

A Rare Case of Congenital Teratoma Arising from Hard Palate in a New Born

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Abstract

Congenital teratoma is a rare malformation, and few papers have been published about it. We present a case of large teratoma arising from the hard palate in a neonate. The obstructive mass caused maternal polyhydramnios. The child was in respiratory distress as a result of airway obstruction, and endotracheal intubation was done immediately after birth. The palatal mass was removed successfully on 1st day of life. The typical components of a teratoma were identified including mature neural glial tissue.

Keywords: Teratomas; Hard palate; Germ layer; Intact survival with emergency medical & surgical management; Polyhydramnios; Teeth and hairs

Case Summary

A female neonate was born at 39 weeks gestation on 15 Feb, 2012 to a 24 yrs old primigravida. The mother was a booked case but antenatally no congenital abnormality was detected. Polyhydramnios during pregnancy suggested impaired fetal swallowing. Mother was admitted in this hospital at term. Taken for emergency LSCS because of non-progress of labour. The neonate was born by LSCS with a large mass was coming through the mouth and neonate had developed respiratory distress. Neonate was immediately intubated. The intubation was difficult and bag and tube ventilation was given. And neonate was immediately transferred to NICU on T-piece ventilation. Examination showed an obstructive polypoid mass that included areas covered in skin and hairs, and cystic, ulcerated lobes (Figure 1).



Figure 1: Large teratoma of the hard palate with an endotracheal tube in place.

Computed tomography showed a complex mass protruding from the oral cavity. 3D analysis showed arises from the palate. It consisted of cystic, fat, bony, and neural elements, with defined teeth within the bone. Though there was no evidence of intracranial involvement. The

tumour was cauterised from its base and deliver the mass excised on 1st day of life (Figure 2). Nasal integrity was confirmed. The temporo-mandibular joints moved well although. After the tumour had been removed no cleft soft palate. Postoperatively the palatal wound epithelialised well.

By 2nd day of life neonate was extubated and we have applied nasopharyngeal airway for 07 days followed by hood and then room air. Feed by naso gastric tube for 14 days and then paladai. USG abdomen-normal scan, ECHO- no congenital heart disease. 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. There were elements of focal necrosis. The tissues overall were mature with the degree of maturity expected in a neonate. It was diagnosed as teratoma epipalatus (mature teratoma).



Figure 2: After removal of teratoma



Figure 3: Large teratoma of the hard palate after removal.

Discussion

The term teratoma refers to a group of complex tumour having various cellular or organoid components reminiscent of normal derivatives from more than one germ layer. A teratoma may develop in almost any area of the body, but usually in median sites [1]. They have a reported incidence of 1 in 20000-40000 live births [2], 4 times more common in girls than boys, with around 2%-9% of these in the head and neck [2]. The most common sites are the sacrococcyx, anterior mediastinum, testicle, ovary, or retroperitoneum [2]. There is confusion over the derivation of the word teratoma (from the Greek 'teras'=monster [3]. 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. Oropharyngeal teratomas should be subdivided according to their site. However, regardless of their anatomical site, "epignathus" is the most used term for an oropharyngeal teratoma. 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. The presence of primitive neural tissue also suggests malignancy. Calcification is four times more common in benign than malignant teratomas, so this may be an important diagnostic indicator. 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] (Figure 3).

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 partheno-genetic 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].

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, 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 [7]. Some authors, including Andze et al., have reported increased fetoprotein concentrations prenatally, which are suspicious of a teratoma [8].

The differential diagnosis is limited, but includes hamartoma, dermoid cyst, and heterotopic gastrointestinal cyst [8]. Teeth within the mass confirm teratoma. 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 sacrococcygeal teratomas that recurred in children. This does not necessarily imply malignancy, although the clinician should maintain follow up.

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