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Congenital Cervical Teratoma: Case Report and Review of the Literature

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Abstract **Case Report**

Cervical teratomas are rare benign tumors in neonates, comprising 3% to 5% of all teratomas in children, A great majority of cervical teratomas are benign tumours. Its prognosis mostly depends on the risk of neonatal respiratory distress, its extension and potential malignancy. We present a case of a cervical immature teratoma in a new born with sub total excision and cure, with review about its etiology, diagnosis, prognosis and treatment.

Keywords: Teratoma, New Born, CT, MRI.

INTRODUCTION

Cervical teratomas are rare tumors in neonates. The incidence of teratomas of the head and neck ranges from 1:20,000– 1: 40,000 births and some sources cite a female predominance [1]. Most commonly found in the sacrococcygeal region, these tumors are rare in the neck region [2].

Diagnosis of a congenital cervical teratoma is possible during an early prenatal ultrasound evaluation, it is typically a large neck mass with solid and cystic components that causes hyperextension of the neck and is frequently associated with polyhydramnios [3]. Ultrasound and MRI evaluation are very useful to differentiate these tumours from other common congenital cervical masses, such as lymphatic malformations (cystic hygromas), through evaluation of their vascularity and soft tissue content [3].

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

CASE REPORT

A 40 days old new born was found to have a voluminous cervical mass detected at birth, Physical examination revealed a large ulcerated lateral lobulated neck mass extending from left side of the neck (Figure 1). The ultrasound showed a large cystized mass containing hyperechoic fatty areas (figure 2). CT scan of the neck showed large cervical mass extended to the scalp, containing dystrophic calcification and a large area of fat and a large area of mass effect on the airway (figure 3), MRI scan showed a solid and cystic mass in the neck associated with calcifications and involving multiple compartments with significant mass effect on the airway (figure 4). The infant benefited from a subtotal excision of the mass with anatomopathological examination which concluded to an immature teratoma.

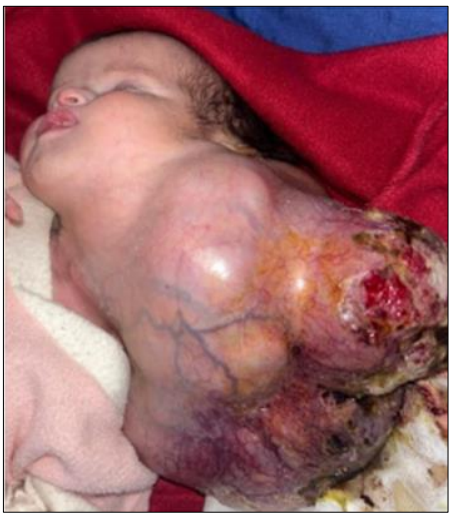

Figure 1: The sacral CT showed a well-limited lytic bone mass (a,b), centered on the S3-S4 and S5 vertebral bodies, which extends inside the vertebral canal and the endopelvic region, and comes into close contact with the low rectum (white arrow); without significant enhancement (d).

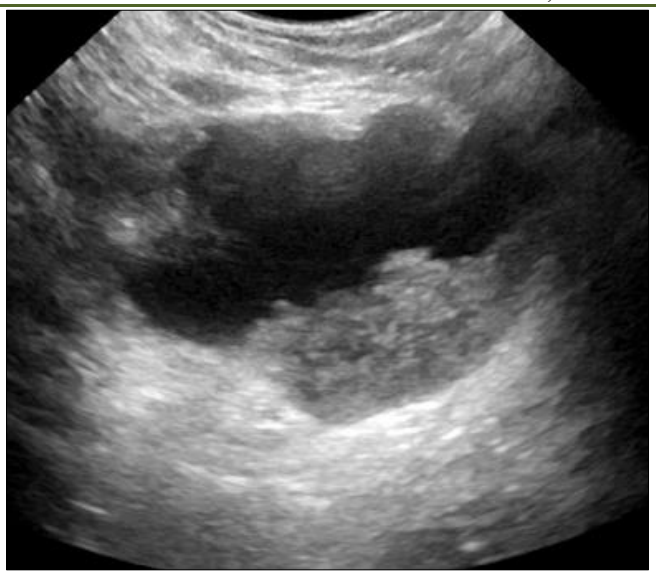

Figure 2: large cystized mass containing hyperechoic fatty areas

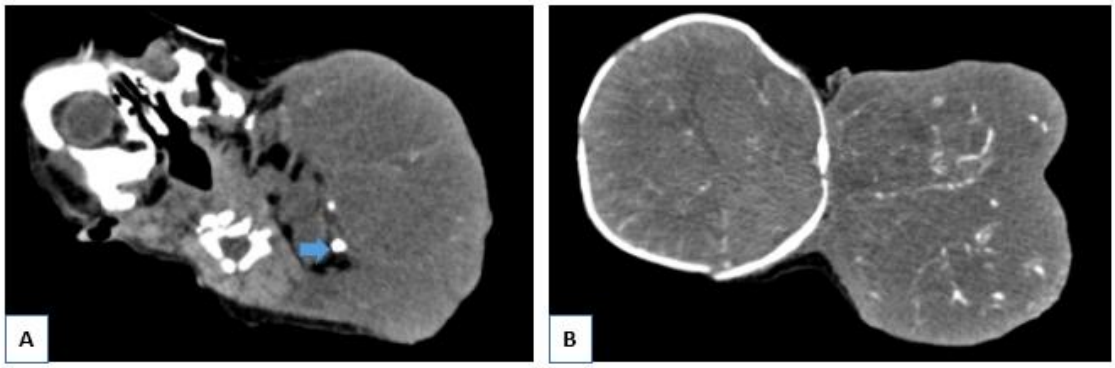

Figure 3 : CT scan of the neck without (A)and after injection of contrast product (B) showed dystrophic calcification (blue arrow) and a large area of fat with enhancement of the tissue portion. This masse is extended to the ipsilateral scalp.

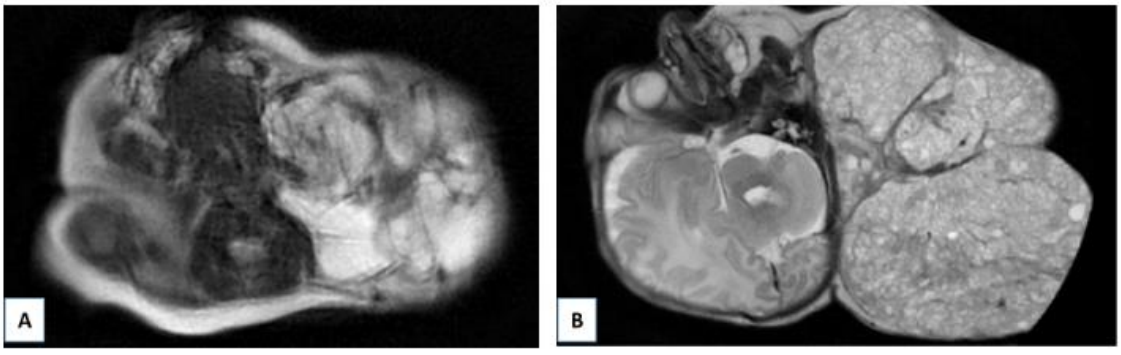

Figure 4 : MRI scan showed a solid and cystic mass in the neck extended to the scalp, associated with calcifications

DISCUSSION

Cervical teratomas are rare benign tumors in neonates, comprising 3% to 5% of all teratomas in children [4]. The vast majority of teratomas in newborns contain immature tissue components but are still considered benign [5]. The most commonly accepted theory suggests teratomas evolve from ectopic pluripotent cells that fail to migrate [6,7]. Teratomas are classified as mature or immature depending on the degree of cell differentiation. Most teratomas occur as an isolated lesion, but sometimes are part of a syndrome,

such as Klinefelter syndrome, trisomy 13, trisomy 21, or Beckwith-Wiedemann syndrome [8].

Cervical teratomas usually arise in the anterior and midline region of the neck, likely owing to involvement with the thyroid gland [9]. The tumor can displace or infiltrate structures through out the anterior neck, but most tumor is usually well encapsulated [9].

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 [10].

On ultrasound, cervical teratomas appear as large multiloculated [11], On computed tomography, teratomas appear as multiloculated cystic lesions with focal areas of fat attenuation and other areas of calcification multiseptated mass lesions with cystic and solid components and scattered areas of calcification. MRI usually shows a circumscribed heterogeneous, cystic and solid uni or multiloculated tumor [11].

The main differential diagnoses for foetal neck masses include lymphangiomas or cystic hygromas, cervical teratomas, haemangiomas, branchial cysts, cervical neuroblastomas, soft tissue sarcomas, and congenital cervical thyroid goitres. One differentiating feature of foetal neck masses is on their location, as teratomas are frequently anteriorly located and along the midline, whereas lymphangiomas or cystic hygromas, haemangiomas and branchial cleft cysts are more posterior and lateral in location [12].

Surgical resection is the standard of care for cervical teratomas. However, the top priority in a neonate with a cervical teratoma is airway management [7]; Patients with cervical teratomas often require an ex utero intrapartum procedure (EXIT) to secure the airway at delivery. Prenatal MRI imaging can assist with evaluating the extent of airway obstruction and determining the need for an EXIT procedure [13]. 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 [6]. Another procedure may be used: operation on placental support (OOPS); 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 [14].

With good presurgical planning and complete surgical excision there is no recurrence and few complications [15]. The recurrence can occur in less than 10% of operated patients and can be treated with further surgery or chemotherapy [16,17]. Postoperative surveillance should include assessing AFP levels at monthly intervals in infancy and yearly thereafter, up to 3 years of life [18,19]. MRI scanning twice a year for the first 3 years of life also has been suggested [20].

CONCLUSION

Teratomas are unusual tumors derived from all 3 germs cell layers: endoderm, mesoderm, and ectoderm, with varying proportions. The cervical teratoma is a rare entity. Various imaging modalities play a significant role in the early diagnosis. An early complete surgical approach to congenital cervical teratomas allows good results, with low rates of complication and recurrence.

Abbreviations:
CT: computed tomodensitometry

MRI: magnetic resonance imaging

AFP: alpha foeto-protrein

Conflict of Interest: All authors state that they have no conflicts of interest.

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