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Case report

Oral teratoma (epignathus) in a newborn: A case report

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ABSTRACT

An oropharyngeal teratoma (epignathus) is a rare malformation composed of cells from all three germinal layers and few papers have been published about it. Epignathus arises from the palate or pharynx and protrudes from the mouth. We present a large skin-covered teratoma that arose from the hard palate in a neonate. A total surgical excision was performed and histology examination showed mature teratoma with no malignant features.

1. 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]. In Greek teratoma means; monsterous tumour. It is a tumour composed of multiple tissues foreign to the normal organ from which it arises. Teratomas are true neoplasms originating from pluripotent cells and are composed of tissues from all three germinal layers, usually benign in nature. The most common sites are the sacrococcyx, anterior mediastinum, testicle, ovary, or retroperitoneum [1,2]. Teratomas of head and neck are exceedingly rare and only about 10% of teratomas are found in this area. Nasopharynx and cervical region are the most common sites [3–8,23]. Epignathus; is commonly used to describe a congenital teratoma in the oropharyngeal region, with an estimated incidence of one in 35,000 to one in 200,000 live births [5,25,29]. We describe a female neonate with a large oral teratoma originating from the anterior hard palate that was successfully treated with surge.

2. Case report

A 2.6 kg female neonate born at 38 weeks gestation in August 2010 by vaginal delivery to a 22-year-old mother (G2 P1) who had irregular antenatal examinations during pregnancy and obstetric history was unremarkable. Apgar score of the newborn was normal. Examination showed a large, bony, fleshy, trapeziform mass 6 cm × 5 × 3 cm in diameter with smooth surface covered on one side with numerous fine and coarse hair and protruding from her oral cavity (Fig. 1).

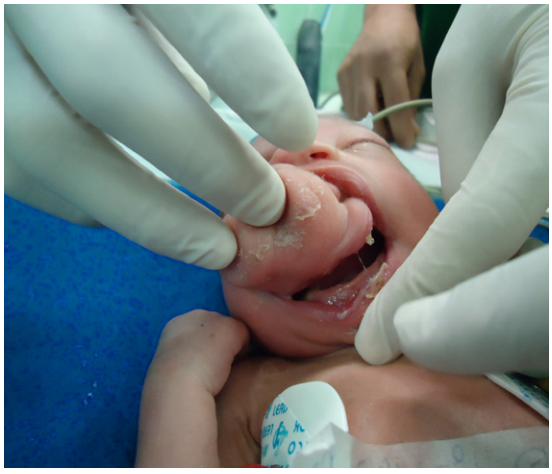

Fig. 1. A large, bony, fleshy, trapeziform mass protruding from her oral cavity.

Due to the oral mass, feeding was not possible and there was mild respiratory distress. She was referred from her hospital of birth to our hospital, At the initial examination the mass was mobile and attached to a stalk that originated from the hard palate (Fig. 2).

Computed tomography showed a complex mass protruding from the oral cavity. It was viewed three-dimensionally and seen to arise from the palate. It consisted of cystic, fat, bony, and neural elements, with defined teeth within the bone (Fig. 3).

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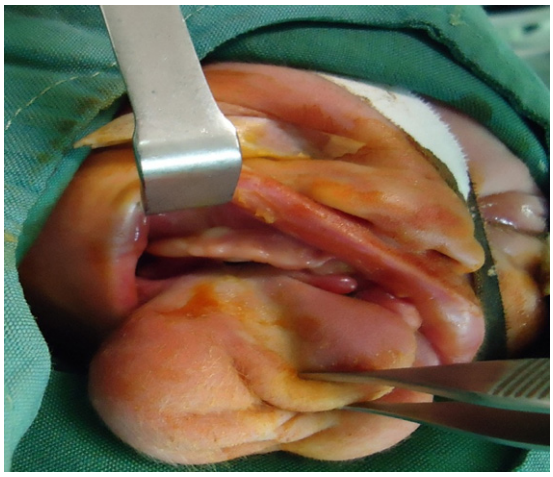

Fig. 2. The mass was mobile and attached to a stalk that originated from the hard palate.

As there was no associated anomaly and the mass was localized, an excisional biopsy was planned. Under general anesthesia, the mass was ligated and totally excised at 10th day (Fig. 4).

After excision of the mass, hemostasis achieved (Fig. 5).

Histopathological examination of the mass revealed mature teratoma with mature keratinising squamous epithelium, skin adnexa, adipose tissue, teeth, neurological tissue and bone formation. Smooth muscle fibers were scattered throughout of specimen (Fig. 6).

Postoperatively the palatal wound epithelialised well. She had a nasogasteric tube at the age of 2 weeks. Masseteric function and swallowing slowly improved over several weeks. The infant remained asymptomatic and was discharged home on day 28 of life. At present, she is well and symptom-free at 2 month follow-up.

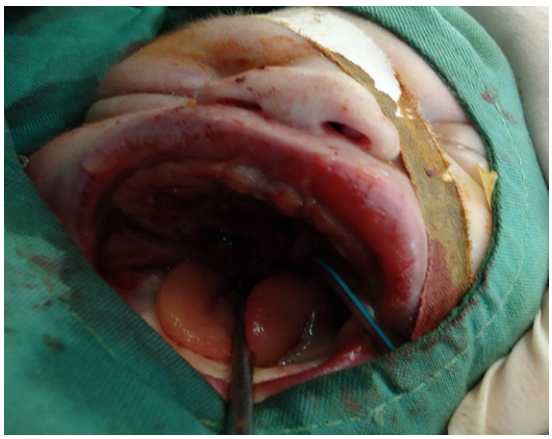

Fig. 5. Oral cavity after excision of the mass.

3. Discussion

Teratomas contain all three primordial germ cell layers (ectoderm, mesoderm and endoderm) [5–8,28] and histologically they may be mature, immature or malignant [6]. They may arise from different sites of the body, the most common site in the newborn is the sacrococcygeal region, (accounting for nearly 40% of the total cases) [9]. Less than 5% occur in the head and neck [9,26]. Epignathus is a teratoma arising from the oropharynx [6,29,30]. When

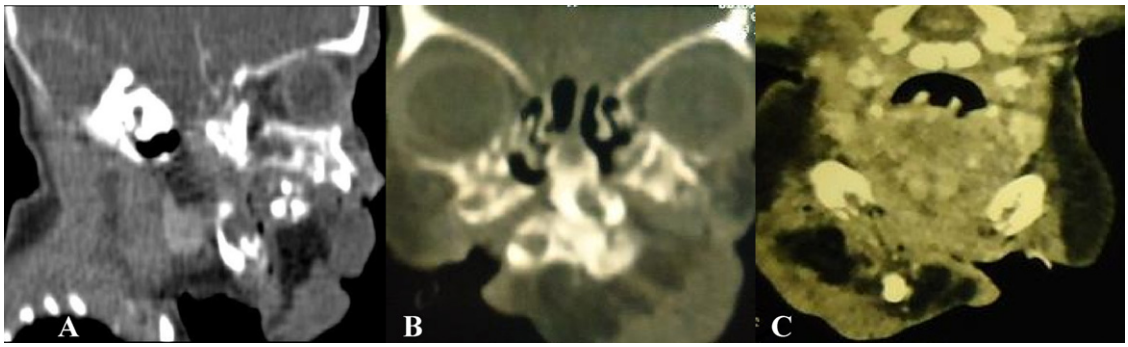

Fig. 3. CT showed a complex mass protruding from the oral cavity. (A) Sagital and (B), coronal and (C) axial views.

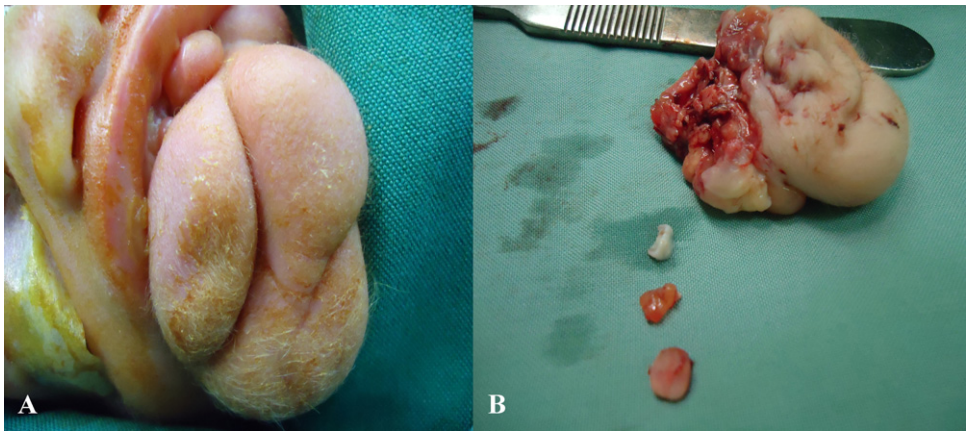

Fig. 4. (A) Dorsal view of mass with fine and coarse hair, (B) ventral view and teeth.

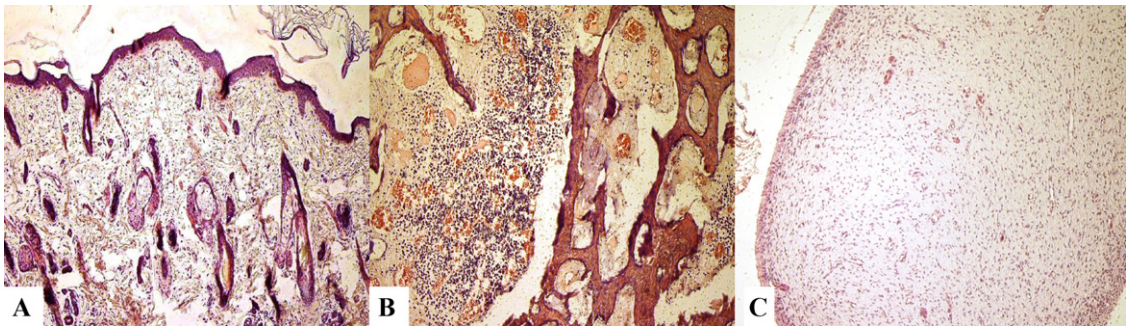

Fig. 6. Histopathology of the mass (mature teratoma): (A) skin and related appendages (ectodermal derivatives). (B) Trabecular bone and bone marrow components (mesodermal derivatives). (C) Neural tissue (endodermal derivative).

large, they fill the oral cavity and can protrude externally, distort facial anatomy and often cause respiratory embarrassment at birth. Our case was noted to have an oral mass causing respiratory and feeding problems. Teratomas are more common in female [3,10,11,13]. Carney et al. found malignant teratoma to be more common in men in a ratio of 5:4 [14].

When present during early childhood, they are usually benign [3,12,23]. As most reported cases, our patient is female and the tumour is benign. There are at least three hypotheses about the aetiology of these lesions [15]. 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 [16]. In our case, incomplete division of blastula may provide enough cells for the production of a teratoma. Tharrington et al. presenting a case of nasopharyngeal teratoma in his review of 850 patients with teratoms [4,10]. It is described in nasopharynx and oropharynx as a solid polyp lesion covered by skin with hair and sebaceous glands and consists mainly of fibro adipose tissue, vascular tissue, foci of smooth and striated muscle, bone or cartilage and glandular tissue [17,27]. In our case the tumour was composed of a fully developed single-root tooth and disorganized mixture of mature mucin secreting glands, adipose tissue, skeletal muscle, and bone covered with skin.

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 [18,22,24]. Some authors, including Andze et al. have reported increased in fetoprotein concentrations prenatally, which are suspicious of a teratoma [19]. The differential diagnosis is limited, but includes hamartoma, dermoid cyst, and heterotopic gastrointestinal cyst [18]. Teeth within the mass confirm teratoma.

Resection is the treatment of choice, as there may be a small chance of malignant transformation the longer they are left [20]. 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 [21]. Exclusion of intracranial extension is an important part of preoperative management [5]. Mortality rate associated with large teratomas in the head and neck are generally high in the absence of a well-prepared resuscitation team or meticulous delivery planning to secure the airway, as the majority of these teratomas are associated with obstruction of airway and difficulty in intubation. In the patient described here, it hampered feeding and mild respiratory distress without need to emergency airway management. In our case, the tumour was easily removed. Benign teratomas may recur after excision. Carney et al. discussed three cases of mature sacrococcygeal teratomas that recurred in children [14]. This does not necessarily imply malignancy, although the clinician should maintain follow-up. This report describes serial imaging studies during the pregnancy including sonography, MRI, and CT with delivery by the ex utero intrapartum treatment (EXIT) procedure. Teratomas arising from the oral cavity are rare in the newborn; only 13 cases have been reported in the literature. In conclusion, we described a rare case of epignathus teratoma.

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