

Case Report

Epignathus (Oropharyngeal Teratoma) in 20 Week Fetus: A Case Report

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Abstract

Epignathus sometimes called (oropharyngeal teratoma) is a very rare cervical teratoid tumor that arises from the oropharyngeal region; it is not often consistent in origin, number, and differentiation of tissues. It usually have components from all three germ layers and can contain cartilage, fat, and bone.

In this paper we present the case of ultrasound scan of pregnant female in 20 week with fetal cervical tumor arising from oropharyngeal region.

Keywords: Epignathus; Teratoma; Germ cell; MRI

Introduction

Epignathus solid tumor arise from sphenoid bone, palate, the pharynx, the tongue and jaw [1-3]. The predilection is slightly more in female the estimated incidence is ~ 1 in 35,000 to 200,000 births.

The tumor rarely may present in young but it is classically presents in utero or in the neonatal period [4]. It typically arises from around the basisphenoid (Rathke's pouch) in the palato-pharyngeal region, and with progressive growth fills the buccal cavity and finally protrudes out of the mouth. When there is associated swallowing impairment may Associations with polyhydramnios, mid line facial defects like cleft lip and/or palate.

Case Presentation

Healthy pregnant woman presented to her obstetrician for a routine visit. Physical examination was normal, Antenatal ultrasound scan was seen as a complex mass protruding from the fetal oral cavity with mixed echogenicity (Figure 1) [5]. Accompanying polyhydramnios presented which mean there is swallowing impairment (Figure 2). This case marked as raised maternal serum alpha feto protein (MSAFP) (Figure 3).

Discussion

Fetal tumors and teratomas

Fetal tumors are rare; teratomas are considering the most frequent type, which have an incidence of between 1:20,000 and 1:40,000 live births [6]. Teratomas may present in several locations. The rare form of fetal teratoma originating from the base of the skull is named epignathus. MRI is useful for better delineation of anatomy (Figure 4) [7].

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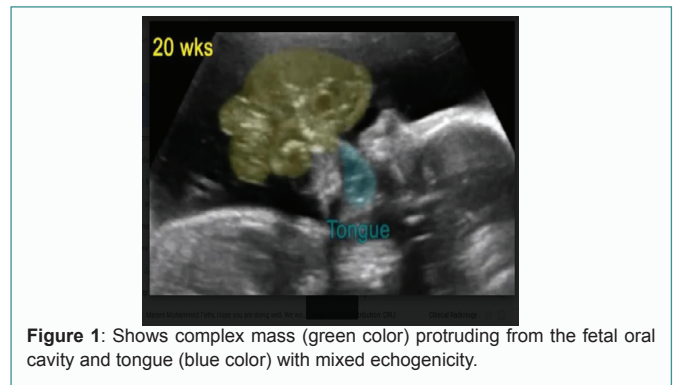


Figure 1: Shows complex mass (green color) protruding from the fetal oral cavity and tongue (blue color) with mixed echogenicity.

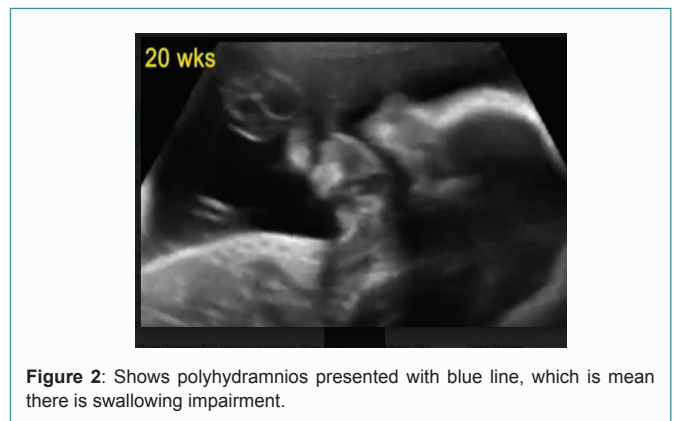


Figure 2: Shows polyhydramnios presented with blue line, which is mean there is swallowing impairment.



Figure 3: Shows complex mass protruding from the fetal oral cavity and tongue with mixed echogenicity.



Figure 4: Aspect of the fetus with the craniopharyngeal teratoma (epignathus) following termination of pregnancy at 24+4 gestational weeks (middle). Demonstration of the two parasitic fetuses with the larger first fetiforme body appending the epignathus and the second parasitic fetus which was found in the uterine cavity during Cesarean section (right). Reproduction of a drawing of a case examined by Baart de la Faillie in 1875, published 190722 (left), Urban & Fischer Verlag.

Treatment and prognosis

The prognosis is generally poor (often as a result of its location) and part of the high mortality rate is caused by severe airway obstruction in the neonatal period [8]. The tumor may also demonstrate rapid growth. An EXIT procedure can be offered in selected cases [9].

Complications

Development of fetal hydrops: often due to tumor shunting/using a lot of blood flow the tumor can spread superiorly destroying the skull base and brain tissue [10]. This in turn can give range complications such as fetal hydrocephalus neonatal death usually occurs due from airway obstruction and consequent asphyxia after birth.

Differential diagnosis

In general consider other tumors from the oropharyngeal region: oropharyngeal rhabdomyosarcoma epulis/congenital oral granular cell myoblastoma [11,12]. For small lesions on a sagittal ultrasound scan also consider: macroglossia prominent philtrum/premaxillary tissue from a midline facial defect (can also be an association) [13,14].

Conclusion

We present here a rare of the largest fetal tumor arising in a oropharyngeal location, extension from sphenoid bone, palate, the pharynx, the tongue and jaw. Ultrasound has very high sensitivity and low false-positive rates in identifying fetal epignathus prenatally. The risk of chromosomal abnormalities is very low in fetuses with teratoma, and their prognosis depends on the location and size of the tumor and any associated perinatal complications.

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