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CASE REPORT

Rare congenital tumor as an atypical cause of neonatal stridor: case report of a rare case of neonatal stridor and atypical evolution of late airway obstruction due to congenital oropharyngeal teratoma (epignathus)

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Teratoma,
Respiratory Insufficiency,
Child,
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Abstract

Background: Epignathus is the term used for congenital oropharyngeal teratoma, a rare condition however with great potential to cause fetal airway obstruction, attending with respiratory insufficiency and being able to cause perinatal death. Usual imaging exam in pregnancy follow-up such as ultrasonography can detect mass-type lesions occupying the head and neck region of the fetus, also commonly associated with polydramnium. The early diagnosis of Epignathus assists in planning management as delivery and adequate procedures to guarantee the fetal airway. **Case:** We report an atypical case of a 44-day-old baby girl, born without complications, but with parents reports of mild stridor from birth without investigation, progressed with severe respiratory insufficiency requiring orotracheal intubation in emergency service. At the time, a mass was detected on oropharynx obstructing airway. After the surgical exeresis of the mass, histopathological study revealed mature teratoma. **Conclusion:** Neonatal stridor, however mild, requires an adequate investigation of the probable etiology. In medical practice, cases of severe stridor associated with symptoms such as respiratory effort or difficulty feeding tend to be more valued. Epignathus is not a common cause of neonatal stridor, especially in cases of late manifestations.

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INTRODUCTION

Congenital teratomas are defined as true neoplasms with embryonic origin constituted by cells from the three germinative layers (endoderm, mesoderm and ectoderm)[1,2]. Epignathus is the term used for teratomas from the oropharynx[3]. Although rare they are relevant due to the high risk of obstruction on fetal airways. They may implying difficulty or even impossibility to do orotracheal intubation due to anatomical distortion, potentially leading to perinatal death[4]. The diagnosis of Epignathus usually occurs in intrauterine period during the standard prenatal assessments, since this pathology can cause structural changes in the fetal face in the early stages of pregnancy[5]. Magnetic Resonance Imaging (MRI) allows better accuracy to diagnosis because it correlates extension of the mass and involvement with nearby structures. MRI allows differential diagnosis and therapeutic decisions.

Neonatal stridor is an abnormal respiratory sound that signs possible anomaly in the airway causing laryngeal / tracheobronchial obstruction. It is one of the most common manifestation of respiratory discomfort in the newborn, and its early recognition and etiological investigation are fundamental, even when it is a discrete manifestation2. In this article we present a rare case in which a child with oropharingeal teratoma, who was not identified during prenatal and neonatal periods, despite institutional delivery, evolved atypically with insidious inspiratory stridor and late symptomatic manifestation of airway obstruction.

CASE REPORT

A 44-days-old baby girl was admitted at Pediatric Intensive Care Unit due to inspiratory stridor associated with severe respiratory failure. The baby was born without records of intercurrences, by cesarean delivery (due to mother desire), with 39 weeks pregnancy, 2760g of weigth, Apgar 8/9. The progenitor was 30 years old, G3P0A2, underwent regular prenatal follow-up with periodic ultrasonography without alterations. Morphological ultrasonography, performed around 20th week of gestation, did not reveal alterations in oral cavity or polydramnio. After brith, the baby followed to joint lodging, received breastfeeding and still at hospital she presented mild stridor without association with signs of respiratory effort. The child was discharged 2 days after delivery.

The parents identified increase of mild inspiratory stridor in the first week of life, associated with frequent pauses during breastfeeding, but without episodes of cyanosis. However there was progressive and gradual worsening of stridor becoming more noticeable after the 10th day of life (when intense crying started due to cramping). The newborn was consecutively evaluated on an outpatient basis by 6 paediatricians, however the complaint of stridor was not appreciated because of the benign another signs of respiratory distress. In the seventh medical evaluation, which occurred with 30-days-old, hypothesis of laryngomalacia was raised, but no complementary evaluation was performed through endoscopic examination (flexible videonasofibroscopy).

At 40-days-old while an episode of intense crying the baby had peripheral cyanosis that evolved with spontaneous resolution. The mother reported that during regurgitation after breastfeeding, she visualized a pinkish mass externalizing in the oropharynx. At 44-days old baby girl had successive episodes of cyanosis and short-term apnea during crying, with progressive worsening, evolving with significant respiratory discomfort. She was then taken to emergency room care at a pediatric hospital.

The baby was admitted at neonatal intensive care unit with respiratory insufficiency and required orotracheal intubation. During intubation it was visualized a solid tumor occupying posterior portion of the oropharynx (Figure 1). Initiated then investigation of congenital head and neck tumor. A MRI was performed to excluded Central Nervous System involvement and identified tumor with insertion into the lateral wall of rhino/oropharynx transition, with no extension to structures of the skull base. It was decided for an endoscopic surgical approach that was recorded in video. During the surgical procedure, a solid, pediculated, smooth surface lesion was then identified on the lateral wall of the hypopharynx. The tumor had approximately 5.0 cm (Figure 2), was photographed and sent for histopathological analysis that revealed mature teratoma.

In immediate postoperative period, the baby already presented a normal respiratory pattern, being discharged on the second day after the procedure. Regular outpatient follow-up was maintained and to date the child presents with no sequelae and no tumor recurrence.

DISCUSSION

Congenital teratoma is a complex tumor with embryonic origin composed of pluripotent cells of the germinative layers (ectoderm, mesoderm, endoderm). It presents irregular growth and can vary from cystic to solid lesions, even benign or malignant. Considered one of the most common types of neonatal tumors (25-30%)[1], congenital teratoma has an incidence of 1: 4000 live births[2,3]. About 80% occurs in the sacrococcygeal and gonadal regions, followed by the retroperitoneum and mediastinum regions. The head and neck region correspond to 1-9% of the cases of congenital teratoma, being more commonly affected the cervical region followed by the nasopharynx[7].

Teratoma originated in the oropharynx is called Epignatus, extremely rare, with an incidence of 1: 35,000 – 200,000



Figure 1. During the surgical procedure : a solid pinkish mass, with smooth surface.

live births, with female preponderance, in a proportion described in the literature of 3: 1[5].

Although rare it is of great importance due to the potential of fetal airway obstruction and perinatal death. According to Mutaz et al 2013[9], the term epignathus etymologically should be reserved for mandibular teratomas, however it is widely used in the literature to refer to teratomas arising in the oral cavity or pharynx. Teratoma epignatus is still considered of obscure origin, however there are theories regarding etiology that describe implantation or congenital inclusion of layers of germ cells; incomplete attempt to form twin Siamese; and, the most accepted theory, the desorganized growth of pluripotent cells of Rathke sac[8,9]. It may present alone, but according to the literature 6% is associated with midline abnormalities such as cleft palate, bifid tongue and duplication of the pituitary[7].

The definitive diagnosis is made through the histopathological analysis of the lesion surgically removed[10]. However, we found a description of prenatal diagnosis in the literature from the mid-1970s through the USG in regular prenatal follow-up[11].

Early diagnosis may occur accidentally 20-50% in the second trimester of gestation during ultrassonography evaluation[5]. Bi/three-dimensional USG can detect cervical or pharyngeal mass with solid-cystic components as early as 15 weeks of pregnancy and may externalize through the fetal mouth, causing a head hyperextension. In addition to 1/3



Figure 2. The tumor after endoscopic exeresis.

may be associated with polydramnio[11]. Research should be complemented with Magnetic ResonanceImage to identify mass characteristics, measure its extent and correlate with adjacent structures, including intracranial structures. Early diagnosis of teratoma epignathus allows better planning for management of pregnancy, decision of delivery and maintenance of the fetal airway. In the literature, we found protocols of surgical treatment considering EXIT (Ex-Utero Intrapartum Treatment Procedure) as an option for airway protection and attempt to avoid fetal death when early diagnosis of teratoma epignathus[5,7,12].

Stridor is one of the most common symptoms associated with neonatal respiratory distress, a problem present in about 7% of newborns. It is an acute respiratory sound, and may be inspiratory, expiratory or biphasic, caused by obstruction of the neonatal airways. It may present alone or associated with signs of respiratory effort and has among the most common causes laryngomalacia (70%), followed by vocal folds paralysis and congenital subglottic stenosis[2,6]. Appropriate management of neonatal stridor regardless of intensity (mild, moderate, and intense) includes anamnesis with a complete

history (description of the type of stridor, start time, triggering factors and improvement), a thorough physical examination and a complementary exam for immediate identification of the etiology. The endoscopic view of the airway through flexible laryngoscopy allows the anatomical and laryngeal dynamics to be valued in an accessible, reliable and sedation-free manner[6].

Head and neck masses are extremely uncommon causes of stridor in newborns and, when present, tend to occur early, with the majority still in the delivery room, which differs from the reported case, where it is initially mild and without association with other symptoms, hove late evolution of obstruction, not being performed early diagnosis.

Congenital teratoma of the oropharynx, called Epignatus, is rare and, when present, tends to manifest itself early with alterations in prenatal imaging tests or with obstruction of the airways soon after delivery.

Stridor in the neonatal period is a symptom that should be evaluated even when it is mild or not associated with signs suggestive of respiratory discomfort. Early investigation should be performed until adequate identification of the etiology, preferably with a pediatric otorhinolaryngological evaluation, allowing adequate management, a faster treatment and avoiding greater respiratory complications in infants, including respiratory discomfort and potential risk of death due to obstruction of the airways.

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Interest conflicts

Case report without any conflict of interest.

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