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Case report

A large teratoma of the hard palate: a case report

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Abstract

Congenital teratoma is a rare malformation, and few papers have been published about it. We present a large teratoma that arose from the hard palate in a neonate. The obstructive mass caused maternal polyhydramnios and was identified prenatally by ultrasonography. The mother went into labour at 35 week's gestation at home. The child was in respiratory distress as a result of airway obstruction, and a tracheostomy was done when she was 4 hours old. She also had major cardiac abnormalities. The palatal mass was removed successfully at 4 weeks of age. The typical components of a teratoma were identified including immature neural glial tissue.

Keywords: Teratoma; Epipalatus; Palate

Introduction

Teratoma was defined by Weaver et al. as a tumour consisting of multiple tissues that are not indigenous to their site of origin.[1] They are true neoplasms of presumed primordial germ-cell derivation, which typically consist of tissues from all three embryonic germ layers: ectoderm, mesoderm, and endoderm.

A teratoma may develop in almost any area of the body, but usually in median sites.[2] They have a reported incidence of 1:4000 live births with around 2%–9% of these in the head and neck.[1]The most common sites are the sacrococcyx, anterior mediastinum, testicle, ovary, or retroperitoneum.[1,2]

There is confusion over the derivation of the word teratoma (from the Greek 'teras' = monster). Some authors incorrectly call lesions that do not have derivatives of layers of all three germ cells "teratoma". "Epignathus" is commonly used to describe a congenital teratoma in the oropharyngeal region, but should be reserved for tumours that arise from the jaw, specifically the alveolus of the mandible.[3] Oropharyngeal teratomas should be subdivided according to their site.[3] However, regardless of their anatomical site, "epignathus" is the most used term for an oropharyngeal teratoma.

We describe a large teratoma in a neonate who had complex congenital heart defects.

Case report

A white female infant was born prematurely after 35 weeks' gestation in August 2004. The mother was seen prenatally at 19 weeks. Incidental ultrasonography showed that the fetus had a univentricular heart and a large mass protruding from its mouth that arose from the hard palate. Polyhydramnios during pregnancy suggested impaired fetal swallowing.

Unfortunately, the mother went into labour at home and gave birth by spontaneous vaginal delivery. The child was in respiratory distress and was initially supported by bag and mask ventilation. She was immediately transferred to hospital where a nasopharyngeal tube was inserted. From there she was transferred to the tertiary referral centre and a tracheostomy carried out when she was 4 hours old.

Examination showed an obstructive polypoid mass that included areas covered in skin and hairs, and cystic, ulcerated lobes (Fig. 1).

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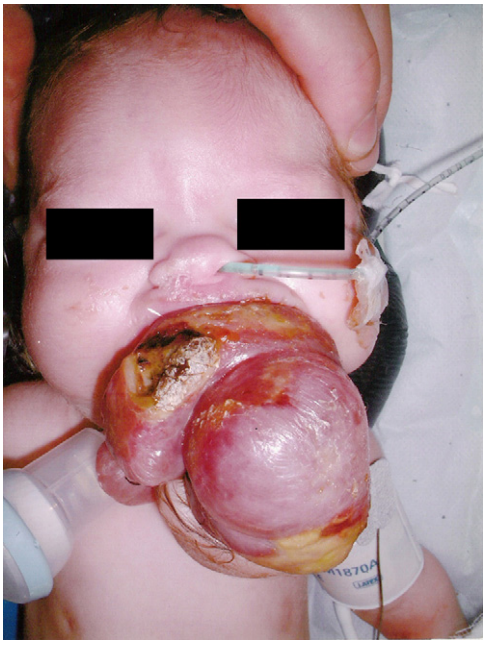

Fig. 1. Large teratoma of the hard palate with a tracheostomy in place.

Computed tomography showed a complex mass protruding from the oral cavity (Fig. 2). It was viewed three-dimensionally and seen to arise from the palate with a thin element extending into the nasal septum. It consisted of cystic, fat, bony, and neural elements, with defined teeth within the bone. There was a tiny defect in the floor of the base of the skull, though there was no evidence of intracranial involvement. A magnetic resonance scan confirmed that there was no intracranial extension. Genetic testing failed to show a syndrome, and amniotic fluid was of normal karyotype. On auscultation there was a 4/6 ejection systolic murmur. A univentricular heart and persistent ductus arteriosus had been diagnosed antenatally and was confirmed postnatally as a double-outlet left ventricle with transposition of the great arteries and tricuspid atresia. Evidence of cardiac failure was seen by day 9, but cardiac surgery was delayed until after treatment of the craniofacial anomaly because of risk of infection and poor weight gain.

The tumour was excised at 4 weeks (Fig. 3). Cystic areas were decompressed and the pedicle (roughly 2.5 cm in diameter) was sectioned to deliver the mass. Nasal integrity was confirmed. The temporomandibular joints moved well although the micrognathic mandible was depressed posteriorly. After the tumour had been removed, a cleft soft palate became apparent. Postoperatively the palatal wound epithelialised well. She had a gastrostomy at the age of 8 weeks.

By 7–8 weeks the neonate required continuous positive airway pressure, and later, ventilation. At 12 weeks her pulmonary artery was banded and the persistent ductus arteriosus ligated with pronounced improvements in her symptoms of heart failure.

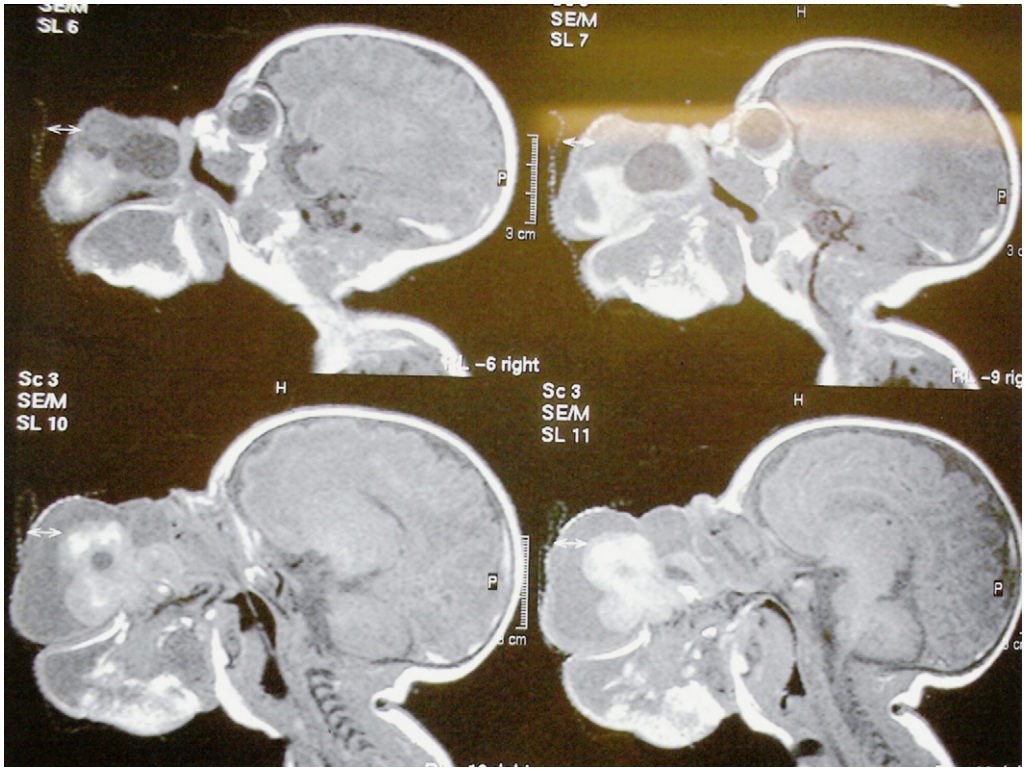

Fig. 2. Sagittal CT view: Pedicled mass from the hard palate, with areas of high and low attenuation. No intracranial extension is apparent.

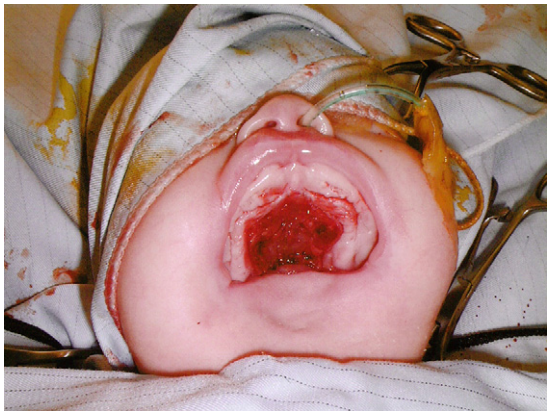

Fig. 3. Palatal defect after the teratoma has been removed.

Histopathological examination

The mass was not encapsulated, and there was a mixture of well-defined tissues derived from the ectoderm, mesoderm, and endoderm. Skin, fat, bone, cartilage, squamous and respiratory epithelium, lymphoid tissue, salivary gland type tissue, and neuroglial tissue with choroid plexus and ependyma, were all present within the mass. There were elements of focal necrosis, but only one small focus of undifferentiated neuroblasts. The tissues overall were immature but consistent with the degree of maturity expected in a neonate. It was diagnosed as teratoma epipalatus. We were uncertain about whether this should be regarded as an immature teratoma, or as a mature teratoma with immature elements "appropriate to age".

Discussion

Teratomas show progressive uncoordinated growth and are true neoplasms.[4] Neonatal teratomas are usually benign, there being a higher incidence of malignancy in teratomas in adults. Calcification is four times more common in benign than malignant teratomas, so may be an important diagnostic indicator. The presence of primitive neural tissue also suggests malignancy. The presence of a teratoma should alert the clinician to the possibility of other germ cell tumours, the most worrying of which in neonates is the yolk sac tumour (named by Huntington and Bullock).[4] Half of malignant teratomas contain this lesion, which is an embryonal carcinoma previously named endodermal sinus tumour. There was no yolk sac tumour within our teratoma.

There are at least three hypotheses about the aetiology of these lesions.[5] It has been suggested that the tissues of a teratoma derive from totipotential cells sequestered during embryogenesis. Another theory is that germ cells may give rise to teratomas by parthenogenetic development. Finally, a teratoma may originate from incomplete formation of Siamese twins. In our case, incomplete division of blastula may provide enough cells for the production of a teratoma but insufficient for a complete twin. This theory accounts for the formation of the most highly differentiated teratoma known as 'fetus in fetu', which has a reported incidence of 1 per 500, 000 births.[6]

There is little knowledge about predisposing factors because of the rarity of the tumour. An incidence of between 1: 35, 000 and 1: 200, 000 live births has been suggested, but it is generally thought that there is no ethnic or geographical pattern to incidence. Most reports do not suggest sex as a risk factor, but some described a slight female predominance,[4] and Carney et al. found malignant teratoma to be more common in men in a ratio of 5:4.[4]

The teratoma may be diagnosed antenatally on ultrasound or magnetic resonance, which permits early multidisciplinary management. The tumour presents as a cystic, and solid lesion that originates from the palate. This diagnosis is confirmed by polyhydramnios,[8] which is thought to be a consequence of impaired fetal swallowing and it has been proposed that cardiac decompensation caused by circulation within a large vascular teratoma may also contribute. Some authors, including Andze et al., have reported increased α-fetoprotein concentrations prenatally, which are suspicious of a teratoma.[7]

The differential diagnosis is limited, but includes hamartoma, dermoid cyst, and heterotopic gastrointestinal cyst.[8] Teeth within the mass confirm teratoma.

Antenatal diagnosis allows a controlled environment to be planned for the birth with multidisciplinary care. In our case labour began at home, and respiratory obstruction could have been life-threatening had the nasal airway not been patent, and ventilation with bag and mask not begun.

This teratoma may present as an obstructive mass causing respiratory embarrassment and an immediate threat to life, which demands immediate establishment of an airway, often with tracheostomy. It may also present as a small pedunculated tumour with no functional obstruction. Resection is the treatment of choice, as there may be a small chance of malignant transformation the longer they are left[4,9] The neonate's prognosis worsens as the size of the tumour increases. The ultimate prognosis of lesions with intracranial involvement is poor and operation is inappropriate.[10]

Benign teratomas may recur after excision. Carney et al.[4] discussed three cases of mature saccrococcygeal teratomas that recurred in children. This does not necessarily imply malignancy, although the clinician should maintain follow-up.

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