Congenital Nasopharyngeal Teratoma with a Cleft Palate: Case Report and a 7 Year Follow Up

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Abstract Teratomas are benign tumors containing cells from ectodermal, mesodermal and endodermal layers. They occur in about 1 in every 4,000 births and most commonly in the sacrococcygeal region, followed by the ovaries. Congenital epignathus teratomas are rare embryological neoplasms localised in the region of head and neck. An epignathus is found in approximately 1:35,000–1:200,000 live births. This accounts for 2–9% of all teratomas. Size and location of the neoplasm in the oronasopharynx is variable. Teratomas are partly undiagnosed at the time of birth. They may exist with an intracranial extension or as small polyps. Large epignathi can lead to difficult management during and after birth. We present a case of a female infant with a combination of nasopharyngeal teratoma and a cleft palate with successful multidisciplinary management via a staged surgical approach. The epignathus presented as a huge mass extending out of the mouth of the infant girl. On the day of birth debulking of the extraoral portion of the tumor, followed by intraoral extirpation and cleft palate repair at a later stage, was performed. The results of the histopathological examination confirmed diagnosis of a congenital teratoma.

Keywords Epignathus · Nasopharyngeal teratoma · Cleft palate

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Introduction

Tumors of the oral cavity and nasopharynx in the newborn are not common. An epignathus refers to a teratoma of the oropharyngeal region composed of cells from the ectodermal, mesodermal, and endodermal layers [1]. These tumors may vary in structure and degree of differentiation. Epignathi that arise from the palate or pharynx and protrude from the mouth result in life-threatening airway obstruction and usually cause asphyxiation shortly after birth [2, 3]. Patients with relatively small or simple tumors have survived, however, [4, 5]. Polyhydramnios occurs in these cases and has been attributed to difficulties in swallowing as a result of the large intraoral and extraoral mass [6, 7]. We present a case of a massive nasopharyngeal teratoma in a newborn girl with a cleft palate. Debulking of the extraoral mass, followed by complete intraoral resection, was performed. Cleft palate repair was planned at 1 year of age.

Case Report

A baby girl was born vaginally as a full term spontaneous delivery, with birth weight of around 3.2 kg. The infant had a partly soft and partly hard, pedunculated tumour that measured 20 cm × 16 cm × 8 cm (Fig. 1) attached to the nasopharynx dividing the soft palate with associated cleft palate. The obstructing mass caused immediate neonatal respiratory distress, so the extraoral part, which weighed 430 g, was excised by ligating the stalk of the tumour near the base of the skull by a transoral approach under general anesthesia using oral intubation. The excised specimen had the following dimensions: 20 cm × 16 cm × 7 cm (Fig. 2a). It was lobulated and had rudimentary skin and

Fig. 1 Clinical photograph taken at birth shows a multilobulated, pedunculated reddish mass protruding from the oral cavity

hair as well as calcified bones and teeth with cystic and solid areas with the density of fat. Histopathological examination confirmed the diagnosis of benign teratoma with differentiation into ectodermal, mesodermal, and endodermal tissue (Fig. 2b). After uneventful follow up for 1 year the palatal defect was repaired (Fig. 3a, b). The patient had regular follow-ups and responded well with no recurrence detected even after 7 years. Postoperatively, the patient had macrostomia and maintained an open mouth posture (Fig. 4). Macrostomia and anterior open bite corrections were planned at a later stage.

Discussion

The term teratoma, derived from the Greek word “teraton” (meaning monster), was initially used by Virchow in 1863 [8]. These tumors have been considered parasites with independent growth potential to disrupt adjacent normal tissues with increase in size [3]. Classification may be made into four groups: (1) dermoid cyst (epithelial lined with skin elements, composed of ectodermal and mesodermal cells), (2) teratoid cyst (all three germ layers but poorly differentiated), (3) teratoma (three germ layers differentiated into specific tissues or organs), and (4) epignathi (oral tumors with developmental fetal organs and limbs) [9]. Another pathological variation of epignathi is fetus-in-fetu, which may be considered incomplete twinning of monozygotic twins at a primitive stage when axial development begins [10]. Fetus-in-fetu has vertebral development, internal organs, and formation of limbs. Generally, an epignathus is a highly developed oral tumor; however, the term specifically refers to a jaw or alveolar point of origin. With regard to the nomenclature, it has been suggested that head and neck teratomas with other sites of origin deserve a specific designation: ‘epipalatus’

Fig. 2 **a** Gross specimen weighing around 430 g with the following dimensions: 20 cm × 16 cm × 7 cm. **b** Photomicrograph showing mature teratoma cells with squamous epithelium and keratinous debris, adipose tissue with duct-like and glandular elements

when of palatal origin, ‘episphenoid’ when of sphenoidal origin, and ‘epicranium’ when of cranial origin [3]. In the case presented, the patient had a tumor arising from the base of the sphenoid. It was composed of fibrous tissue, cartilage, bone, neuroepithelium, hepatic tissue, and lung tissue. The pathogenesis of epignathi is uncertain. One theory suggests that failure of fusion of midline structures during embryogenesis leads to a midline nasopharyngeal teratoma and cleft palate [11, 12]. Another theory suggests that pluripotential primordial germ cells are misplaced and grow unchecked without the normal organizational influences of surrounding cells and factors [13, 14]. Krafka [15] explains that the axial orientation of the epignathi is caused

Fig. 3 **a** Clinical photograph showing cleft palate. **b** Clinical photograph showing closure of cleft palate

Fig. 4 Clinical photograph of the patient showing macrostomia and anterior open bite

by secondary embryonic axes at foreign sites. Dohman and Sjoval [16] advocated a multistage treatment of an epignathus, including an immediate debulking on the first day, followed by removal of residual tumor in 5 weeks and closure of the cleft defect at 14 months of age. In summary, successful staged resection of a massive epignathus, nasopharyngeal teratoma was achieved in a newborn. In the literature, epignathi and teratomas of the skull base have been considered incompatible with life [15]. With involvement of the skull base; lesions may extend both extracranially and intracranially in an hourglass formation. Newborns with an epignathus that involves the base of the sphenoid should undergo imaging (CT scan or MRI). Resection of smaller pedunculated epignathi has been successfully performed. Larger epignathi extending extraorally are problematic for the neonatal airway and have

led to respiratory distress and death [16]. The main differential diagnosis is encephalocele or meningoencephalocele, and it is important to know before deciding on the operation. In 6% of all the cases teratomas are associated with malformations such as cleft palate, bifid tongue, and bifid uvula [17]. Our patient had cleft palate.

Conclusion

Infants with nasopharyngeal teratoma with cleft palate often have a poor prognosis or are stillborn. Diagnosis on prenatal ultrasonography allows for early consultation with surgical subspecialists, discussions with parents regarding possible developmental delays, and appropriate preparations for delivery including cesarean section and tracheostomy in a tertiary care center. A good outcome after resection of complex, giant tumors like this one is possible under optimal conditions. The multidisciplinary and staged surgical approach to the management of the lesion in this patient were essential to the successful outcome.

Conflict of Interest None.

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