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Case Report

Early Prenatal Diagnosis of Congenital Ranula

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ABSTRACT

Congenital ranula (CR) is a rare tumor. Few cases of prenatal diagnosis (PD) of CR have been reported in the literature and diagnosis usually is beyond 20 weeks of gestation. We report a case of a pregnant patient with 16 weeks of gestation, where ultrasound (US) visualized in her fetus, a cystic tumor protruding in his oral cavity. Basis and complementary studies, including fetal echocardiography, were normal. Serial two-dimensional (2D) and three-dimensional (3D) US studies monitored fetal growth, amniotic fluid, progressive changes in the cystic mass, and maxillary movements. At 38.5 weeks of gestation, a cesarean section was performed. At 17th neonate days old, the neonate was done the simple and complete excision. Early PD of CR is possible. In this case was done as early as at 16 weeks of gestation. This is the first CR reported case with the earliest PD (16 week of gestation). We recommend that during the first trimester US, the oral cavity must be well explored, to rule out tumors.

Keywords: Congenital ranula, Mucocele, Congenital cystic malformation, Congenital high airway obstruction syndrome, Noninvasive prenatal diagnosis

INTRODUCTION

Congenital ranula (CR) is a rare tumor, caused either by a mucus extravasation cyst secondary to obstruction and rupture of sublingual or minor salivary glands, or, less commonly, to a mucus retention cyst, originating from dilatation of an obstructed submandibular gland duct; and therefore surrounded by epithelium. Varies from 0.74 to 1.2/1000.[1-6] Few cases of prenatal diagnosis (PD) of CR have been reported in the literature and diagnosis usually is beyond 20 weeks of gestation.[1-3] We report a CR case, diagnosed as early as at 16 weeks of gestation, and review the literature.

CASE REPORT

A 21-year-old pregnant patient, with no important personal or family history, G3 P2, during her routine prenatal control, at 16 weeks of gestation; by two-dimensional (2D) ultrasound (US) [Figure 1a], the open mouth was viewed in the fetus, and a rounded image of anechoic or cystic appearance, homogenous, and thin-walled, with no flow to the color Doppler, was visualized in the oral cavity. Three-dimensional (3D) US [Figure 1b] confirmed this image. There were no other congenital malformations or polyhydramnios. During prenatal care, she had a severe urinary infection that warranted hospitalization and parenteral treatment. She then continued antenatal care on an outpatient basis, her complementary studies including fetal echocardiography were normal and rejected the cytogenetic study. Serial 2D and 3D US monitored fetal growth, amniotic

fluid and progressive changes in the cystic mass, and observed maxillary movements and were unremarkable. US was performed using a GE Voluson 730 Expert and a Medinson-Samsung Accuvix XG. At 38.5 weeks of gestation, an elective cesarean section was done; obtaining a male newborn, alive, 2.900 g, 49 cm, with Apgar scores of 8/10 and 9/10 at 1' and 5' of life. On clinical examination, the infant had a tumor on the ventral side of the tongue, 5 cm × 5 cm [Figure 2]. The patient was discharged 24 h postoperatively and the post-operative period was uneventful. A newborn was approached by a multidisciplinary team including obstetricians, neonatologists, and maxillofacial surgeons; he maintained the suction reflex and was fed orally without needing a nasogastric tube. At 17 days old, he had decreased 800 g and simple and complete surgical excision was performed [Figure 3 top, middle and bottom left], the post-operative period was also uneventful [Figure 3 bottom center]. Four weeks later, the child was healthy, showed a normal-sized tongue, and had no feeding difficulties [Figure 3 bottom right]. Histological report confirmed the diagnosis.

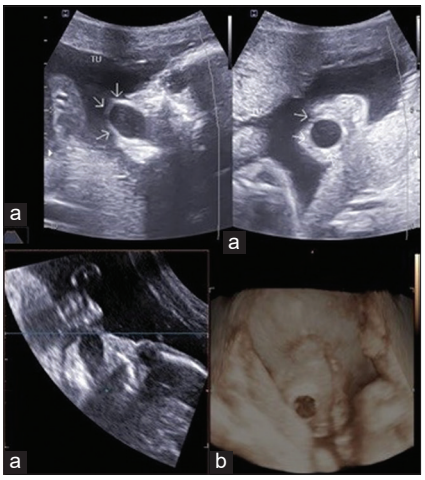

Figure 1: A 21-year-old woman at 16 weeks of gestation 2D ultrasound (top and bottom left), showed in the fetal oral cavity a rounded image of anechoic or cystic aspect, homogeneous, thinwalled, with no flow to the color Doppler assessment. 3D image (bottom right), showed the cystic mass in the fetal oral cavity.

Figure 2: Neonate's physical examination showed a cystic tumor on the ventral side of the tongue.

DISCUSSION

PD of congenital head and neck masses is becoming more frequent with the improvement of US technology, although it can be difficult to establish the etiology, which often is done by the histological examination.[1,7] CR may be diagnosed by prenatal US. With 2D US, is noted at the level of the floor of the mouth, a cystic, anechoic and avascular with color Doppler mass, which sometimes moves with fetal swallowing movements, displacing the tongue upward; that, in cases of large dimensions, can cause polyhydramnios.[3,5] The lesion lacks color Doppler flow inside and since it has elastic characteristics, it changes in shape with the reflex movements of suction. Other described PD method is 3D. US, which can play a role as adjuvant to 2D US, allowing a spatial evaluation of the relation between the mass and the oral cavity of the fetus. 3D US and magnetic resonance can also help diagnosis and differential diagnosis.[1] In the present case, diagnosis of the cystic lesion with 2D US was facilitated by the US findings of homogeneity, thin and regular walls, and an avascular mass to the color Doppler [Figures 1a and b]. The findings with 3D

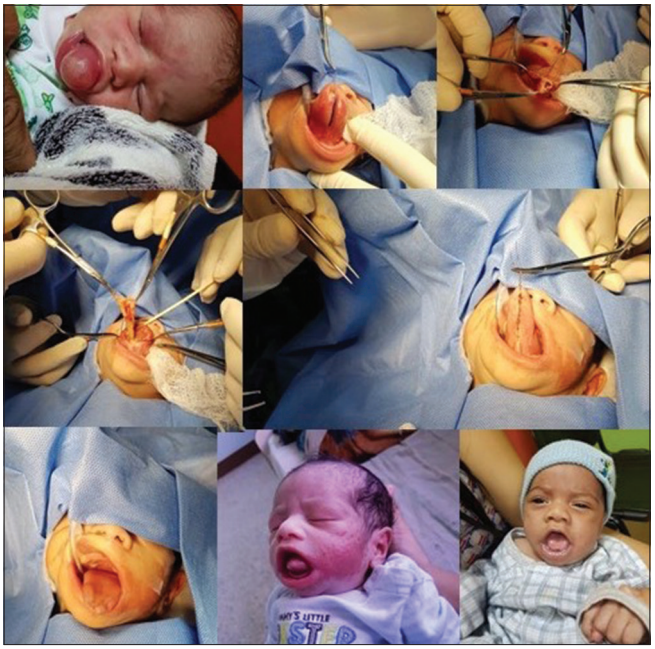

Figure 3: At seventeen days of age, complete surgical excision of the cystic tumor was performed (top, middle and bottom left) in the neonate. Bottom center, the neonate on the first postoperative day. Bottom right, the neonate, 30 days after surgery.

US were similar to the previous ones. Correct PD is extremely important because, generally, such neonates present varying degrees of airway obstruction, which requires the presence of a multidisciplinary team at the time of birth.[1,5]

The literature reported that gestational age in which the PD is done is: Swiss Society of Neonatology,[8] 20 weeks. Onderoglu *et al.*[2] and Marques *et al.*,[5] 21 weeks; Pires *et al.*,[3] 25 weeks; Fernandez Moya,[4] 26 weeks; and Rios *et al.*,[1] 29 weeks. This case was diagnosed at 16 weeks of gestation.

Differential diagnosis must be done with epignathus, epulis, oropharyngeal teratoma, gingival cysts (palatine), hamartomas, lymphangiomas, thyroglossal duct cyst, congenital anomalies of the submandibular canal, heterotopic gastric cysts, and enterocystomas.[2-5] Epignathus is an unusual lesion estimated to affect between 1 in 35.000 and 1 in 200.000 live births. It is a benign teratoma mostly attached to the palatal or pharyngeal surface. Occasionally, these lesions may have an intracerebral extension. They are usually unresectable and affected infants usually die secondary to respiratory compromise. It is crucial that a timely tracheostomy is available during birth, while the fetomaternal circulation is uninterrupted.[2] The congenital epulis is a rare smooth mass, which occurs on the alveolar ridges, more common in the upper maxillary and in the female sex. In 10% of cases is a multiple lesion. It is a self-limiting lesion that responds to conservative excision.[2,9] The gingival or palatal cysts of the newborn are common and self-resolving. They originate from embryonic epithelial rests.[2] Vascular hamartomas represent proliferation of lymphatic or blood vessels.[2] Oral lymphangiomas can present superficially mostly on the tongue or they can less commonly present as deeper, poorly defined masses.[2]

Antenatal management involves US follow-up and early detection of the main obstetrics complications (polyhydramnios and preterm birth),[7] and other fetal morphological alterations. In our case, antenatal evolution was normal. US follow-up focused on fetal growth, amniotic fluid, maxillary movements and changes in the size, and position of cystic mass and showed no changes.

If possible, delivery should be at term, in a hospital at the tertiary level. The delivery route should be considered according to individual evolution, with a team of neonatologists and maxillofacial surgery. If the lesion is of large dimensions and/or obstruction of the upper airway is suspected, causing respiratory distress, before interrupting the fetoplacental connection, ex-uterus intrapartum treatment (EXIT) must be executed.[2,3,5] We discussed the mode of delivery and decided a cesarean section to avoid airways obstruction and rupture of the cyst. There was not necessary the aspiration or drainage of the cyst or the EXIT treatment.

Treatment of the ranula may be observation, aspiration, marsupialization, or surgical excision.[3,5] Cases of spontaneous remission have been reported. The most appropriate and definitive treatment should be surgical excision, just as marsupialization or excision is chosen, with a recurrence risk of 5%.[3,5] The most common surgical complication is the rupture of the ranula during surgery, which does not increase the risk of recurrence.[3,5] In our case, simple surgical excision was performed, which evolved normally [Figure 3] and there was no rupture. It is thought that prenatal needle aspiration for tumor decompression prevents postnatal airway obstruction and obviates the EXIT procedure or neonatal intubation, especially if it is performed before birth.[3]

The most important neonatal complications are the loss of suction reflex, respiratory distress, language disorders, and interference with chewing and swallowing.[3,5] The present newborn decreased weight 800 g.

CONCLUSIONS

We report the first CR case with the earliest PD (16 weeks of gestation) and recommend that during the first trimester US, oral cavity must be well explored, to rule out tumors.

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Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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