Odontogenic Myxoma of the Maxilla: A Case Report with Review of Literature

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ABSTRACT

Odontogenic myxomas (OMs) are rare benign mesenchymal tumors of head and neck with a potential for local infiltration and recurrence. They appear to originate from the dental papilla, follicle or periodontal ligament in mandible and less commonly in maxilla. They usually present in second or third decade of life as slowly progressive space occupying lesion in the jaw, giving a mixed radiopaque-radiolucent appearance. The treatment is considered to be wide local excision in view of high recurrence along with curettage. Here, we present a case of odontogenic myxoma of maxilla, in 18 years old adult, with a brief review of literature, clinical, radiological, histopathological characteristics.

Keywords: Mesenchymal tumor, Myxoma, Odontogenic.

INTRODUCTION

In the international histological classification of odontogenic tumors, odontogenic myxoma (OM) is defined as a benign odontogenic tumor of mesenchymal origin that is locally invasive and consists of rounded and angular cells lying in abundant mucoid stroma. The average age of patients with myxoma is 25 to 30 years. Odontogenic myxomas represent approximately 1 to 17.7% of all odontogenic tumors. Being among the rarest variety of benign tumors, they are characterized grossly by mucoid or gelatinous grayish-white tissue.

Myxomas can occur anywhere in the jaws but have a predilection for the molar and premolar regions of the mandible and maxilla. Maxillary myxomas often extend into the sinus. It is an asymptomatic lesion that shows an infiltrative growth pattern. Such lesion causes a destruction of the medullar bone and expansion of the cortical bone. Frequently, the lesion is perforating and invading the adjacent soft tissues and cortical bone. The neoplasm occurs within a similar distribution both in males and females.

Radiologically, the appearance may vary from a unilocular to a multicystic lesion with a well defined or diffuse margin. A unilocular appearance may be seen more commonly in children and in anterior parts of the jaws. Microscopically, these lesions are characterized by stellate and spindle-shaped cells embedded in a richly myxoid extracellular matrix, with scarce collagen; those cases with higher amounts of collagen may be denominated as myxofibroma.

Here, we report a case of odontogenic myxoma of maxilla, in 18 years old adult.

CASE REPORT

An 18-year-old male presented with chief complaint of painless swelling in right middle third of the face since one and a half years. His past medical history was of no relevance and general physical status was satisfactory. Extraoral examination revealed swelling in middle third of the face which extends from ala of the nose to the tragus of ear which is localized and nontender (Fig. 1A). On intraoral examination, there is obliteration of buccal vestibule in the region of 12 to 16 (Fig. 1B) with negative for aspiration. Reconstructed panoramic image using Dentascan Software revealed opacification of the right maxillary sinus and the size of the pathology to be 35 x 33.9 mm extending from 12 to 16 region (Fig. 1C). Incisional biopsy of the lesion was performed. On microscopic examination, loosely arranged angular, spindle and comma shaped cells resembling dental papilla in a delicate myxoid stroma were noted (Fig. 1D). Numerous cells demonstrate long, thin, fine anastomosing processes that tend to intertwine the collagenous matrix which was suggestive of odontogenic myxoma (Fig. 1E).

The tumor was surgically excised under general anesthesia. The gross macroscopic examination of the excised specimen showed a white gelatinous appearance. Microscopic examination of the same confirmed the diagnosis of incisional biopsy. Patient recovery was...
uneventful and upon 1 year follow-up showed no evidence of recurrence.

DISCUSSION

Odontogenic myxoma is a tumor considered to be arising from embryonic mesenchymal elements of dental anlage, such as dental papilla and Hertwig’s epithelial root sheath. Evidences supporting the odontogenic origin are: (a) its occurrence is almost exclusive, only to the tooth bearing areas in the jaws, (b) occasional association with an unerupted tooth or a missing tooth, (c) occurrence in younger individuals, (d) histological resemblance to dental papilla, and (e) occasional presence of odontogenic epithelial island.

The OM cells articulate extracellular membrane molecules, like fibronectin, type 1 collagen and tenasin resembling human immature dental papilla stem cells. Nestin, a marker for progenitor cells, was affirmative in the stromal neoplastic cells of the OM. So, Martinez-Mata G et al suggested that the possible origin of tumor cells are from the dental papilla cells, fibroblasts or myofibroblasts. Using the notch signaling, Nakano et al concluded that the differentiation level of the tumor tissue is similar to that of cap stage.

Zhang et al, classified radiographic appearances of OM into six types—type I: Unilocular well-defined radiolucency, type II (multilocular): two or more compartments with multiple interlaced osseous trabeculae described as honey comb, soap bubble or tennis racquet radiolucency, type III: lesion located in alveolar bone, type IV: lesion involving the maxillary sinus, type V (mouth eaten appearance): larger radiolucent area with irregular borders and type VI: combination of bone destruction and bone formation giving sun ray appearance.
On gross tissue examination, myxoma appears as a mass of characteristically mucoid, white gelatinous slimy material. Microscopically, odontogenic myxomas demonstrate spectrum of fibrous connective tissue stroma from myxoid to densely hyalinized and from relatively acellular to cellular nature. The tumor shows loosely arranged stellate or spindle shaped cells resembling dental papillary cells in a delicate myxoid stroma. It was sometimes called odontogenic fibromyxoma, because cells were active with granular cytoplasm, dense diffuse collection of fibrils and fibroblasts.

Histopathological differential diagnosis of OM should include a developing dental papilla, dental follicle and non-odontogenic tumors with myxomatous degeneration, such as myxoid neurofibroma, myxoid osteosarcoma, mesenchymal chondrosarcoma, myxoid lipoma, myxoid liposarcoma, myxoid fibrosarcoma, and chordomyxoid fibrosarcoma. Histochemical studies of the mucoid intercellular substance have revealed the presence of large quantities of hyaluronic acid.

Ultrastructural and immunohistochemical studies have shown that the tumor cells positively react with vimentin and muscle-specific actin. Several studies on S-100 and glial fibrillary acidic protein positivity have given conflicting reports. The treatment for the OM is quite variable. The current literature recommended therapy depends on the size of the lesion and on its nature and behavior and can vary from simple curettage and peripheral ostectomy up to segmental resection.

REFERENCES