Diagnosis and Management of Fetal Cervical Masses

Diagnóstico e Abordagem de Massas Cervicais Fetais

Keywords: Fetoscopy; Head and Neck Neoplasms/congenital; Prenatal Diagnosis; Teratoma/congenita
Palavras-chave: Diagnóstico Pré-Natal; Fetoscopia; Neoplasias da Cabeça e Pescoço/congénito; Teratoma/congénito

Dear Editor,

Cervical teratomas are an important cause of cervical neoplasms and represent a rare entity, corresponding to less than 5% of all pediatric teratomas and the main complication is neonatal airway obstruction.[1,2]

We describe a case report of a female newborn with a right-sided cervical mass detected during an ultrasound performed at 24 weeks gestation measuring 32 x 29 x 19 mm (Fig. 1A). At 34 weeks, increased dimensions (75 x 51 x 55 mm) and excessive accumulation of amniotic fluid was observed. Due to high risk of airway obstruction, *ex-utero intrapartum* treatment (EXIT procedure) was performed at 35 weeks, under maternal general anesthesia and fetal cardiotocographic surveillance. This procedure is performed to secure the fetal airway through intubation before delivery and umbilical cord clamping while maintaining uteroplacental circulation. There were no contraindications such as fetal distress or severe maternal disease, and fetal intubation was successful with no post-procedure complications (Fig. 1B). Alpha-fetoprotein and beta-HCG levels were normal. Surgical excision was performed on the sixth day of life, revealing a 70 x 50 mm capsulated heterogenous pre-tracheal lesion with significant structural compression of trachea, esophagus, right carotid artery and right internal jugular vein. The histological examination was compatible with thyroid teratoma, revealing an expansive and lobulated embryonal tumor with multinodular pattern of three germlines compressing unaltered thyroid tissue, without evidence of neuroepithelium, immaturity, or somatic malignancy. After surgery, she presented transient hypocalcemia requiring oral calcium supplementation. Elective extubation was performed on day 12 of life with transition to continuous positive airway pressure and after five days, she was weaned off to spontaneous breathing. Post-operative management of stridor due to iatrogenic unilateral vocal cord paralysis and hypothyroidism included transient non-invasive ventilation and levothyroxine, respectively. Follow-up showed no other long-term complications.

Prenatal detection of these masses is essential to ensure proper management. Although less than 5% of congenital cervical teratomas are malignant, the risk increases with diagnostic and therapeutic delay.[2] Mortality can reach 80% if not properly treated, mainly due to obstructive respiratory complications.[3] The EXIT procedure has revolutionized the management of congenital malformations with upper airway obstruction.[4] Surgical excision should be performed expeditiously to reduce respiratory impairment and to prevent complications such as ulceration, infection, coagulopathy and malignant transformation.[2] A surgical mortality rate of 15% has been reported, and tumor recurrence is generally considered to be the result of incomplete excision.[2]

Prenatal ultrasound of cervical masses allows planned intervention during the perinatal period. Prompt airway management and early excision are crucial aspects in order to ensure a successful outcome.

AUTHOR CONTRIBUTIONS

BS: Drafting and writing of the manuscript.
MJO, RC, LGM: Critical review of the manuscript.
ACF: Writing and critical review of the manuscript.

PROTECTION OF HUMANS AND ANIMALS

The authors declare that the procedures were followed according to the regulations established by the Clinical Research and Ethics Committee and to the Helsinki Declaration of the World Medical Association updated in 2013.

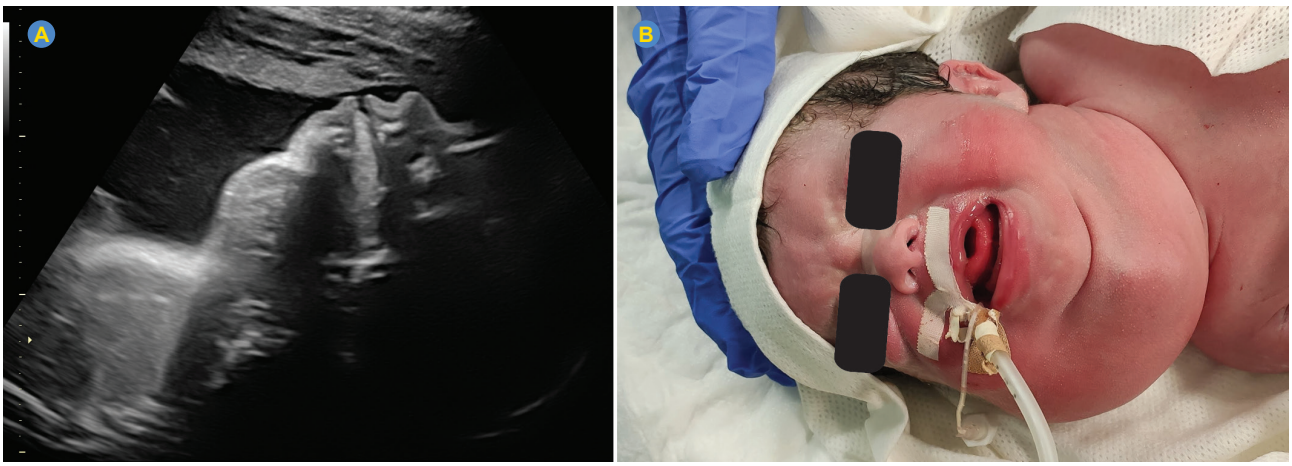

Figure 1 – Ultrasound imaging of a right-sided cervical mass first detected at gestational week 24 (A); Newborn elective intubation performed using the ex-utero intrapartum treatment procedure (B).

DATA CONFIDENTIALITY

The authors declare having followed the protocols in use at their working center regarding patients' data publication.

PATIENT CONSENT

Obtained.

COMPETING INTERESTS

The authors have declared that no competing interests exist.

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Reply to "A Case Report about the Management of Hereditary Angioedema with Normal Complement Levels during Pregnancy"

Resposta a "Caso Clínico sobre a Abordagem do Angioedema Hereditário com Complemento Normal Durante a Gravidez"

Keywords: Angioedemas, Hereditary/complications; Complement C1 Inactivator Proteins; Pregnancy Complications
Palavras-chave: Angioedema Hereditário/complicações; Complicações na Gravidez; Proteínas Inativadoras do Complemento 1

Dear Editor,

The recently published article by Pinto *et al* highlights a rare disorder called hereditary angioedema (HAE) diagnosed in a pregnant woman.[1] This disease is characterized by recurrent and unpredictable episodes of swelling of the upper airways and gastrointestinal tract.[1,2] The laryngeal oedema may cause fatal asphyxiation.[2]

The most common forms of HAE are types I and II, caused by deficiency and disfunction of C1 inhibitor (C1-INH), respectively.[2] A less prevalent form of HAE with normal C1 inhibitor (HAE-nC1-INH) has the distinctive characteristic of showing normal complement levels and affects mainly women, while men are often asymptomatic carriers.[3] In the largest Portuguese cohort that included 126 patients, HAE-nC1-INH was diagnosed in five patients (4%).[4]

The pathophysiology of HAE-nC1-INH is unknown, although evidence suggests the role of excess generation of bradykinin, causing vasodilation and vascular permeability.[5] Several mutations have been identified in families with HAE-nC1-INH, namely, in coagulation factor XII.[1]

Estrogens can interact with most of the steps of the cascade generating bradykinin.[5] Oral contraceptives, menstruation, pregnancy, and hormone replacement therapy represent important triggers.[5] However, pregnancy has shown a variable course in intensity and frequency of the attacks, even in the same patient, suggesting that the hormonal changes are not the only influencing factors.[3] Mechanical trauma due to uterine growth and fetal movements have also been proposed as aggravating factors.[3] However, a recent study revealed that attacks were more common during the first trimester (41.7%).[3]

Moreover, symptoms during pregnancy may be misdiagnosed, especially if the timing matches that of the onset