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Fetal Nasopharyngeal Teratoma: An Autopsy Case Report

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ABSTRACT

Teratoma is a rare type of tumor that can contain fully developed tissues and organs, including hair, teeth, muscle, and bone. Teratomas are most common in the tailbone, ovaries, and testicles, but can occur elsewhere in the body. These tumors originate from all three embryonic germ layers: ectoderm, mesoderm, and endoderm. Nasopharyngeal teratomas are responsible for a high birth mortality rate from acute respiratory distress. We report a rare case of pharyngeal mass in a fetus with antenatal ultrasound and autopsy findings. A 31-year-old primigravid woman with history of insulin-dependent diabetes mellitus and treated hypothyroidism presented with severe polyhydarmnios, prenatal ultrasound revealed a heterogenous solid cystic mass in submandibular region with no vascularity on Doppler exam. The postmortem and pathologic examination confirmed the nasopharyngeal teratoma with extensive cervical extension. The aim of this study was updating current knowledge about this disease.

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Introduction

In 1863 Virchow used the term “teratoma” that derived from the Greek word “teraton” (meaning monster(1,2). Teratoma is a true neoplasm and is the most common mass encountered in neonates (3). This is consists of tissues from all three embryonic germ layers: ectoderm, mesoderm, and endoderm (4-6).

Teratomas occur in 1 of 4000 live births with head and neck teratomas accounting for less than 5% of the total (5, 7, 8) and representing 25–35% of neonatal tumours (9). Head and neck teratomas are most commonly cervical with the nasopharynx being the second commonest location (10, 11). Teratomas in the upper areo-digestive tract (UADT) are rare neoplasms, accounting for fewer than 2% of all teratomas (12).

The appearance of nasopharyngeal teratomas varies according to the size, degree of tissue heterogeneity and degree of tissue maturation (9). The most common location for teratomas to be seen within the UADT mucosa is the nasopharynx; other less commonly involved sites include the oral cavity (tonsil, tongue, and palate), sinonasal cavity and the ear and temporal bone. Nasopharyngeal teratoma presents as a mass protruding into the oral cavity or pharynx causing dysphagia and/or airway obstruction (9,13).

True teratoma is a congenital neoplasm of tridermal origin (14). Teratomas may be associated with maternal polyhydramnios, preeclampsia, preterm delivery and respiratory compromise after birth due to upper airway ob-

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struction by the mass and stillbirth (2,15).

A fetal teratoma originating from the base of the skull is called pharyngeal teratoma or epignathus and is associated with other anomalies (16,17). Epignathus is a rare type of teratoma for which accurate prenatal diagnosis is definitive to plan proper peripartum management (10). It usually carries a poor prognosis (15).

Here we report a case of fetal pharyngeal teratoma with ultrasound postmortem findings and review literature.

Case report

A 31-year-old primigravid woman with history of insulin-dependent diabetes mellitus and treated hypothyroidism, underwent a first-trimester ultrasound examination at 13 weeks, which was normal. Neither the first trimester screening test (ultrasound and biochemical markers screening test for aneuploidies) nor second trimester anomaly scan and quadrant blood test (Alpha-fetoprotein (AFP), free β-human chorionic gonadotropin (β-HCG), free estriol, and inhibin A) were performed.

She had no positive history for stillbirth or previous abortion and no malformed neonates within her first relatives. Sonographic examination by a Medison V20 with multi-frequency 3-5 MHz convex probe at 29 weeks of gestation, revealed a single live fetus with biparietal diameter of 72mm, head circumference of 267 mm, femur length of 55mm and abdominal circumference of 251 mm. The placenta was located on the uterine fundus. The amount of amniotic fluid was severely increased (Amniotic fluid Index was 40 cm).

There was a heterogeneous solid-cystic appearance mass with diameters of 48*32*21mm filling the submandibular region bilaterally caused partial opening of the mouth and deformity of profile fetal face (Fig 1a&b). No calcified or echogenic elements were found within the mass and no vascularity was detected by color Doppler ultrasound.

Other fetal organs were unremarkable. The differential diagnosis included cystic hygroma, hemangioma and teratoma. After counseling the parents decided to terminate the pregnancy at the 30 weeks' gestation. Cesarean section was performed and a 1300gm live girl was born but because airway access was too difficult due to the mass, intubation was failed and it shortly died.

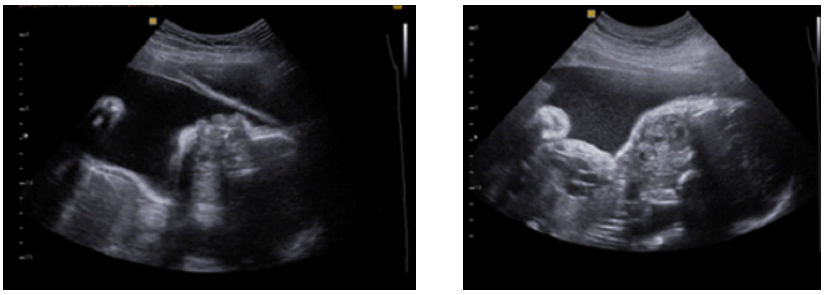

Figure 1a&b. Sagittal sonogram of the fetal head at 30 weeks' gestation shows a complex mass containing solid-cystic elements (arrows) filling the pharynx (a) Coronal view of fetal neck and upper thorax reveals extending mass into the cervical region

The autopsy revealed an exophytic multi-cystic mass in submandibular and suprahyoid region measured 5×3×2 cm which was attached to the nasopharynx and extended into left maxillary sinus superiorly and cervical region inferiorly (fig 2).

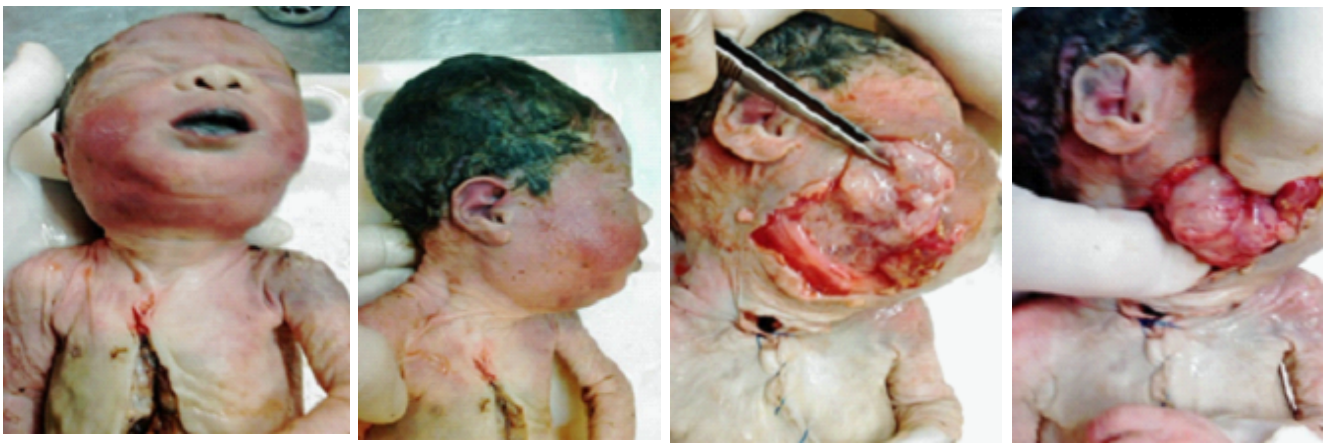

Figure 2. Photographs of the infant reveals bulging of submandibular region and opening mouth due to the pharyngeal mass

Histological analysis showed a mixture of maturing elements of ectodermal, mesodermal and endodermal origin including cartilage, epithelial and neuronal tissue compatible with the diagnosis of pharyngeal teratoma (Fig 3a&b). There were not any other fetal abnormalities.

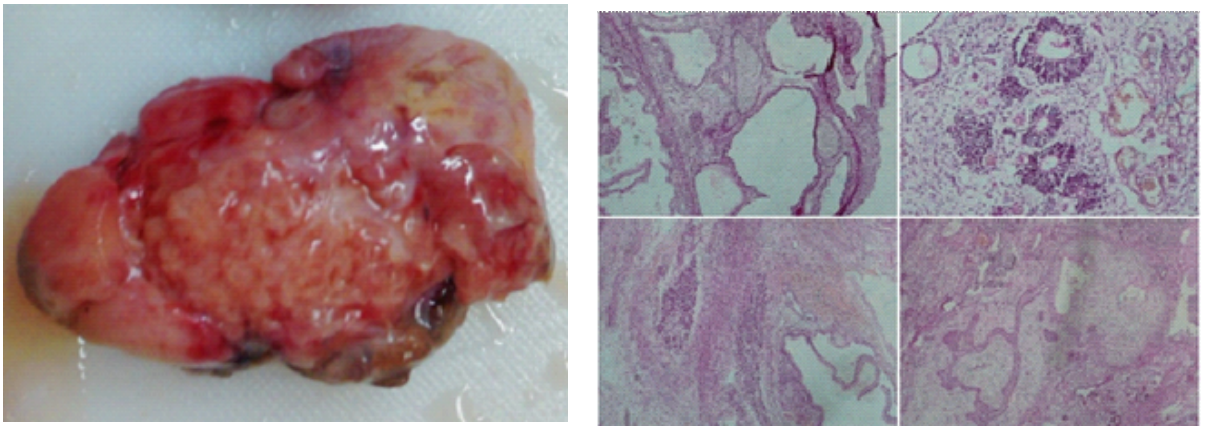

Figure3a&b. Photograph of the teratoma revealed encapsulated solid cystic mass(a) in histopathologic examination a mixture of maturing elements of ectodermal, mesodermal and endodermal origin including cartilage, epithelial and neuronal tissue (b).

Discussion

Here is a report a rare case of pharyngeal mass in fetus with antenatal ultrasound and autopsy findings. High-quality prenatal imaging plays a key role in identifying neonatal teratomas, determining an appropriate airway management strategy and planning for surgical resection (18). Teratomas are tumors derived from pluripotent cells and made of one or more of the three germ cell layers (2,13). Brain tissue is the most frequent component along with cartilage, bronchial epithelium and ependyma-lined cysts (19). They are most commonly located in the neck and the nasopharynx being the second commonest site (10). Teratomas usually occur more frequently in females than in males (9, 19, 20). It can be associated with elevated maternal serum αFP, which requires a detailed ultrasound exam of the fetus. Although elevated levels of MS-αFP have also been reported in fetal defects such as neural tube defects, tracheoesophageal fistula, esophageal obstruction, sacrococcygeal teratoma, and anorectal atresia (21).

In our case maternal serum αFP level was not checked. Epignathus or nasopharyngeal teratoma may be associated with cranial anomalies like palatal clefts, hemicrania and anencephaly. These defects were not detected in the present case. The diagnosis can be suspected by ultrasound exam, especially in epignathus with a protruding mass from the mouth (2).

No environmental risk factors is reported in pathogenesis of pharyngeal teratoma (16). Additionally, no association with some karyotype abnormalities is proven and these lesions are not thought to be inherited in a Mendelian or polygenic fashion (15,20).The prognosis of pharyn geal teratoma is very poor (15). Intrauterine fetal demise in up to 50% and neonatal demise of 50% of the survivors have been reported. Only six cases of infants surviving a pharyngeal teratoma have been reported over the last 10 years (19).

Conclusion

Fetal pharyngeal masses including teratoma can be suggested by ultrasound but definite diagnosis is made by a pathological analysis. The teratoma may be diagnosed antenatally on ultrasound, which permits early multidisciplinary management.

Conflict of Interest

The authors declare no conflict of interest.

References

1. Izadi K, Smith M, Askari M, et al. A patient with an epignathus: management of a large oropharyngeal teratoma in a newborn. J Craniofac Surg. 2003;14:468-472.
2. Singhvi I, Pawar JGJISJ. Oral teratoma-the devil's curse: a case report. J Craniofac Surg. 2003;14:468-472.
3. Venkatesh IH, Venkatesha L, Sathi V, et al. Nasopharyngeal teratoma: An unusual cause for respiratory distress in a neonate. J Craniofac Surg. 2003;14:468-472.
4. He J, Wang Y, Zhu H, et al. Oral Medicine, Oral Pathology, Oral Radiology,, Endodontology. Nasopharyngeal teratoma associated with cleft palate in newborn: report of 2 cases. Oral Surg Oral Med Oral Pathol Oral Radiol Endod. 2010;109:211-216.
5. Thong HK, Lamry NA, Kamalden TMIT, et al. Congenital nasopharyngeal teratoma: a potential cause of neonatal airway obstruction. RMJ. 2018; 43: 562-564.
6. Kaplan I, Allon I, Shlomi B, et al. A comparative study of oral hamartoma and choristoma. J Interdiscip Histopathol. 2015; 3: 129-134.
7. Tonni G, De Felice C, Centini G, et al, obstetrics. Cervical and oral teratoma in the fetus: a systematic review of etiology, pathology, diagnosis, treatment and prognosis. Arch Gynecol Obstet. 2010;282:355-361.
8. Tiwari L, Baijal N, Puliyel JMJIP. Nasopharyngeal teratoma as a cause of neonatal stridor. Indian Pediatr. 2009;46:1097-1098.

9. Diakité C, Bénateau H, Dakpé S, et al. Management of nasopharyngeal teratomas associated with cleft palate. Int J Oral Maxillofac Surg. 2019;48:291-297.
10. Shah F, Raghuram K, Suriyakumar G, Dave A, Patel V. Congenital teratoma of nasopharynx. Ind J Radio Imag. 2002;12:201–202.
11. Mishra N, Mandelia A, Naranje K, et al. Case of nasopharyngeal teratoma: challenges in the management. BMJ Case Rep. 2019;12:e230105.
12. Anand B, Raman EV, Rakshit SHJIJoO, et al. Mature Nasopharyngeal Teratoma in a Neonate: Case Report and a Review of Literature. Indian J Otolaryngol Head Neck Surg. 2019;71:1775-1778.
13. Huang Y, Pan HJIjoc, pathology e. A fetus with a mass in the oral cavity: a rare case of oral immature teratoma. Int J Clin Exp Pathol. 2017;10:7890-7892.
14. Haghighi K, Milles M, Cleveland D, et al. Epignathus teratoma with bifid tongue and median glossal salivary mass: report of a case. J Oral Maxillofac Surg. 2004;62:379-383.
15. Clement K, Chamberlain P, Boyd P, et al. Prenatal diagnosis of an epignathus: a case report and review of the literature. Ultrasound in Obstetrics and Gynecology: Ultrasound Obstet Gynecol. 2001;18:178-181.
16. Sarioglu N, Wegner R, Gasiorek-Wiens A, et al. Epignathus: always a simple teratoma? Report of an exceptional case with two additional fetiforme bodies. Ultrasound Obstet Gynecol. 2003;21:397-403.
17. Brodsky JR, Irace AL, Didas A, et al. Teratoma of the neonatal head and neck: a 41-year experience. Int J Pediatr Otorhinolaryngol. 2017 Jun;97:66-71.
18. Calda P, Novotna M, Čutka D, et al. A case of an epignathus with intracranial extension appearing as a persistently open mouth at 16 weeks and subsequently diagnosed at 20 weeks of gestation. J Clin Ultrasound. 2011;39:164-168.
19. Varras M, Akrivis C, Plis C,et al. Antenatal sonographic diagnosis of pharyngeal teratoma: our experience of a rare case with review of the literature. Obstet Gynecol Int. 2009;2009:180643.
20. Bornstein E, Boozarjomehri F, Monteagudo A, et al. Diagnostic and prognostic aspects in the sonographic evaluation of a fetus with an oral mass. J Ultrasound Med. 2009;28:689-693.