiratory system revealed equal air entry on both the side with mild conducted sounds and inspiratory stridor. Rests of the systemic examinations were within normal limits. On local examination, there was a neck mass which was solitary, oval, of size about 9 × 5 × 6 cms in midline anteriorly having firm consistency with moderate mobility in all the directions [Figure 2]. Swelling was non-pulsatile, and there were no skin changes.

Initial imaging was done by X-ray neck and ultrasound. These were stipulated calcifications within the mass as shown by X-ray. Ultrasound showed the mass to be consisting of solid matrix with occasional cystic areas filled with hemorrhagic fluid having freckles of calcifications and well-defined capsules all around with neck vessels pushed laterally and some degree of tracheal displacement

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suggestive of the teratoma with mass effect. Computed tomographic scan was obtained to further rule out the organ of origin, vascular involvement, and thoracic extension of the mass lesion. CT showed the mass to be of size 8 × 6 × 5 cms and heterogeneous with scattered, nodular calcification extending superiorly into sub-mandibular and inferiorly up to supraclavicular region [Figure 3]. Invasive investigations like fine needle biopsy were not attempted due to the fear of hemorrhage within the tumor leading to further aggravation of respiratory distress and complicating the pre-operative clinical scenario. Corroborative evidences of the mass to be teratoma were obtained by means of tumor marker studies like estimation of alpha-fetoprotein and beta human chorionic gonadotropins. Alpha-fetoprotein level was significantly high in the range of 83,000 ng/ml though there was no rise in beta HCG. As the neck teratomas mostly arise from and completely replace the thyroid tissue, thyroid function test was also obtained, but T_3, T_4, and thyroid-stimulating hormone levels were within normal range.

After pre-operative stabilization and investigations, patient was taken up for the surgical excision of the mass. General anesthesia with endotracheal intubation was done though securing the airway was difficult. Transverse incision directly over the mass was given. Below the platysma muscles, the well-encapsulated mass was lying above the strap muscles of the neck, attached only with the fibrous bands and few small caliber vessels feeding the mass. Complete enucleation of the mass without capsule rupture was attained, and specimen was sent for histopathological examinations. No ventilatory support was needed in post-operative period. Feeds were allowed in 2nd post-operative day, and patient was discharged on 4th day. Repeat alpha-fetoprotein and beta HCG at the end of 1 month, 3 months, and 1 year were within normal range.

Histopathology report came out as lobulated grey-white mass predominantly solid with areas of cystic changes. Cysts were ranging from 1 mm to 1 cm and filled with mucinous material. The solid parts consisted of cartilages and bony spicules. Multiple sections studied from tumor showed mature as well as immature elements derived from all 3 germ layers. Mature elements comprised of nests of squamous cells, glands, mature cartilage, occasional bony tissue, neural tissue, and smooth muscle tissue. Immature elements included neuroepithelial elements, occasional group of blastemal cells, and immature cartilage in myxoid stroma. Mitosis was in the range of 2/10 HPF. Normal thyroid tissue was not seen in the section studied. Final diagnosis was immature cervical teratoma; grade II.

DISCUSSION

The word teratoma is derived from Greek, which means monstrous. Large neck swellings in neonatal period are uncommon. Though teratoma are known to occur anywhere in the body and commonest location being sacral region,

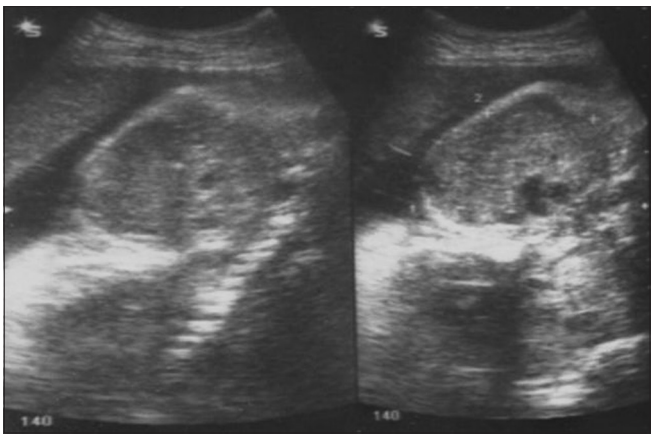

Figure 1: Ultrasound of the fetus revealed a solid mass in the anterior aspect of the neck

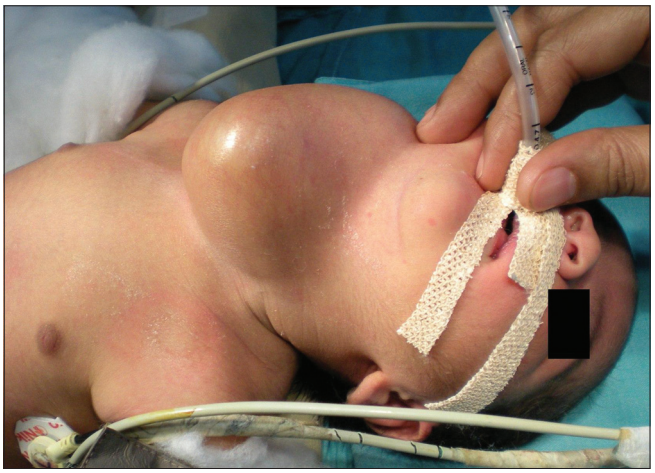

Figure 2: A giant solitary mass seen in the neck

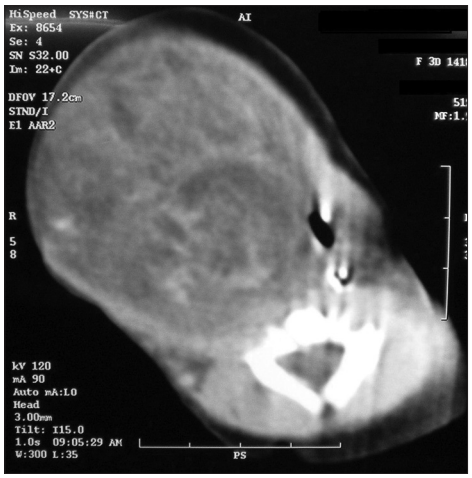

Figure 3: Computed tomographic showed a heterogeneous mass with nodular calcification extending superiorly into submandibular and inferiorly up to supraclavicular region

cervical presentation constitutes only small percentage of 1.6% to 9.3% of all pediatric teratomas, roughly equating to 1 per 40,000 births.[1] Globally, over 150 cases of teratoma in pediatric population have been reported. We could find only 3 case reports from India, and only 1 of them is surviving.[2]

Exact cause of teratoma remains obscure. No relation to the maternal age or specific ethnicity could be established. Speculations regarding probable genetic changes on comparative genomic hybridizations are being sought and they include 1p21.1 amplification, 9p22 deletion, and 17q21.33 1-copy gain.[3] Associated malformations include imperforate anus, Chondrodystrophia fetalis, hypoplastic left ventricle with pulmonary hypoplasia, cystic fibrosis, absence of corpus callosum, and arachanoid cyst, though very rare.

Degree of malignancy depends upon the grade of maturity of the components tissues and the majority the reported neonatal cervical teratomas belong to mature category though immature and highly malignant teratomas have also been reported. Teratomas commonly are classified using the Gonzalez-Crussi grading system: 0 or mature (benign); 1 or immature, probably benign; 2 or immature, possibly malignant (cancerous); and 3 or frankly malignant. If frankly malignant, the tumor is a cancer, for which additional cancer staging applies.[4] The presented case was found to be immature teratoma grade II due to fewer mature epithelial foci and comparable neuroepithelium with common mitotic figures not more than 40 per high power fields.

Such neck masses, which are formed at early gestation period and acquire gigantic dimensions in late gestation period, present with peculiar set of sign and symptoms. The compression over the esophagus causes mild to moderate hydramnios. Associated compression over the trachea also causes tracheomalacia increasing the post-operative morbidity.

The EXIT procedure, or ex utero intrapartum treatment procedure, is a specialized surgical delivery procedure used to deliver babies who have airway compression. Originally developed to reverse temporary tracheal occlusion in patients who had undergone fetal surgery for severe congenital diaphragmatic hernia, EXIT procedures have been shown to be useful for management of other causes of fetal airway obstruction, like giant cervical teratomas.[5]

Once stabilized at birth, investigations before any definitive procedure include baseline hemogram and blood biochemistry, USG, CT scan/MRI, FNAC and biopsy, thyroid and parathyroid function test, serum alpha-fetoprotein and beta HCG, transcription factors GATA-4 and GATA-6, and genetic studies.[6] Even the fine needle biopsy can be done in the presenting case, as the probability of bleeding is almost zero, and the fear to aggravate the respiratory problems is not evidence-based. Cyto/histologic result prior to the operation procedure would be appreciated to know whether the baby needs another pre-operative treatment. Treatment options for the newborn are mainly primary surgical excision and if malignancy is proved, then chemotherapy, radiotherapy, or combination of both is used.[7]

Long-term outcome with cervical teratoma is still to be elucidated. According to available data, there are risks for serious thyroid conditions, hyperparathyroidism, hypothyroidism, developmental delay and mental retardation, malignant transformation, recurrence, metastasis to regional lymph nodes. Also, only one case of occurrence among siblings has been reported. Follow-up recommendations warrant AFP levels to be obtained at birth, at 1 month, 3-month intervals in infancy, and yearly thereafter, up to 3 years of life. MRI scanning twice a year for the first 3 years of life is also suggested.[8] Prognosis depends upon several factors, most important being degree of maturity of tissues and completeness of resection. Others include airway obstruction at birth and associated anomalies.[9]

CONCLUSION

The fine needle biopsy can also be done without fear of bleeding. Cyto/histologic diagnosis is necessary before going for the operation to know whether the baby needs another pre-operative treatment. CT scan is also helpful for staging and diagnostic purpose. Surgery of these mostly benign malformations is very challenging for a multidisciplinary team. Long-term follow-up is done by estimation of relative levels of pre-and post-operative levels of tumors markers.

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