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CLINICAL CASE OF AN OROPHARYNGEALTERATOMA (EPIGNATHUS) IN A PATIENT WITH CONGENITAL CLEFT LIP AND PALATE

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Abstract:

Oropharyngealteratomas (Epignathus) are rare, congenital tumors, which can develop from the sphenoid bone, the hard or soft palate, the pharynx, the tongue or the jawbone. A teratoma is a tumor that develops from two or three different embryonic cells. The malformation consists of multiple tissue types and is usually found in an anatomical region of the body where these tissues should not naturally occur.

We present a case study of an oropharyngealteratoma in a one-year old female patient with congenital unilateral cleft lip and palate. We observed a significant increase in tumor volume from birth, to the point that the tumor almost completely occupied the oral cavity and oropharynx of the patient. This led to a condition of significant pulmonary distress.

Keywords: *Epignathus, Cleft Lip and Palate, Oropharyngeal,Teratoma.*

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INTRODUCTION

Epignatus is a rare orofacial teratoma, which develops from the sphenoid bone, hard and soft palate, pharynx, tongue and jaw. A teratoma is a rare tumor which consists of multiple tissue types, developing from two or three embryoniccells, the presence of which is unusual for those organs and anatomical regions of the bodyin which the malformation is located [1, 3].

Teratomas develop most commonly from the base of the skull and then grow into the nose cavity, mouth, hard palate and mandible. The intensive growth of the malformation changes the anatomy of the maxillofacial area and can cause asymmetry to the face and obstruction of the oral cavity and oropharynx. Nasopharyngeal teratomas are usually found on the left side of the nasopharynx and oropharynx (73%), but can be found on the back of the soft palate (15.4%), and also at the base of the tongue (11.6%) [1].

CASE REPORT:

In our case report a 13-months-old girl was diagnosed with an oral epignathus teratoma and an uncommon combination of orofacial malformations including cleft lip and palate. The patient was also diagnosed with several medical conditions; congenital abnormality on the right hand; Second degree Bronchopulmonary dysplasia. Intracranial compensatory hydrocephalus and delayed motor development.

A full term normally delivered 3.7 kg female was referred to us with a huge mass attached to the tongue and protruding out of the mouth. A second smaller mass could be visualized arising from the lateral oropharynx, through the open mouth [Fig.1].

At the age of 3 months, 13 days, the child underwent a biopsy of the surface of the tumor and was diagnosed with a mature oropharyngeal teratoma. Due to the rapid growth of the teratoma which caused a respiratory obstruction at the age of 3 months, 23 days, the patient developed pneumonia it necessary for a tracheostomy to be performed.

Due to the size of the tumor, the child had–severe respiratory distress. Local examination revealed a large, irregular mass 4 cm X 5 cm in size protruding out of the mouth. The consistency was variegated. The baby presented with a cleft lip and palate with a defect of the skin and prolabium on the right-side of the upper lip. The right alar of the nose was considerably flattened, the tip of the nose pulled downward with the nasal septum displaced to the left. The cleft palate was filled with tumor masses exhibiting a smooth surface, was painless on palpitation and displayed a soft consistency. The teratoma was located within the oral cavity and was approximately 5x6 cm in diameter, causing obstruction of the velopharyngeal ring and considerable distress to the respiratory function. There was a bone defect in the alveolus; a large fragment of alveolar bone protruded vertically and anteriorly. The incisors had erupted from the larger fragment of the alveolar bone.

An Ultrasonography examination taken at 28 weeks showed the malformation, which at that point was 20 mm by 15 mm in diameter, consisting of both cystic and solid components. The ultrasound also showed unilateral cleft of the lip, alveolus and palate. At the age of 1 month, a clinical examination revealed that the tumor, was obstructing the throat, and was thus causing breathing difficulties. A CT of the head and neck showed a complex mass protruding from the oral cavity, the congenital cleft of the lip, alveolus and palate. It was viewed three-dimensionally and seemed to originate from the palate.

The patient underwent surgery and an endotracheal intubation was done. Both masses were excised using a surgical scalpel [Fig. 2]. Cheiloplasty was performed after the excision of the tumor masses. A histopathological examination of the mass revealed a mature teratoma with keratinising squamous epithelium, adipose tissue and a tooth follicle [Fig. 3].

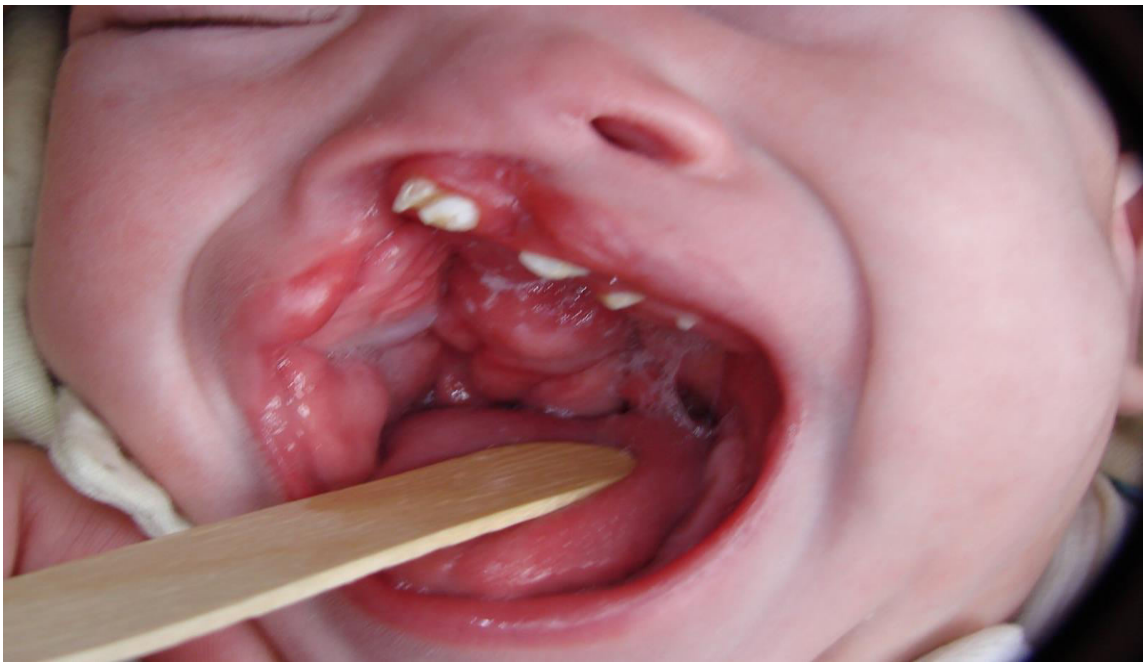

Fig. 1. A large mass attached to the tongue and protruding out of the mouth.

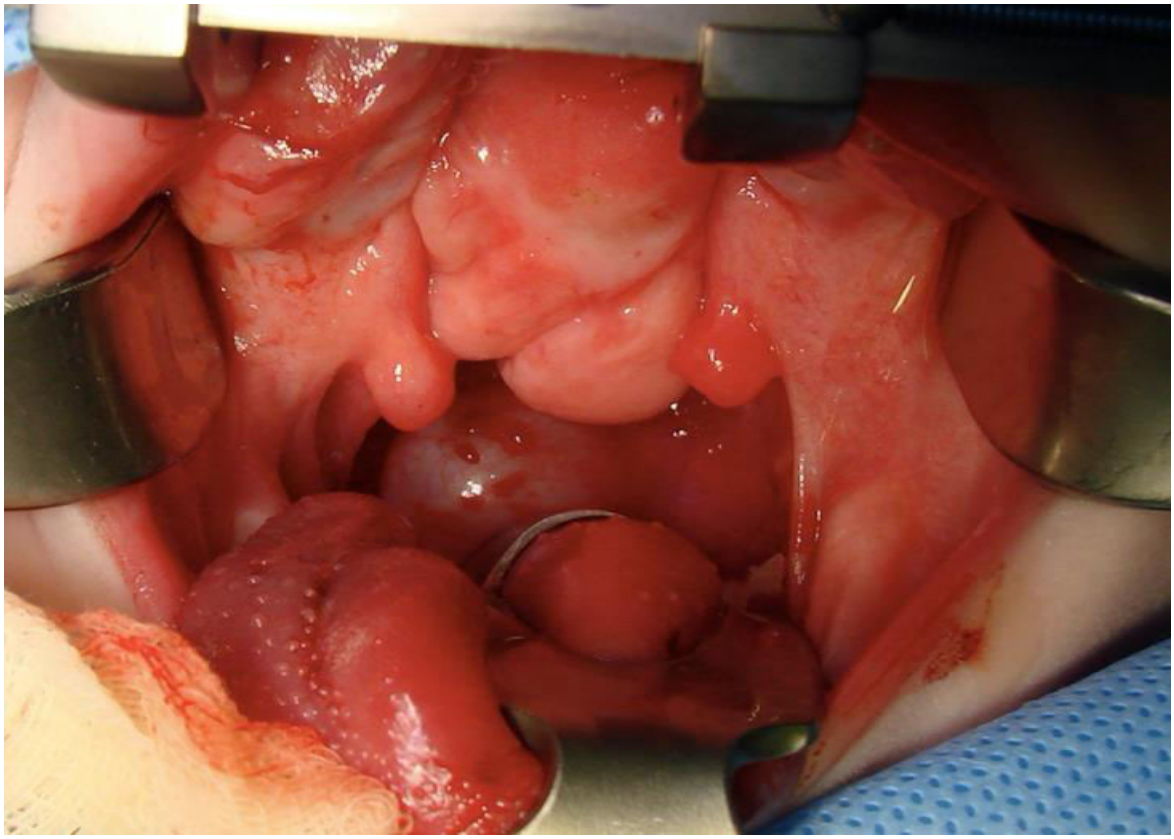

Fig. 2. Teratoma masses protruding through cleft palate.

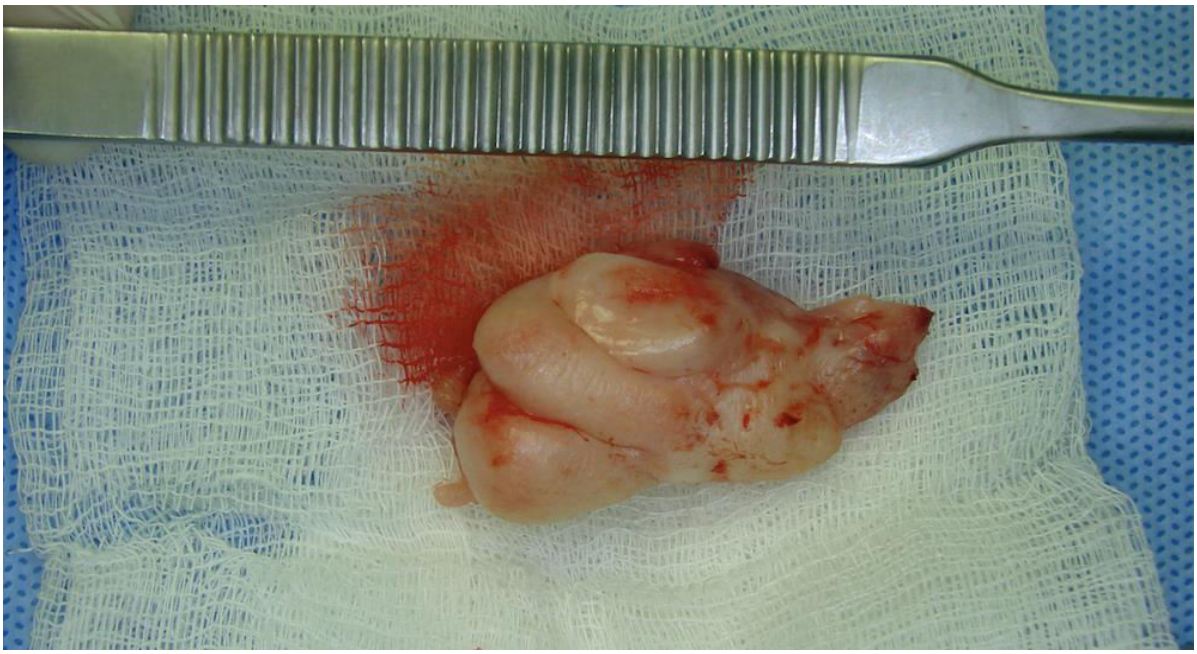

Fig. 3. A mature teratoma with keratinising squamous epithelium, adipose tissue and a tooth follicle.

DISCUSSION:

Teratomas are rare malformations containing cells from ectodermal, mesodermal and endodermal layers with a variable degree of differentiation [6].

Although teratomas are relatively rare tumors [2], in reviewed literature we found several reports of its different localization in neonates [4, 5]. Less than 2% of these arise from the oropharyngeal cavity [12, 14]. These tumors usually contain recognizable organs. Some authors suggest that teratomas arise from totipotential primordial germ cells. One theory suggests germ cells may give rise to teratomas by parthenogenetic development. Other theories, that teratomamay originate from incomplete formation of Siamese twins [8]. In our case the tumour was composed of keratinizing squamous epithelium, adipose tissue and a tooth follicle.

Teratoma can be associated with other midline anomalies (Pierre Robin syndrome, meningoencephalocele, cleft lip, cleft palate or combined with other congenital malformations [5, 6]; For example, in our case, the teratoma of the oral cavity and oropharynx was accompanied by severe congenital deformity of maxillofacial area, with a complete cleft lip and palate. It seems evident that rare cases of tumors and congenital maxillofacial require further detailed study.

Resection is the treatment of choice, as there may be a small chance of malignant transformation the longer they remain [10]. However,–this depends on the site of the tumor. Unless the teratoma is expanding massively into the cranial area, resection of tumor may be attempted.

Initial treatment should be directed toward airway management and feeding problems [9]. In our case report the patient experienced severe respiratory distress, due to a fast growth of the tumor masses.

Cleft lip and palate patients experience severe respiratory distress and nutrition problems. When associated with oropharyngeal tumors, can lead to high mortality rates. In our case report we aimed to ensure the airway and feeding by removing the mass, and performing primary cheiloplasty. In the post-operative period, the patient's respiratory functions and feeding was stabilized. The patient is currently under longterm observation.

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