Case Report

Prenatal Diagnosis and Immediate Successful Management of Isolated Fetal Epignathus

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Abstract

Epignathus or enigmatic teratoma is rare congenital tumor, arising from the sphenoid region of the palate or pharynx. A 30-year-old pregnant woman presented at 34 weeks + 2 days' gestation with a well-defined solid mass measuring 6.47 cm × 5.7 cm arising from the fetal oral cavity. The color flow mapping showed that the mass was well vascularized, especially in its center, and its blood supply was originated from the fetal oral cavity. The newborn has no chromosomal abnormalities or other associated anomalies and/or intracranial extension immediately and successfully managed after delivery to highlight the importance of prenatal diagnosis and the importance of multidisciplinary team management.

Keywords: Diagnosis, epignathus, fetal, management, prenatal

Introduction

Epignathus or enigmatic teratoma is rare congenital tumor, arising from the sphenoid region of the palate or pharynx. The incidence of the enigmatic teratoma is 1 in 35,000–1 in 200,000 live births.[1] The tumor produces airway obstruction and is associated with high neonatal mortality rate (80%–100%).[2,3] Epignathus is usually detected in the second or third trimesters using the two-dimensional (2D) ultrasound;[2,3] it can be detected early using the three-dimensional (3D) ultrasound and/or magnetic resonance imaging (MRI) technology.[4,5]

This case report represents the prenatal diagnosis and the immediate successful management of fetal epignathus.

Case Report

A 30-year-old pregnant woman, P1 (previous cesarean delivery), referred to Ain Shams University Fetal Care Unit as pregnant female 34 weeks + 2 days' gestation with fetal umbilical cord mass for further evaluation. Further evaluation showed that she is pregnant 35 weeks' gestation ± 1 week with estimated fetal weight of 2.639 kg, good amniotic fluid volume, normal umbilical cord, and a well-defined mass measuring 6.47 cm × 5.7 cm arising from the fetal oral cavity [Figure 1].

The color flow mapping showed that the mass was well vascularized, especially in its center [Figures 2 and 3], and its blood supply was originated from the fetal oral cavity [Figure 4].

The studied case managed by multidisciplinary team consists of senior obstetrician, neonatologist, anesthetist, and pediatric surgeon.

Fetal MRI was done before delivery to study the relation of the mass to the fetal airways and to exclude intracranial extension of the mass.[1] T2-weighted MRI showed a hyperintense mass with a stalk projecting from the palate and upper lip, measuring 6.7 cm × 5.7 cm in size with no evidence of fetal airway obstruction and/or intracranial extension or central nervous system-related anomalies.

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She delivered by elective cesarean section a baby boy – 3.550 kg and Apgar score 7, 8, and 9 at 1, 5, and 10 min, respectively, at 39 weeks' gestation[6,7] according to the hospital protocol with the prescribed oral mass by ultrasound protruding from the newborn oral cavity [Figures 5 and 6].

Immediately after delivery and neonatal resuscitation, the mass was surgically excised 6–8 h after delivery [Figure 7].

The histopathological microscopic examination of the excised mass showed the presence of hair follicles with mature adipocytes and keratinized squamous epithelium

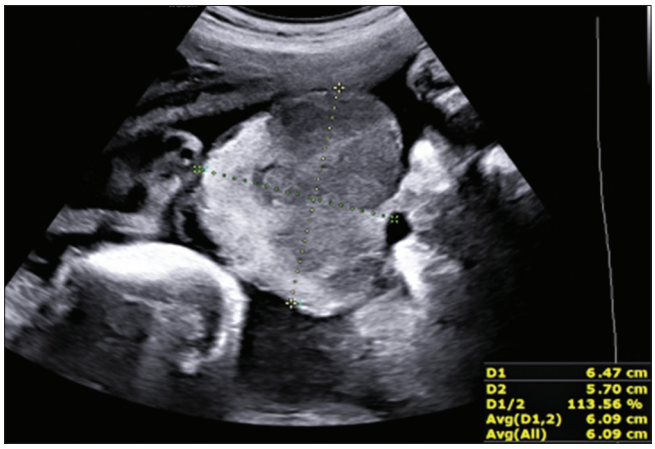

Figure 1: Well-defined mass measuring 6.47 cm × 5.7 cm arising from the fetal oral cavity

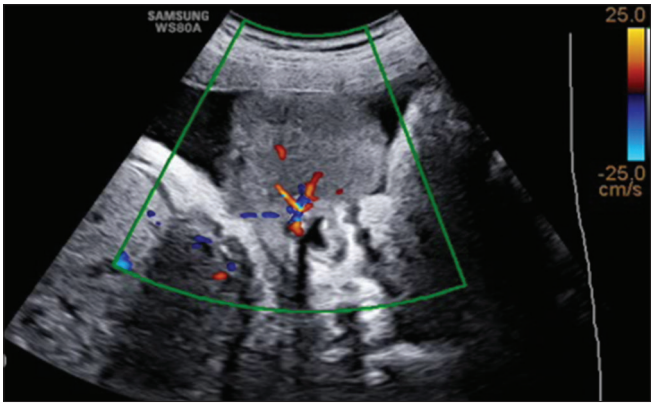

Figure 2: Color flow mapping showed that the mass was well vascularized, especially in its center

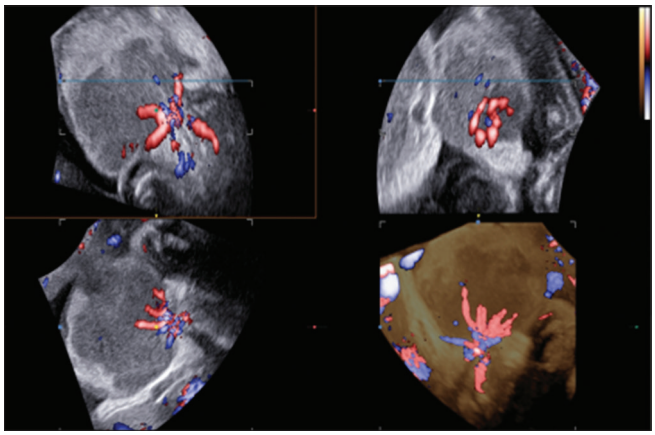

Figure 3: Three-dimensional color flow mapping showed that the mass was well vascularized, especially in its center

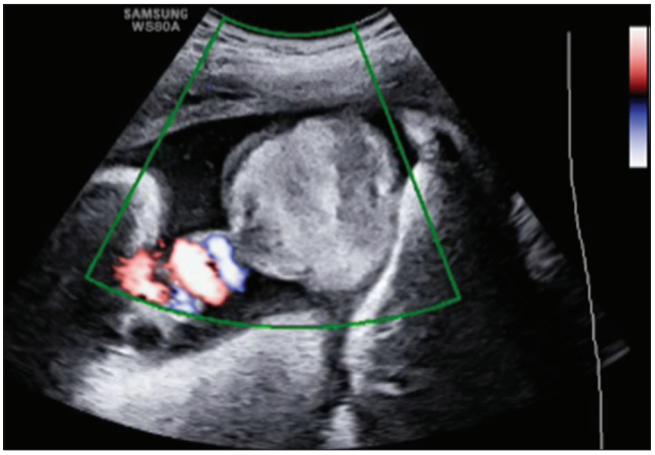

Figure 4: Color flow mapping showed the blood supply of the mass originated from the fetal oral cavity

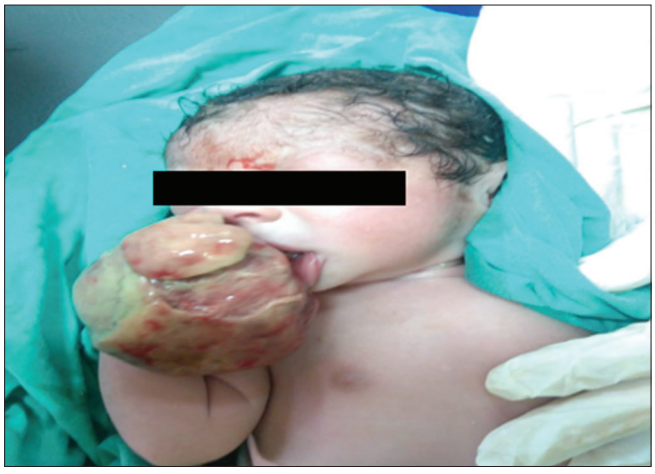

Figure 5: Studied neonate immediately after delivery

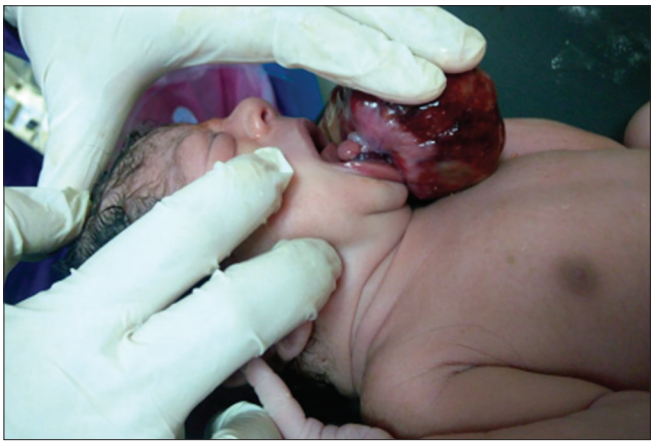

Figure 6: Studied neonate 8 h after delivery, before general anesthesia and excision of the mass

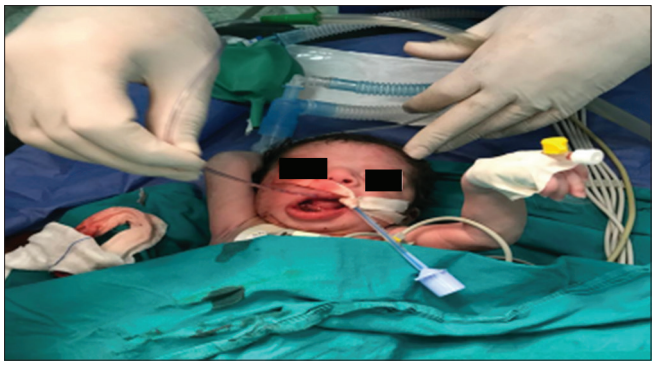

Figure 7: Studied neonate during recovery from general anesthesia after removal of the mass

consistent with mature fetal enigmatic teratoma or epignathus.

The newborn admitted to the neonatal subcare unit for 2 postoperative days and discharged from the neonatal subcare unit in good general condition without any postoperative complications after chromosomal studies which showed (46, XY) normal karyotype.

Discussion

Epignathus or enigmatic teratoma is usually associated with other fetal malformations in 6% of cases.[8] Cleft palate is the most common malformation associated with epignathus because the tumor prevents closure of the palate.[8] Other associated malformations are bifid tongue and/or nose, glossoptosis, diaphragmatic, and inguinal hernias.[8,9] Bifidity and glossoptosis indicate early development of the epignathus with subsequent anterior positioning of the tongue and impaired fusion of the primitive tongue buds.[1] Functional mandibular microretrognathia can be seen secondary to glossoptosis due to impaired fetal jaw's growth.[8,9]

This case report represents a case of isolated epignathus without chromosomal abnormalities or other associated anomalies and/or intracranial extension immediately and successfully managed after delivery, and the newborn was discharged from the neonatal subcare unit 2 days after surgery to highlight the importance of prenatal diagnosis and the importance of multidisciplinary team management.

Other authors reported that epignathus may be associated with embryonic defects and/or chromosomal abnormalities.[1,8]

Epignathus is usually diagnosed by the 2D ultrasound as a protruding mass related to the fetal oral cavity, partially solid and partially cystic, and is associated with polyhydramnios in 30% of cases due to obstruction of the fetal mouth and impaired fetal swallowing.[1]

The suspicious diagnosis of epignathus can be confirmed with the 3D ultrasound in most of the cases,[4] and the reconstructed images from the 3D volume data can provide more detailed information about the origin and the extension of the epignathus.[5]

MRI is a complementary diagnostic tool for epignathus to detect the relation of the tumor to the fetal airway and intracranial structures.[1] The MRI done before delivery showed no evidence of fetal airway obstruction and/or intracranial extension or central nervous system-related anomalies.[10]

If epignathus produces fetal tracheal obstruction, some authors suggest delivery by cesarean section with immediate tracheostomy and intubation before umbilical cord clamping while the newborn is on placental support, followed by surgical removal of the tumor after resuscitation and stabilization of the newborn to correct the airway or the tracheal obstruction.[11,12]

The prognosis of epignathus depends on the size, associated polyhydramnios, associated other fetal anomalies, and the degree of intracranial extension.[1,9] Malignant degeneration rarely occurs in epignathus.[1] Large epignathus interfering with fetal swallowing may lead to polyhydramnios and prematurity with subsequent pulmonary hypoplasia and respiratory distress syndrome following preterm labor and/or preterm rupture of fetal membranes.[1] Poor prognosis usually encountered with epignathus is associated with embryonic defects and/or chromosomal abnormalities.[1,12]

Conclusion

Epignathus or enigmatic teratoma is a rare fetal teratoma arising from the sphenoid region of the palate or pharynx. The studied newborn has no chromosomal abnormalities or other associated anomalies and/or intracranial extension immediately and successfully managed after delivery to highlight the importance of prenatal diagnosis and the importance of multidisciplinary team management.

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Informed consent

The parents of the studied neonate signed a written consent for publication of their neonate data and photos.

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Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Moon NR, Min JY, Kim YH, Choi SK, Shin JC, Park IY. Prenatal diagnosis of epignathus with multiple malformations in one fetus of a twin pregnancy using three-dimensional ultrasonography and magnetic resonance imaging. Obstet Gynecol Sci 2015;58:65-8.
2. Kumar SY, Shrikrishna U, Shetty J, Sitaram A. Epignathus with fetiform features. J Lab Physicians 2011;3:56-8.

3. Tonni G, Centini G, Inaudi P, Rosignoli L, Ginanneschi C, De Felice C, *et al.* Prenatal diagnosis of severe epignathus in a twin: Case report and review of the literature. Cleft Palate Craniofac J 2010;47:421-5.
4. Takagi MM, Bussamra LC, Araujo Júnior E, Drummond CL, Herbst SR, Nardozza LM, *et al.* Prenatal diagnosis of a large epignathus teratoma using two-dimensional and three-dimensional ultrasound: Correlation with pathological findings. Cleft Palate Craniofac J 2014;51:350-3.
5. Allen LM. Prenatal 3-dimensional imaging techniques in the sonographic evaluation of an oral mass: Comparison with postnatal imaging modalities. J Ultrasound Med 2011;30:561-8.
6. Abdelazim I, Farghali MM, Elbiaa AA, Abdelrazak KM, Hussain M, Yehia AH, *et al.* Impact of antenatal oxytocin infusion on neonatal respiratory morbidity associated with elective cesarean section. Arch Med Sci 2017;13:629-34.
7. Farghali M, Abdelazim I, Abdelrazek K. Delayed second twin delivery: benefits and risks. J Matern Fetal Neonatal Med 2019;32:1626-32.
8. Tonni G, De Felice C, Centini G, Ginanneschi C. Cervical and oral teratoma in the fetus: A systematic review of etiology, pathology, diagnosis, treatment and prognosis. Arch Gynecol Obstet 2010;282:355-61.
9. Pasupathy M, Narayanan PV, Mani V, Adenwalla HS. A case report of nasopharyngeal teratoma with a cleft palate and an inguinal hernia. J Plast Reconstr Aesthet Surg 2011;64:1525-7.
10. Durukan H, Gundogan S, Çevikoğlu Kıllı M, Dilek TU. Oral mass of a fetus incidentally found during second trimester: Ultrasound survey and postnatal prognosis of the baby. Case Rep Obstet Gynecol 2018;2018:6590710.
11. Sichel JY, Eliashar R, Yatsiv I, Moshe Gomori J, Nadjari M, Springer C, *et al.* A multidisciplinary team approach for management of a giant congenital cervical teratoma. Int J Pediatr Otorhinolaryngol 2002;65:241-7.
12. Noah MM, Norton ME, Sandberg P, Esakoff T, Farrell J, Albanese CT. Short-term maternal outcomes that are associated with the EXIT procedure, as compared with cesarean delivery. Am J Obstet Gynecol 2002;186:773-7.