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Case report/Kazuistyka

An unusually giant myxoma of the maxilla in a child – A case report

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ABSTRACT

Myxoma of maxilla is rare and slow growing benign and locally aggressive mesenchymal tumor. Odontogenic myxoma (OM) in maxilla is less common and more aggressive than that of the mandible. Commonly this tumor occurs in second and third decades of life, being rare in children and older adults. This tumor often grows without symptoms and presents as a giant size swelling. Clinical presentations show a slow growing painless mass marked by facial asymmetry leading to gross cosmetic disfigurement. Pain and paresthesia are rare, thus this tumor may reach to the considerable size before patient seeking the treatment. We report a rare case of huge myxoma over the left maxilla in an 11-year-old boy.

Introduction

Myxoma in the maxilla is an uncommon tumor and rarely encountered in patients of pediatric age group. This tumor originates in the mesenchymal portion of the dental follicles. Myxoma is more often seen in mandible than maxilla. This tumor is not encapsulated and characterized by a proliferation of a few rounded cells, fusiform and star cells with myxomatous stroma. Islands of odontogenic epithelial tissue may be scattered in stroma, which are important for diagnosis. This neoplasm proliferates slowly, but due to its aggressiveness and high chance of recurrence, need radical management. When it involves the maxilla, odontogenic myxoma (OM) can invade the maxillary antrum and then diagnosed later one only when having growth to very large mass. It has property of aggressiveness with infiltration to surrounding tissues because of lack of capsule, so removal of this tumor completely is difficult [1]. Vircho coined the term myxoma in 1863 for neoplasms those had histological similarity to the mucinous substance of the umbilical cord [2]. Often this lesion appears in the second and third decades of life whereas it is rare in children and adults with more than 50 years of age. Despite its benign nature, there is high chance of recurrence after simple curettage only and in certain cases need adequate excision.

Report

An 11-year-old boy was referred to the outpatient Department of Otorhinolaryngology for a growing mass in the left

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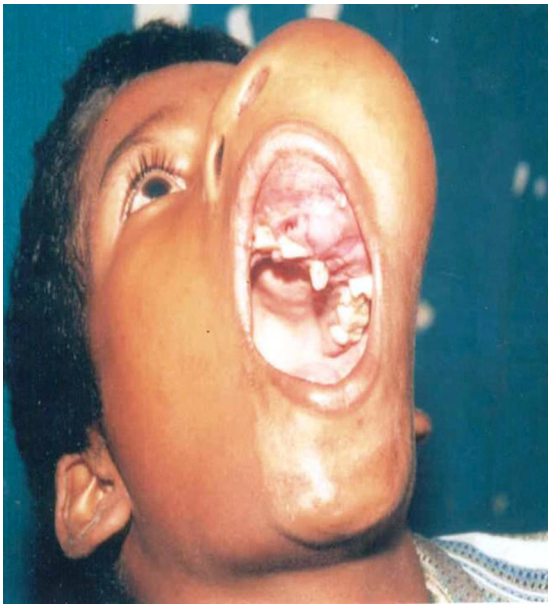

Fig. 1 – Patient showing a large swelling over the left maxilla

maxillary area. The patient reported that such growth as evolved during 1 year period. There was no history of trauma, pain, tooth extraction or fever. On examination, there was a giant swelling at the left side of the face with obliteration of left naso-maxillary groove (Fig. 1). The skin overlying the mass was normal. He had partial left nasal obstruction. There was no bleeding, ulceration or sensory loss of upper lip, oral structures and teeth. An intraoral examination revealed no swelling in the palatal area. The rest of the clinical examination of the head and neck was unremarkable. The computed tomography (CT) scan showed an expansile mass on the left maxilla which completely

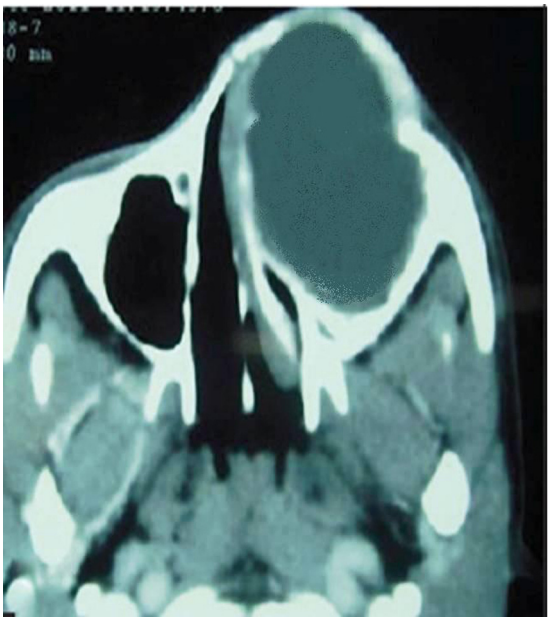

Fig. 2 – Computed tomography (CT) scan showing the mass in the left maxilla and it's extention

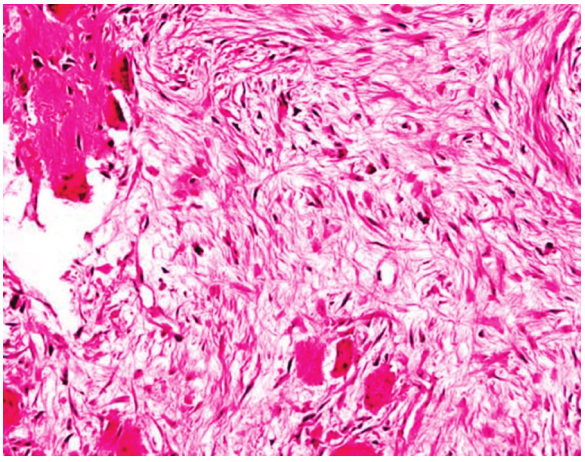

Fig. 3 – Microphotograph of hematoxylin and eosin section revealing stellate shaped cells in the fine fibrillary stroma (100×)

obliterated the maxillary antrum (Fig. 2). An incisional biopsy through sublabial approach confirmed the diagnosis of odontogenic myxoma (OM). The main treatment was surgery *via* sublabial route, resecting the tumor mass with a margin of normal tissue. In this patient for avoiding future facial asymmetries and dental abnormalities, complete enucleation of the mass was done through sublabial approach. The mass was sent for histopathological examination which confirmed the diagnosis of OM. Histopathologically, the tumor consists of loosely arranged spindle cells with serpentine nuclei within a variable fibrous and myxoid stroma (Fig. 3). Postoperative recovery was uneventful and since then the patient is under regular follow up. He will be monitored long term clinically and radiologically for recurrence.

Discussion

Odontogenic myxoma (OM) is a benign, mesenchymal, slowly proliferative and locally aggressive tumor with high chance of recurrence. Odontogenic maxillary myxoma was first described in the literature by Thoma and Goldman in 1947 [3]. Several studies show female predilection for maxillary myxoma [4]. It usually occurs in the 2nd and 3rd decades of life, rarely in children or adults over the 50 years of age [5]. Our case was male with age of 11 years. Even the origin of the myxoma is still obscure; an origin from dental follicle seems to be the most accepted explanation. Often the OM is encountered in the mandible than maxilla. However, one study by Zimmerman and Dahlin reported, myxoma can be equally seen in both mandible and maxilla [4]. Myxomas are usually seen intra-orally mostly at the posterior part of the mandible and rarely extra-orally [6]. The maxilla and anterior part of the mandible are rarely affected by the myxomas. The myxoma may be diffused or well defined, unilocular or multilocular. Myxoma is characterized by gelatinous or mucous grayish-whitish tissue that replaces the spongy bone and displaces the cortical plates of