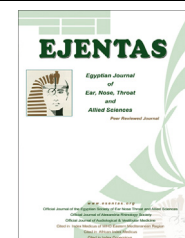




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CASE REPORT

Congenital teratoma of the neck: A case report and literature review



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Abstract Teratomas are unusual tumors derived from all 3 germ cell layers: endoderm, mesoderm, and ectoderm, with varying proportions. The cervical teratoma is a rare entity. Its prognosis mostly depends on the risk of neonatal respiratory distress, its extension and potential malignancy. Surgical management must be as complete as possible to avoid recurrences and malignant transformation. We present a case of a cervical immature teratoma in an infant with total excision and cure. No recurrence has been reported. The aim of our study is to review the diagnosis, management and outcomes of congenital cervical teratomas.

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1. Introduction

Teratoma is a term derived from a Greek word meaning monster. Classically it is composed of tissues from the three embryonic germ layers: ectoderm, mesoderm and endoderm.¹

Most commonly found in the sacrococcygeal region, these tumors are rare in the neck region. The cervical area is exceptionally affected.²

The symptoms are mostly attributed to the mass effect of these lesions, making the cervical tumors potentially lethal.³

The aim of our work by reporting a rare case of congenital teratoma is to produce a review about its etiology, pathology, diagnosis, prognosis and treatment.

2. Observation

A baby girl of a 20-year-old primigravida mother was found to have a voluminous cervical mass detected at birth. Although the patient remained asymptomatic except slight dyspnea.

On local examination, there was a solitary left neck mass which was irregular, of size about 6 cm having firm consistency with moderate mobility and no skin changes (Fig. 1).

MRI showed a 62*57*47 mm mass, with solid and cystic components with contralateral airway displacement and posterior displacement of the carotid. Additional CT scan revealed tumoral calcifications (Fig. 2).

At 20 days of age, the infant underwent surgical excision. Through a left cervical incision, the tumor surface was exposed

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Figure 1 Newborn with lateral neck lobular mass.

in the subdermal plane. It was a lobulated gray-white mass. A capsule was present, allowing a plane of dissection of the mass from the surrounding structures, which were displaced but not infiltrated (Fig. 3).

The infant was discharged after two days without any particular complaints.

Histopathology report came out as predominantly solid with areas of cystic changes. Multiple sections studied from tumor showed mature as well as immature elements derived from all 3 germ layers. Mature elements comprised of glands, mature cartilage, and neural tissue. Immature elements included neuroepithelial elements, neuroectodermal rosettes, and immature cartilage. Final diagnosis was immature cervical teratoma free of malignant elements (Fig. 4).

Uneventful recovery followed without recurrence.

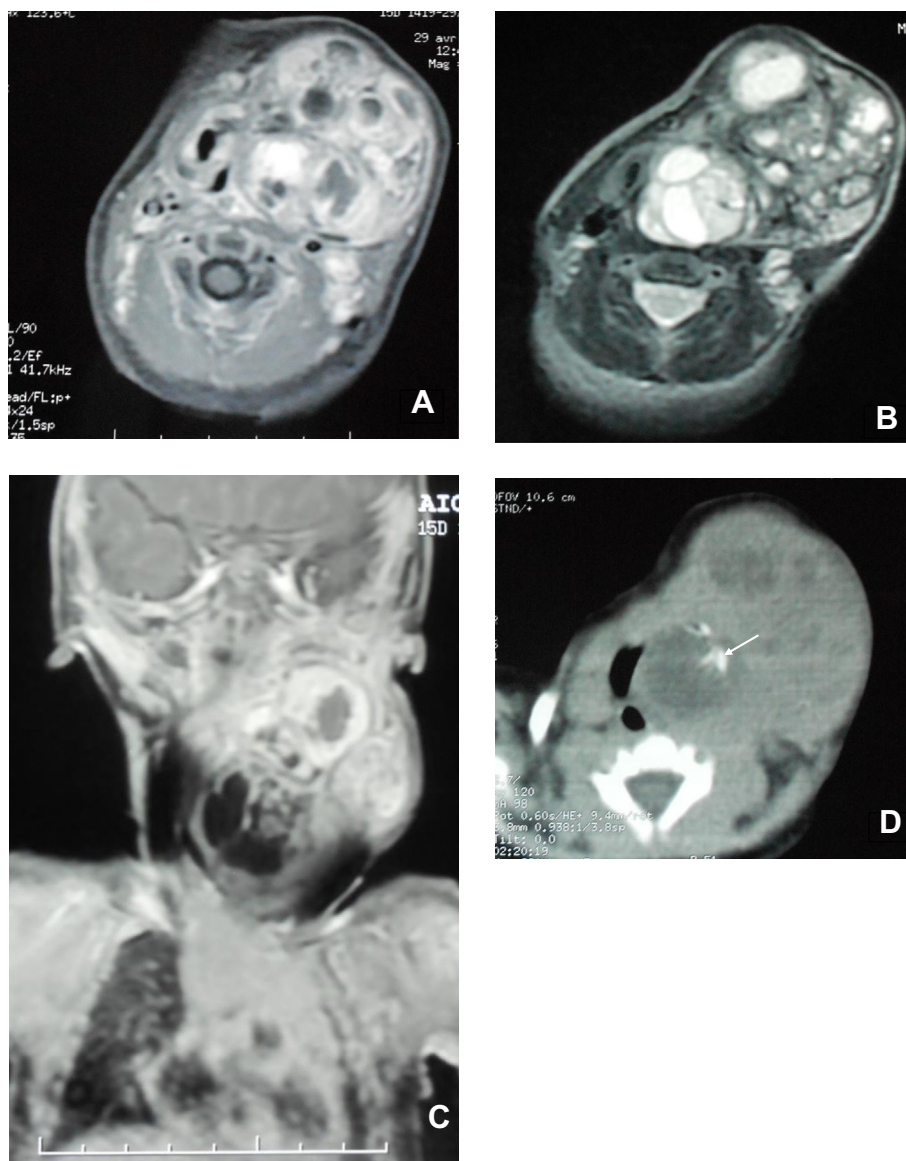


Figure 2 (A) Preoperative axial gadolinium enhanced MRI (B) axial T2 weighted magnetic resonance (B) coronal magnetic resonance showing heterogeneous mass (cystic and solid component) with contralateral airway displacement (D) cervical CT shows a non-homogeneous soft tissue mass with calcifications (arrow).

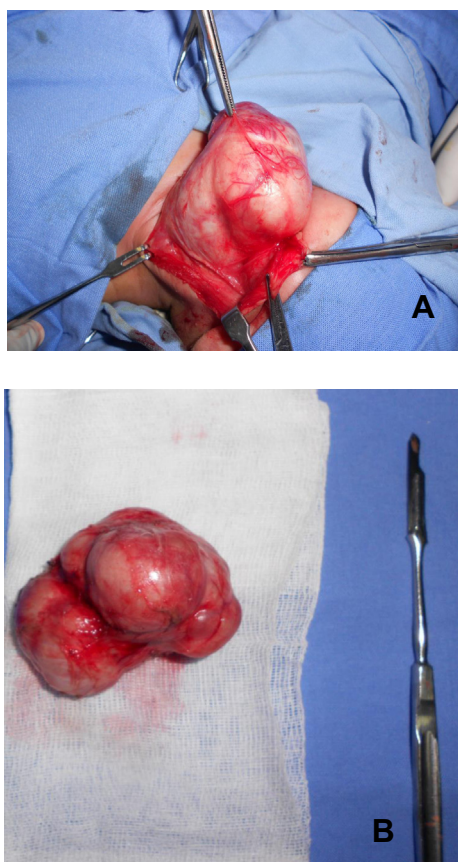


Figure 3 (A) Pictures during the surgical procedure show the typical appearance of the lobular soft tissue mass and (B) tumor totally removed.

3. Discussion

Teratomas are known to occur anywhere in the body the commonest location being sacral region. Cervical presentation constitutes 1.6–9.3% of all pediatric teratomas, roughly equating to 1 per 40,000 births. They predominate in females (3/4 of the cases).⁴

This tumor originates from aberrant germ-cells at the 4th or 5th week of gestation. These ectopic germ-cells undergo proliferation and differentiate in mature (mature teratoma) or fetal (immature teratoma) tissue, depending on the amount of immature elements, most often neuroepithelium.¹

Mature teratomas consist of well-differentiated tissues and may be pure or have a histologic component of a mixed germ cell neoplasm. Immaturity does not equate to malignancy, as was previously thought and less than 5% of congenital cervical teratomas are malignant.^{5,6}

Immature teratomas may be a component of a malignant germ cell tumor of mixed histology. The malignant component of mixed histology tumors that contain teratoma is usually of germ cell origin (yolk sac tumor, germinoma, embryonal carcinoma, or choriocarcinoma) and only rarely of somatic origin.¹

Teratomas have multifactorial etiology, chromosomal abnormalities have been reported are: trisomy 13, ring X-chromosome mosaicism with inactive ring X-chromosome, gonosomal pentasomy 49, gene mutations or abnormalities in early embryonic development.^{7,8}

Clinically, a cervical teratoma appears as a large single mass, although multiple lesions may occur. Airway obstruction is the main complication and is related to the size and site of the lesion occurring in 80% to 100% of cases.⁹ Peripartum mortality is often related to difficulty in establishing an airway after delivery.

The diagnosis can be made in utero on ultrasonography in pregnancy (15–16 weeks). The antenatal diagnosis of large congenital cervical teratomas allows for planned intervention by experienced personnel.¹⁰ Ultrasonography 3D and MRI may enhance the accuracy of the antenatal diagnosis and may help in the selection of newborns requiring early treatment.¹¹

If the mass is found after birth, ultrasonography, computed tomography (CT) and MRI are of primary importance for determining the extension, involvement of adjacent structures, and helps in planning surgery.¹²

Ultrasonography and CT scan show a cystic or heterogeneous mass with calcifications, solid and cystic components. MRI usually shows a circumscribed heterogeneous, cystic and solid uni or multiloculated tumor.¹¹

The differential diagnosis is done with a metastasis from thyroid carcinoma, cystic squamous cell carcinoma of cervical lymph node arising, follicular adenomas of the thyroid, lymphangiomas, and bronchial cysts.^{1,5}

Complete excision should be done in all cases. The procedure must not be delayed because these lesions, although benign, may grow quickly.

If prenatal diagnosis is made two procedures may be used: intrapartum treatment (EXIT) procedures and operation on placental support (OOPS). In the EXIT procedure, after a low transverse uterine incision, the head and at least one hand of the fetus are delivered. The rest of the body along with the umbilical cord and the placenta remain in utero. This allows examination of the airway.² In the OOPS procedure, the baby is completely delivered and the umbilical cord is clamped only after securing the airway with endotracheal intubation, using a laryngoscope or rigid bronchoscope or tracheostomy.¹⁰

With good presurgical planning and complete surgical excision there is no recurrence and few complications.⁹ A regular and long-term follow-up is necessary to detect early recurrences.¹³

The recurrence can occur in less than 10% of operated patients and can be treated with further surgery or chemotherapy.^{7,10}

Regarding the treatment of immature teratomas, Marina et al. found in a retrospective study of seventy-three children with extracranial immature teratomas that more than 85% of patients can be effectively treated with surgical resection alone and close observation.⁷

Follow-up is based on clinical examination and MRI, especially in case of incomplete excision.³ A trimestrial alpha-fetoprotein quantification is recommended by some authors.^{3,5}

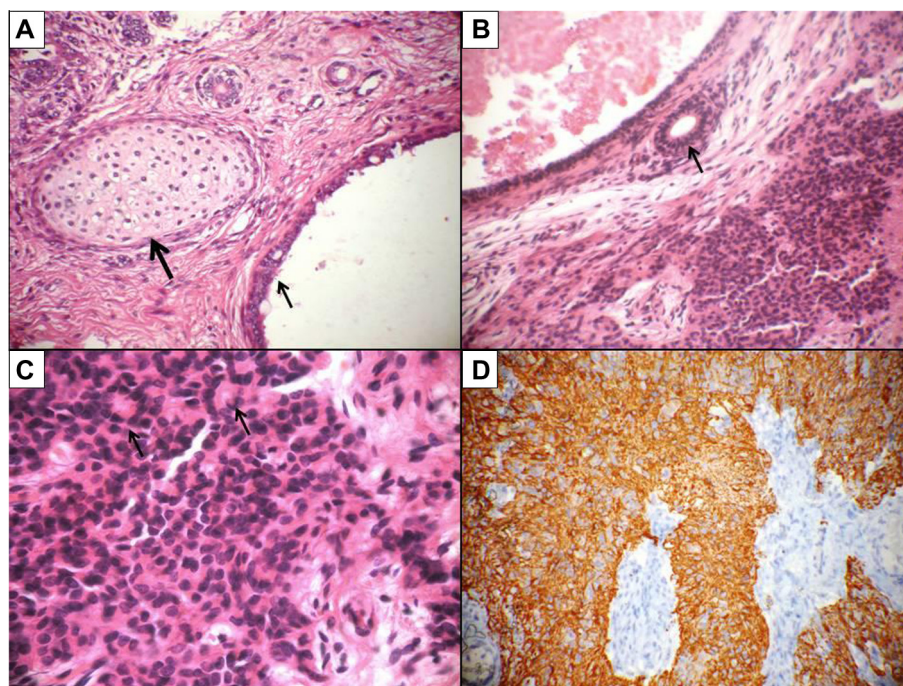


Figure 4 Immature teratoma: (A) Fetal cartilage (thin arrow) admixed with mature tissue (thick arrow) (Hematoxylin eosin $\times 200$). (B) Neuroectodermal tubule (thin arrow) lies in a background of glial tissue (Hematoxylin eosin $\times 200$). (C) Neuroectodermal rosettes are evident within the immature neuroectodermal tissue (thin arrow) (Hematoxylin eosin $\times 400$). (D) Diffuse immunostaining for glial fibrillary acid protein in the immature neuroepithelial component ($\times 400$).

4. Conclusion

Teratomas are rare tumors derived from all three germ cell layers affecting the neck in 3% of all cases. An early complete surgical approach to congenital cervical teratomas allows good results, with low rates of complication and recurrence.

Conflict of interest

No conflict of interest to declare. No ethical approval required.

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