CONGENITAL TERATOMA IN A RARE CERVICAL LOCATION

Edin HUSARIĆ¹, Nešad HOTIĆ¹, Amir HALILBAŠIĆ¹, Emir RAHMANOVIĆ¹, Šekib UMIHANIĆ²

¹Department of pediatrics University Clinical Center Tuzla, Tuzla, Bosnia and Herzegovina, ²Department of otorhinolaringology University Clinical Center Tuzla, Tuzla Bosnia and Herzegovina

Corresponding author:
Edin Husarić
Department of pediatrics
University Clinical Center Tuzla
75000 Tuzla
Bosnia and Herzegovina
ehusaric@hotmail.com
Tel.: + 387 61 194 187
Fax.: + 387 35 250 474

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Objective – Teratomas with sacrococcygeal, mediastinal and gonadal locations are the most frequently occurring pediatric germ cell tumors. Cervicofacial and intracranial locations are rare. Approximately 20% of giant cervical teratoma causes airway compression. We report a congenital cervical teratoma with partial airway compression diagnosed post-natally in a preterm infant. Case report – A 27 year old mother delivered a male infant at 35 weeks gestation after her first uneventful pregnancy. The newborn had a notable anterior neck mass measuring 4x5 cm. The alpha fetoprotein level was elevated at 317.5 ng/ml and β-HCG was less than 1.2 mU/ml. An MRI scan showed a 24x53x27 mm prelaryngeal and paratracheal solid-cystic mass compressing and shifting the larynx and trachea to the right. The tumor was approached via a wide collar incision. It did not communicate with the oesophagus, trachea or thyroid gland and did not infiltrate the surrounding tissues. It was completely excised. Pathological examination revealed an immature teratoma. The recovery was uneventful. Conclusion – Teratoma in infancy may present in an unusual cervical location. Not only giant tumors may compress the airway.

Key words: Cervical teratoma • Congenital.

Introduction

The presentation of teratoma correlates with both age and anatomic site. The locations in infancy and early childhood are generally extragonadal, whereas those presenting in older children more commonly occur in the ovary and testis. Head and neck teratomas account for 5% of all neonatal teratomas (1). Cervical location is extremely rare. Most of these tumors are histologically benign. They are generally large and bulky, often measuring 5 to 12 cm in diameter. In approximately 20% of cases they cause significant airway and esophageal compression in the perinatal period and they are potentially fatal (2).

We report a case of a congenital cervical teratoma diagnosed post-natally, measuring 24x53x27 mm in a preterm newborn with partial airway compression.

Case report

A 27-year-old mother delivered a male infant at 35 weeks gestation by normal vaginal delivery after her first uneventful pregnancy, weighing 2200 g. The neonate presented...
Cervical teratomas are uncommon neoplasms, representing 3% of teratomas in childhood. Although these lesions are histologically benign, they may be large and may cause airway obstruction. Cervical teratomas are usually diagnosed at birth. In-utero ultrasound diagnosis is possible in early pregnancy (15-16 weeks). This may help planning early airway management and surgical intervention (3). Fetal MRI would delineate the anatomy more clearly if needed (4). Up to 50% of cervicofacial teratomas contain calcification and these are often seen on postnatal plain radiographs. A postnatal CT scan may also be used for evaluation (5).

Airway obstruction at birth is life threatening and associated with a high mortality rate, but prognosis is good with airway control and complete surgical excision. However, pressure injury of contiguous structures can limit resectability and adversely affect outcome (6). Airway management of the fetus with prenatally diagnosed tracheal obstruction can be performed through an operation on placental support (OOPS procedure): elective cesarean delivery of the fetal head and thorax under tocolysis, permitted endo-

Discussion

Cervical teratomas are uncommon neoplasms, representing 3% of teratomas in childhood. Although these lesions are histologically benign, they may be large and may cause airway obstruction. Cervical teratomas are usually diagnosed at birth. In-utero ultrasound diagnosis is possible in early pregnancy (15-16 weeks). This may help planning early airway management and surgical intervention (3). Fetal MRI would delineate the anatomy more clearly if needed (4). Up to 50% of cervicofacial teratomas contain calcification and these are often seen on postnatal plain radiographs. A postnatal CT scan may also be used for evaluation (5).

Airway obstruction at birth is life threatening and associated with a high mortality rate, but prognosis is good with airway control and complete surgical excision. However, pressure injury of contiguous structures can limit resectability and adversely affect outcome (6). Airway management of the fetus with prenatally diagnosed tracheal obstruction can be performed through an operation on placental support (OOPS procedure): elective cesarean delivery of the fetal head and thorax under tocolysis, permitted endo-
tracheal intubation, while the oxygen supply of the infant is maintained through the placenta (7).

Ex utero intrapartum treatment (EXIT) continues to be the optimal delivery strategy for patients with prenatally diagnosed giant cervical teratomas and potential airway obstruction at birth. A thorough evaluation of the prenatal images and an experienced multidisciplinary team are key factors for an effective approach to the obstructed fetal airway (8).

Despite the existence of poorly differentiated or undifferentiated tissue in the primary tumor, many infants remain free from recurrence following complete resection of cervical teratoma (1), however a successful outcome may not be obtained. All patients that undergo surgical excision of these tumours must be closely observed for post-operative respiratory distress, even in the absence of pre-operative symptoms (9). Biological markers: alpha fetoprotein and β-human chorionic gonadotropin play an important role in the diagnosis and management of giant cervical teratoma. However serum AFP levels are high during the first trimester, remain elevated at birth and decrease to adult values by 8 months of age, and still may be used for diagnosis and monitoring (10). Serum AFP levels should be checked 3-monthly in infancy and annually thereafter. Rising levels should alert clinicians to the possibility of tumor recurrence.

Imaging studies, twice a year for the first 3 years of life, are also recommended for surveillance. Because the thyroid and parathyroid glands may be removed or affected by tumor excision, the risk of temporary or permanent hypothyroidism must be considered (11).

Conclusion

We report a recent case of congenital cervical immature teratoma with partial airway compression. Total excision via a wide collar incision was performed. The aim of this case study is to report the authors’ experience in managing a case of congenital cervical teratoma to provide a structured approach and help in decision making, when prenatal diagnosis is not made.

Authors’ contributions: Conception and design: EH; Acquisition, analysis and interpretation of data: AH, ŠU; Drafting the manuscript: NH; Revising it critically for important intellectual content: ER, EH.

Conflict of interest: The authors declare that they have no conflict of interest.

References


