

Case Report

Prenatal Diagnosis and Immediate Successful Management of Isolated Fetal Epignathus

Mohamed ElSherbiny Hamed¹, Mohamed Hassan Naser El-Din¹, Ibrahim A. Abdelazim^{1,2*}, Svetlana Shikanova³, Bakyt Karimova³, Sakiyeva Kanshaiym³

¹Department of Obstetrics and Gynecology, Ain Shams University, Cairo, Egypt,

²Department of Obstetrics and Gynecology, Ahmadi Hospital, Kuwait Oil Company, Ahmadi, Kuwait,

³Department of Obstetrics and Gynecology №1, Marat Ospanov, West Kazakhstan State Medical University, Aktobe, Kazakhstan

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.

Address for correspondence: Prof. Ibrahim A. Abdelazim,

Department of Obstetrics and Gynecology, Ain Shams University, Cairo, Egypt.

Department of Obstetrics and Gynecology, Ahmadi Hospital, Kuwait Oil Company, P. O. Box: 9758, 61008 Ahmadi, Kuwait.

E-mail: dr.ibrahimanwar@gmail.com

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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

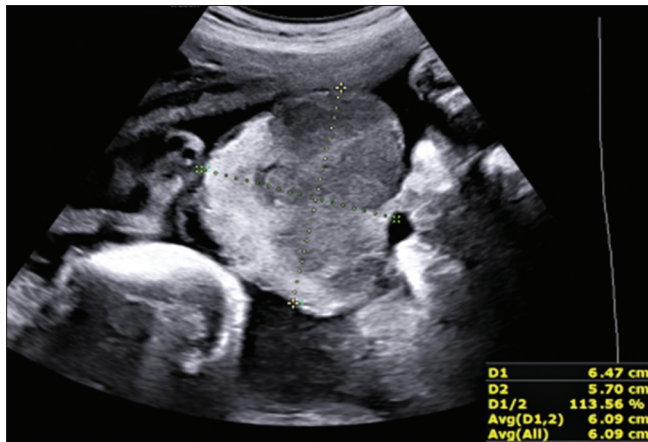


Figure 1: Well-defined mass measuring 6.47 cm × 5.7 cm arising from the fetal oral cavity

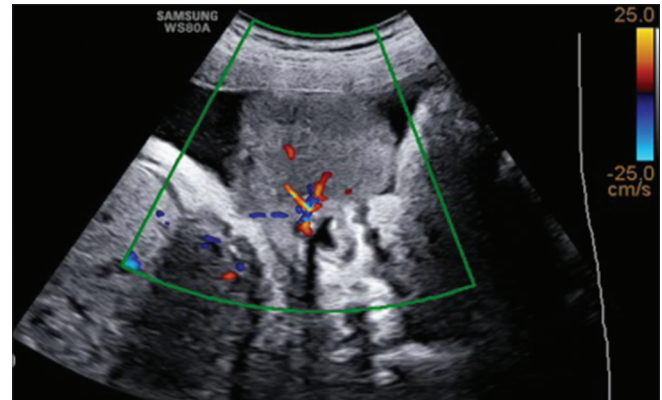


Figure 2: Color flow mapping showed that the mass was well vascularized, especially in its center

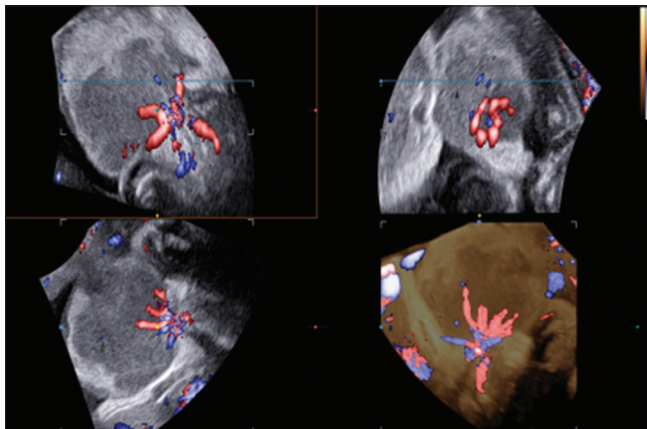


Figure 3: Three-dimensional color flow mapping showed that the mass was well vascularized, especially in its center

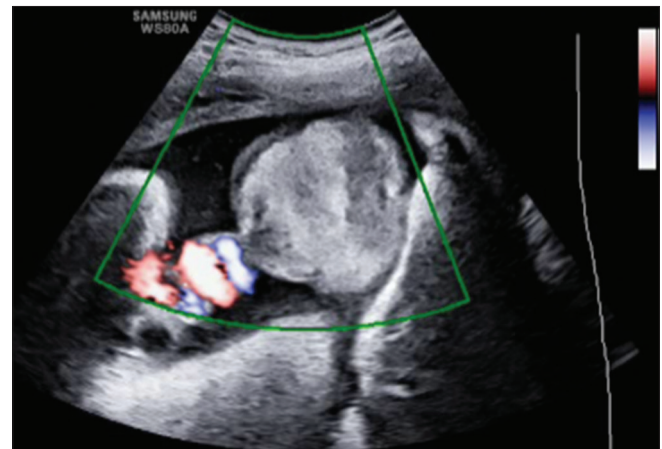


Figure 4: Color flow mapping showed the blood supply of the mass originated from the fetal oral cavity



Figure 5: Studied neonate immediately after delivery



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

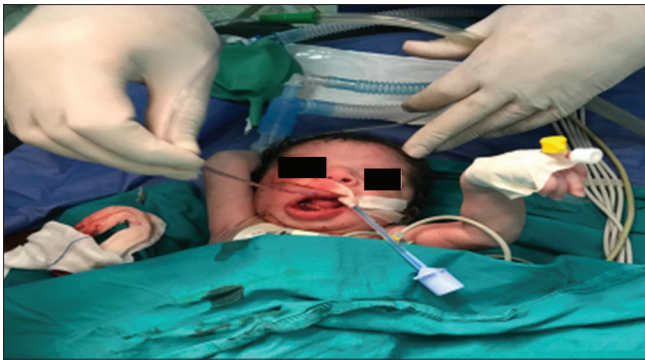


Figure 7: Studied neonate during recovery from general anesthesia after removal of the mass

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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Conflicts of interest

There are no conflicts of interest.

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