

Case Report

Congenital large maxillary teratoma

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ABSTRACT

Teratoma of the maxilla is a rare entity. Congenital intraoral teratoma occurs in 1:4000 births. It is a benign tumor, although malignancy has been described in adults. A 10-year-old male child with this condition is described in this report. This case illustrates a huge mass on the right side of the maxilla. The mass was excised under general anesthesia. Histopathologically, it consisted of all three layers of embryonic elements with predominantly fibrous tissue. Postoperative result was uneventful and no recurrence was detected after 2 years.

Key words: Epignathus, maxilla, neonatal, teratoma

INTRODUCTION

Teratomas are congenital germ cell tumors that contain tissues of variable maturity and have a known malignant potential, which is unpredictable from their histological features or stage of development. Teratomas are composed of various tissues of ectodermal, mesodermal, and endodermal origin. Teratomas occur in approximately 1 in 4000 live births, showing a female preponderance, and have an 18% risk of other congenital malformations, some of which can be incompatible with life. Ten percent of teratomas are found in the head and neck area.^[1,2] The most common sites of origin of it in children are the sacrococcygeal region, gonads, and mediastinum. *In utero* it can cause hydramnios or fetal death.^[3] In newborns, it can cause respiratory distress due to tracheal compression.

CASE REPORT

A 10-month-old male child reported to the Department

of Oral and Maxillofacial Surgery of C.S.M. Medical university, Lucknow, India with chief complaint of swelling over the right side of upper lip and cheek. At birth, he was in good condition but there was a large polypoid growth protruding from the right upper lip. The growth was obliterating right half of the oral cavity and right nasal aperture, pushing right ala upward. Lower lip was not involved. On examination, the lesion measured 6 × 4 × 4 cm. It was cystic, but contained ill-defined rubbery nodules [Figure 1]. Biopsy of lesion done under local anesthesia confirmed the diagnosis of teratoma. Soon after diagnosis, under general anesthesia, it was excised through a right transverse incision combined with massive dissection [Figure 2]. Layer wise suturing was done [Figure 3]. Microscopic examination revealed a cystic mass composed entirely of adult tissue from all three germ cell layers. The cystic cavities were lined by stratified squamous epithelium or pseudostriated respiratory epithelium, and underlying the epithelium were groups of mucous glands, small islands of cartilage, brain tissue, and transitional epithelium. Smooth muscle fibers were scattered throughout.

The child was discharged on the tenth postoperative day, having had an uneventful postoperative course. No further therapy was planned because the lesion was non-malignant. On a follow-up period of 24 months, the patient was found to be disease-free.

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Figure 1: Preoperative photograph showing lesion and its extension



Figure 2: Intraoperative photograph showing extensive dissection



Figure 3: Postoperative photograph after suturing

DISCUSSION

Teratomas are composed of various tissues of ectodermal, mesodermal, and endodermal origin. These tissues exhibit various degree of maturation.^[4]

Teratomas are monstrous lesion that is composed of tissues foreign to the part in which they arise. They are classified in four groups as dermoid cyst, teratoid cyst, teratoma, and epignathus. Dermoid cysts are derived from the endodermal and mesodermal layers. Tumors composed of all three germ layers that are poorly differentiated are called teratoid cyst and those that are well-differentiated are called as teratoma. Epignathi are oral tumors containing fetal organ and structures.^[4] Intraoral teratomas are rare among head and neck teratomas. Giant epignathi that present at birth fill the oral cavity and protrude from the mouth; it may cause respiratory obstruction. Isaacs *et al*^[5] reported that perinatal germ cell tumors have been mentioned in the literature: 16 cases with hard palate origin, 14 with nasopharyngeal origin, 6 with sphenoid origin, and 6 with oropharyngeal origin. Attachment is usually to the maxillary region, sphenoid, the lateral wall of the pharynx, and the palate.^[6-9] On palpation, they are cystic, but this alternates with solid areas. The differential diagnosis is that of cystic hygroma, lymphangioma, encephalocele, lymphovenous malformation, and brachial cyst. Calcification and cysts within a mass are typical of teratomas of the head and neck than of other sites. This lesion should be differentiated from encephalocele, glioma, haemangioma, congenital rhabdomyosarcoma, and neurofibromatosis. Early surgery is indicated. The aim of surgical management was to remove the diseased mass as well as to provide good airway and esthetic in long-term follow-up. Furthermore, the extirpation of mass should be done as soon as possible, although there is no initial upper airway obstruction, as the problem develops eventually. Histologically, these are benign tumors. Malignant degeneration has been observed, mainly in adults.^[10,11] This case is presented because of its rarity and to emphasize that surgery should be performed as soon as the patient permits. Obstruction of airway requires immediate intervention.

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