ODONTOGENIC MYXOMA OF THE MAXILLA IN A LIBYAN FEMALE: CASE REPORT AND REVIEW OF LITERATURE

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ABSTRACT:

Odontogenic myxoma OM is rare odontogenic tumor that is slowly growing, locally invasive lesion with a tendency for local recurrence. Most of OM are expansile and can displace or even resorb the adjacent teeth. We report a case of OM in 22 years old Libyan female who presented with a painless soft tissue swelling at the left side of the hard palate, covered with normally looking overlying mucosal surface with no ulceration or suppuration. The diagnosis of OM was established through its clinical presentation, radiological features and histopathological findings. Further, the advanced imaging techniques, especially CT scans and MR were used in the diagnostic work up and proven to be a useful tool for the diagnosis of OM.
INTRODUCTION:

Odontogenic myxoma (OM) is a rare benign odontogenic tumor that is locally aggressive, slowly growing, persistent and destructive lesion prone to local recurrence. It usually occurs in the tooth-bearing areas of the jaws and believed to be arising from the mesenchymal portion of the tooth germ (most likely from the dental papilla). It has the potential for extensive bony destruction and extension into the surrounding structures. A number of studies revealed that odontogenic myxomas account for 1.5% to 17% of all odontogenic tumors. Almost 75% of OM occur in patients around 23-30 years of age and rarely occur in patients over the age of 50 years or under 10 years of age, with a slight female predilection (1:1.5 male-to-female). It is believed that it occurs almost equally in the maxilla and mandible with a slight predilection for the posterior parts of the mandible. If OM is left untreated, it is invasive and locally destructive. The diagnosis of OM is established through clinical presentation, radiological features and histopathological findings. The advanced imaging techniques, especially CT scans and MR were proven to be a useful tool for the diagnosis of myxoma. The standard treatment for OM is surgical excision with simple enucleation and curettage in order to minimize injury to the facial growth centers. However, a reported recurrence rate of 25% after enucleation, makes it mandatory to follow the patients with myxoma for an extended postoperative duration as recurrences have occurred 15 years after initial surgery.

CASE REPORT:

A case of 22 years old Libyan female was presented to our department in July 2010, complaining of a slowly growing, unilateral soft tissue mass in the left side of the mouth since 7 months. It was associated with mild local pain symptoms and small amounts of blood occasionally coming out on sucking. On clinical examination, there were no abnormal extra oral signs, but intra orally, there was soft tissue swelling measuring about 2 x 4 cm projecting at the left side of the hard palate. The swelling was covered with normally looking overlying mucosal surface with no ulceration or suppuration, Figure 1. The upper left first and second molar teeth were mobile and spaced apart, while the upper left third molar is clinically missing with slight bulging at the maxillary tuberosity area. There were neither neurological symptoms nor bleeding or ulceration in the area and no regional lymphadenopathy. Orthopantomography showed multilocular cyst-like radiolucency extends from the upper first molar extending posteriorly and surrounded completely the second molar and displaced the third molar upward, with a thinning of the maxillary sinus floor. The lesion was separated by straight or curved bony septa giving it a (soap-bubble appearance), Figure 2. Computed tomography (CT) revealed expansile mass arising in the left maxilla and expanded and deformed the base of the right maxillary sinus. As a result, the walls of the sinus were thinned and the teeth were spread out. The growth has perforated the maxillary sinus wall and occupied a considerable space of the sinus, Figure 3. Histopathological examination of biopsied tissues revealed a loosely arranged spindle-shaped satellite cells as well as triangular cells with long processes anastomozing in a mucoid intercellular substance and few areas of inactive odontogenic epithelium. The picture is consistent with the diagnosis of odontogenic myxoma (OM), Figure 4. The lesion was then surgically excised and the patient was kept under regular follow up with no recurrence for the last 3 years.

Fig: 1. The clinical presentation of the lesion intraorally

Fig: 2. Panorama view reveals the hard tissue changes caused by the lesion

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DISCUSSION:
Odontogenic myxoma was first described by Virchow in 1863, as a benign odontogenic tumor of mesenchymal origin that is locally invasive and consists of rounded and angular cells that lie in abundant mucoid stroma. It mostly occurs in the teeth bearing areas of the jaws and should not be mistaken for soft tissue myxoma, which is relatively common lesion of the soft tissues of the body. OM is less commonly seen than odontomas and ameloblastomas and constitutes about 0.5% to 17% of all odontogenic tumors in Europe, Americas and Asia and perhaps occur slightly more commonly in Africa, with relative frequencies between 1% and 19%. For that reason, a pathologist who is not familiar with the histology of a tooth germ can mistake a myxoid dental follicle for an OM. Most of the cases of OM are expansile and can displace and resorb teeth. When located in the maxilla; the tumor frequently involves the sinus and at times (albeit rarity) crosses the midline to the opposing sinus. Radiographically, the majority present as expansile multilocular radiolucency, though some are unilocular with or without scalloped borders, and rare cases present with a diffuse and mottled appearance that can be mistaken for a malignant neoplasm. Grossly, this lesion is gelatinous in nature, making curettage alone difficult; the more fibrotic odontogenic myxomas (also known as odontogenic myxofibroma or fibromyxoma) have more body and are easier to curette. CT images of the OM are seen as osteolytic expansile lesions with bony expansion and thinning of cortical plates or as a soft tissue mass with bone destruction and thinning, and strands of fine lacelike density representing ossifications. The characteristic finding on CT scan may be the strands of fine lacelike density. Histologically, it is made up of loose and delicate fibrous connective tissue. The fibroblasts are stellate and are suspended on a delicate network of collagen fibrils. Small blood vessels are present, as are small odontogenic epithelial islandson occasion. Immunohistochemistry studies suggest that the spindle-shaped cells constituting this neoplasm have a combined fibroblastic and smooth muscle typing, suggesting that it is of myofibroblastic origin. OM must be considered in any differential diagnosis of facial masses and/or odontogenic tumors, and must be ruled out whenever suspected, especially ameloblastoma, dental cysts, non-neoplastic inflammatory processes (nodular fasciitis and giant cell granuloma) and other gelatinous tumors such as embryonal rhabdomyosarcoma, mucoid liposarcoma and neurogenic sarcoma.

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