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Head and neck teratoma: From diagnosis to treatment

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ABSTRACT

Introduction: Head and neck 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. The authors present a retrospective analysis of 6 cervicofacial teratomas and a review of the literature. The aim of the study was to analyse prenatal, neonatal and postnatal management of teratoma.

Materials and methods: Charts of children presenting with a head and neck teratoma, managed by our maxillofacial and plastic surgery unit, were analysed and antenatal, clinical, biological, radiological and pathological characteristics were collected. Surgical treatment, recurrences and surgical outcomes were analysed.

Results: Six patients were included: 2 with a cervical teratoma, 2 with a facial teratoma and 2 with intraoral teratomas. In 2 cases, the lesions were diagnosed antenatally and both patients required neonatal resuscitation. All the patients underwent early surgery, and 3 with complete excisions. All patients with an initial incomplete excision eventually presented a recurrence and therefore second look surgery. No malignant transformation was noted.

Conclusion: Early prenatal diagnosis is crucial to neonatal care. Early surgery and meticulous follow-up are critical in the long-term favourable outcome.

1. Introduction

Teratoma is a term derived from a Greek word: τερας (meaning monster). Classically it is composed of tissues from the three embryonic germ layers: ectoderm, mesoderm and endoderm (Silberman and Meldelson, 1960).

Ectoderm produces skin, teeth, nails, ependyma, choroid plexus, neuroglia and neurons. Mesoderm produces adipose tissue, bone, cartilage and muscle, whereas entoderm generates respiratory and intestinal epithelium, exocrine glands, and solid organs (pancreas, liver, kidney, lung, thyroid). Teratoma 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 foetal (immature teratoma) tissue (Silberman and Meldelson, 1960; Holt et al., 1979).

Epignathus actually refers to teratomas of the oropharyngeal cavity in neonates. These lesions are benign and represent the highest differentiated teratoma in the head and neck region. The ultimate form is defined as representing a foetus in foetus (Kumar and Sharma, 2008).

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Table 1
Antenatal and neonatal data of the included patient.

Patient	Gender	Antenatal diagnosis	Obstetrical history	Prematurity	Delivery	Respiratory distress	β-HCG (UI/l)	α-fetoprotein (ng/l)
#1	Girl	No	None	No	Vaginal	No	Normal	Normal
#2	Boy	No	None	No	Vaginal	No	Normal	Normal
#3	Girl	No	Preeclampsia	No	Vaginal	No	Normal	220,000, fall down after surgery
#4	Boy	Yes	Polyhydramnios	Yes, 33 weeks	C-section	Yes	Normal	Normal
# 5	Boy	Yes	Polyhydramnios	No	C-section	Yes	Normal	Normal
#6	Girl	Yes	None	No	C-section	Yes	21	Normal

C-section: cesarean section.

Congenital teratomas mostly occur in the sacrococcygeal area (60%). The head and neck localization is rare (Iatrou et al., 2013) and represents 5% of all congenital teratomas. Its incidence varies from 2.5 to 5/100,000 live birth. There is no gender predilection (Mahour et al., 1974).

These tumours are often large (Silberman and Meldelson, 1960). Their severity depends on neonatal respiratory distress and malignancy. There is no pathognomonic clinical or radiological characteristic (Koeller et al., 1999; Lloyd and McHugh, 2010), however the presence of calcifications within a heterogeneous mass is strongly suggestive of the diagnosis. Complete surgical excision is considered as the Gold Standard (Anderson and David, 2003; Benouaiche et al., 2007). From the antenatal diagnosis to surgical excision, a multidisciplinary approach is often needed.

Teratoma treatment is well known and accepted. However antenatal and neonatal diagnosis remains sometimes difficult. The aim of this study was to describe antenatal, obstetrical, clinical and radiological characteristics, their management and follow-up for teratoma.

2. Material and methods

We conducted a retrospective study from 1989 to 2013 including neonates presenting with cervical and facial teratoma managed in our maxillofacial and plastic surgery department. All charts were analysed and the following data were collected: antenatal diagnosis, sex, obstetrical history, term of birth, gestational age, respiratory distress, clinical description of the tumours, serum level of alpha-fetoprotein (AFP) and beta-human chorionic gonadotrophin (beta-HCG), and post-operative outcome. All radiological and pathological examinations were reviewed. Surgical treatment, recurrences and surgical outcomes were analysed.

3. Results

Six patients matched the criteria and were included, 3 girls and 3 boys. Antenatal ultrasonography (US) showed the tumour in 3 cases, one at 33 weeks and two at 22 weeks.

Patient #1 presented an intraoral foetal mass and polyhydramnios responsible of prematurity (33 weeks).

Patient #2 presented with a protruding intraoral heterogeneous mass at week 24 and polyhydramnios at 33 weeks. MRI performed at 30 weeks showed a 7.8 cm-intraoral heterogeneous mass extending down to the larynx and externalized through the right nasal fossa.

Patient #3 presented with a cystic lesion of the cheek on US. It was initially diagnosed as a lymphangioma but MRI showed a heterogeneous tumour and teratoma was suspected. All children diagnosed prenatally were delivered by C-section and presented with neonatal respiratory distress, needing nasal intubation (Table 1). For the three remaining patients, delivery occurred normally without respiratory problems.

Alpha-fetoprotein and beta-HCG blood levels are shown in Table 1.

MRI was performed shortly after birth in all patients (Fig. 1). Tumour diameter varied from 5 cm to 13.7 cm (mean 8.7 cm). Tumours showed a cystic component in 5 cases, and a solid component in 4 cases. None displayed calcifications (Table 2). In patient #1, complementary examination showed a malformation of the aortic arch.

All patients were treated surgically (Figs. 2 and 3). Total excision was performed in 3 cases, without recurrences. A second look surgery was eventually required in all subtotal primary excision. Surgical outcomes were considered as good in 4 patients (#1, #4, #5, #6). Patient #2 had an aesthetic average result with a secondary need of fat graft and patient #3 had mandibular growth impairment with the need of osteogenesis distraction (Table 3). Pathological examination revealed tridermal mature teratoma (Fig. 4) in 5 cases and bidermal mature teratoma in one case.

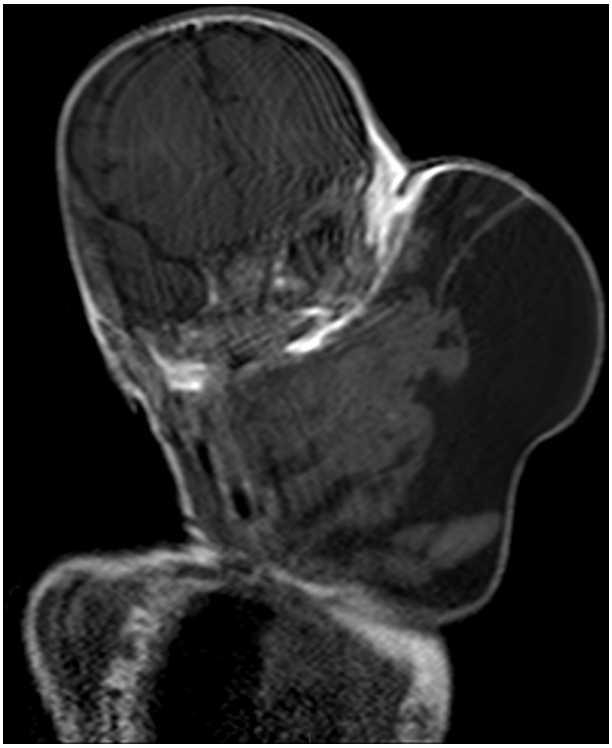

Fig. 1. MRI: Cervicofacial teratoma showing cystic and solid component, and anatomic extension (T1, with gadolinium, coronal slice).