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Desmoplastic ameloblastoma of the maxilla

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ABSTRACT

Aim: Ameloblastoma is known to be a benign, slow-growing but locally aggressive and infiltrative odontogenic epithelial neoplasm with a rare capacity to metastasize. Peripheral ameloblastoma is a rare extraosseous variant with histological characteristics similar to those of the common intraosseous ameloblastoma. Methods: The present study describes a case of a 66-year-old woman with a maxillary peripheral desmoplastic ameloblastoma. Clinical features, histopathology, surgical treatment and follow-up are discussed. Conclusion: A clinical case of maxillary desmoplastic ameloblastoma is reported. Two months after surgery, a temporary prosthesis was applied. After the planned follow up, final rehabilitation will include bone grafting to reconstruct the resected maxillary area followed by implants positioning. Further reports of new cases are however necessary for a better knowledge of the origin, the biological behavior, and the clinical treatment of this particular odontogenic tumor variant.

Keywords: odontogenic tumor, ameloblastoma, maxilla, peripheral desmoplastic ameloblastoma, oro-maxillo-facial surgery, surgical reconstruction, prosthesis, implantology

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INTRODUCTION

According to the 2005 histological classification of odontogenic neoplasms by the World Health Organization (WHO), ameloblastoma is a benign, locally invasive epithelial odontogenic tumor. It represents about 1% of all oral tumors and about 11% of odontogenic tumors. [1] It is primarily seen in adults in the third to fifth decades of life, with almost equal sex predilection. [2]

This neoplasm originates within the mandible or maxilla from epithelium that is involved in the formation of teeth. Potential epithelial sources include enamel organ, odontogenic rests (rests of Malassez, rests of Serres) and the epithelial lining of odontogenic cysts, especially dentigerous cysts. The trigger for neoplastic transformation of these epithelial residues is totally unknown. [3,4]

This is a lesion of adults, and it occurs predominantly in the fourth and fifth decades of life. The age range is very broad, extending from childhood to late adulthood. Mean age have been most commonly between 35 and 45 years. The rare lesions occurring in children are typically unycistic and appear clinically as odontogenic cysts. There appears to be no gender predilection for this type of tumor.

Present only during tooth development, ameloblasts are cells that deposit tooth enamel, which is the hard outermost layer of the tooth forming the surface of the crown. Microscopically, ameloblastoma is exclusively epithelial. The neoplastic epithelium forms sheets, islands and cords in which the peripheral layer is formed by columnar or cuboidal cells that resemble ameloblasts, and the central mass is usually formed by stellate cells resembling the stellate reticulum of the enamel organ. Although this is the classic and usual microscopic picture, variations do occur. The stroma of the tumor consists of fibrous connective tissue and the tumor is not encapsulated. Tumor islands and clusters infiltrate the marrow spaces far beyond the major bulk of the mass and can elicit desmoplastic reaction and phlogistic response. In reality, therefore, the tumor is more extensive than its radiographic shadow and requires a wider excision or curettage than may appear necessary.

It is classified into four distinct histopathological subtypes: solid, multicystic, unicystic, desmoplastic, and extraosseous peripheral.

Ameloblastomas may occur anywhere in the mandible or maxilla. In the mandible, the most favored site is the molar-ramus area. In the maxilla the molar area is more commonly affected than the premolar and anterior regions. These tumors can cause severe expansion of the cortical bones and gross anatomic deformities. Dentition can be affected causing tooth mobility and displacement. [4]

Despite being categorized as benign and slow-growing, ameloblastoma is a locally invasive odontogenic epithelial neoplasm with a high recurrence rate.[5] Recurrence rates are reportedly as high as 15-25% after radical treatment and 75-90% after conservative treatment. [6]

Radiographically, ameloblastomas are osteolytic, typically found in the tooth-bearing areas of the jaws, and may be either unilocular or multilocular.

MATERIAL AND METHODS - CLINICAL REPORT

In October 2020, a 66-year-old woman came to our observation with a 7-month history of an ulcerated lesion on the palatal mucosa of the right maxillary region. In April 2020, the patient had previously consulted an odontostomatologist who performed an incisional biopsy. Having been diagnosed ameloblastoma, the patient underwent several curettages of the lesion with no clinical resolution. Pain and bleeding associated with the lesion were reported. At our clinical examination, there was a sessile mass of about 3 cm extending on the palate from region tooth 21 to region tooth 14 (Figs. 1A,1B). The mass was red and ulcerated. There was bleeding and

pain on palpation. No other lesions in the oral cavity and in the oropharynx, or abnormalities in the head and neck area were reported. CT scan revealed initial bony erosion from 21 to 14 (Fig. 2). A new biopsy was then performed. Histological examination assessed: “peripheral desmoplastic ameloblastoma”. The patient subsequently underwent resection of the lesion with surrounding bone alveolar tissue and removal of the teeth 21, 11, 12, 13, 14 *en bloc* with the surrounding maxillary bone (Fig. 3). A vestibular mucosal flap was rotated and used for closure (Fig. 4). The final pathologic diagnosis was: “peripheral ameloblastoma, with a desmoplastic pattern, with initial aspects of infiltration in the bone tissue, contained within the margins of resection”. No complications were observed during recovery. A temporary prosthesis with the missing teeth was applied after 2 months. No recurrence of disease was observed after 24 months follow-up (Figs. 5A,5B). After the planned follow up, final rehabilitation will include bone grafting to reconstruct the resected maxillary area followed by implants positioning.

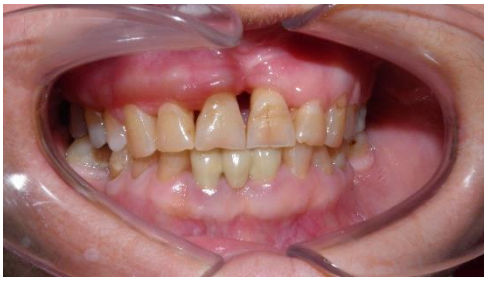

Figure 1A - Ameloblastoma of the palate mucosa extending from region 21 to region 14 with areas of bony erosion.

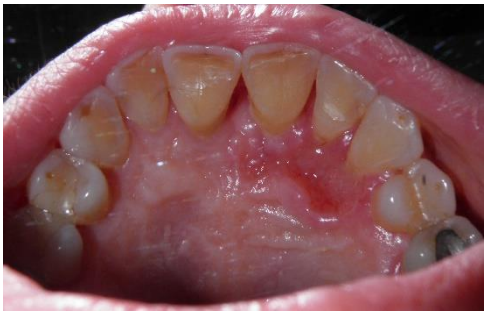

Figure 1B - Intraoral view. Partial palatal mucosa erosion. No evident signs of ameloblastoma.

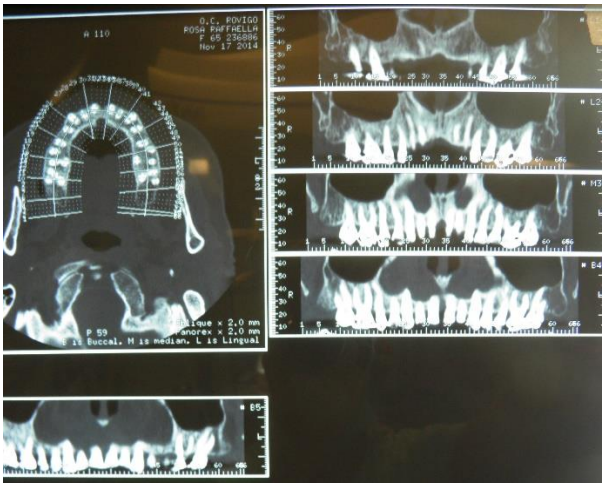

Figure 2 - CT scan. In these CT scan sections initial bony erosion is visible in the maxillary area 21-14.

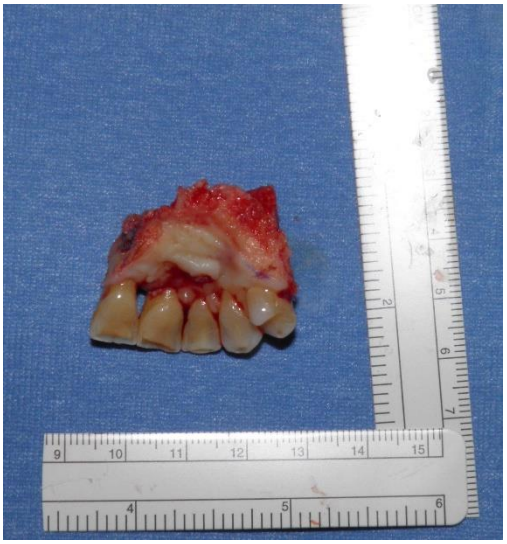

Figure 3 - Surgical specimen including dental elements from region 21 to region 14 en bloc with the corresponding alveolar process bone and the palatal mucosa. Macroscopically erosion of the maxillary-alveolar bone is evident.

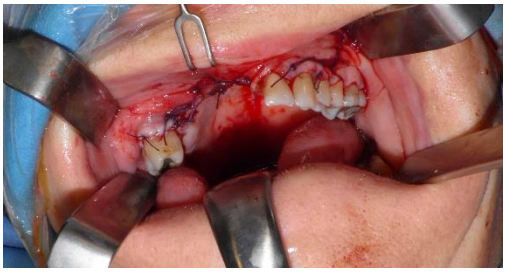

Figure 4 - Final view after closing and suturing a with sliding mucosal palatal-vestibular flaps.

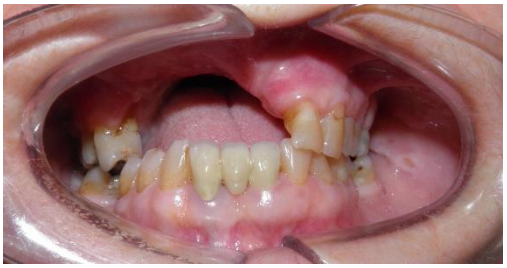

Figure 5A - Intraoral close up: after 24 months: complete healing of the oral mucosa

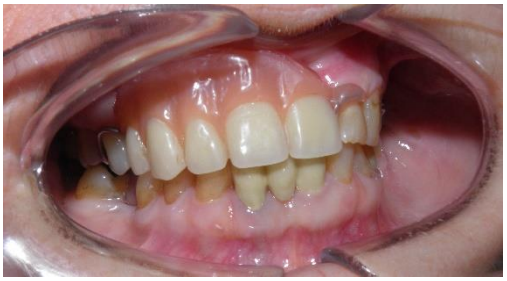

Figure 5B - Intraoral view after prosthetic rehabilitation with a removable prosthesis. No esthetic or functional residual deficits. After the planned follow up, maxillary bony reconstruction and implants will rehabilitate the entire maxillary-dento-alveolar area

RESULTS AND DISCUSSION

Ameloblastoma is a benign, slow-growing but locally aggressive and infiltrative odontogenic epithelial neoplasm with a rare capacity to metastasize. Desmoplastic ameloblastoma, since its first description by Eversole et al. [7,8] is considered a rare variant, with an incidence of 0.9% to 13% among all ameloblastomas. [9,10,11,12]

Histopathologically, desmoplastic ameloblast-tomas are nonencapsulated tumors with extensive collagenous stroma or desmoplasia containing small islands and nests of ameloblast cells. As the tumor is nonencapsulated, the cells infiltrate between the trabeculae of the cancellous bone leaving them intact for some time [13,14]. Ameloblastoma usually occurs in the jawbones, but tumors characterized by similar histological features have also been reported in gum tissues. This group of diseases is referred to as peripheral, extraosseous, or soft tissue ameloblastoma. [15]

Peripheral ameloblastoma is a rare variant, representing approximately 2% to 10% of all ameloblastomas. In particular it occurs in a significantly higher age group, usually between 40 and 60 years of age, than does intraosseous ameloblastoma. Peripheral extraosseous ameloblastomas may occur in the gum and very rarely in the buccal mucosa. They may arise from overlying epithelium or rests of Serres. They exhibit a benign, nonaggressive course and generally do not invade underlying bone. Following local excision, recurrence is rare.

Peripheral desmoplastic ameloblastoma is a very infrequent histological variant. It is a rare event, reported in only three cases involving adult patients and in one case affecting an adolescent patient. This neoplasm shows the clinical features of a noninvasive, slow growing mass of soft tissue covered by normal mucosa of the jaws. [15] It tends to be asymptomatic and to grow very slowly. Peripheral ameloblastomas are usually regular in shape, sessile, firm, and small in size. They are known to erode cortical bone by a mass effect that is referred to as “cupping or saucerization”. [16]

Peripheral ameloblastoma is typically treated by local soft tissue excision to achieve safe margins of 2-3 mm. Recurrence is rare and is more likely the result of incomplete excision rather than aggressiveness of the tumor. [12,13,17]

In this case, a resection of the lesion with surrounding bone tissue was carried out. It could be argued that bone resection would not have been necessary if the reported tumor was actually a peripheral ameloblastoma. None-theless, the resection was performed to ensure negative margins since the potential for widespread destruction and death due to local disease is high because of the fragility of the maxillary bone, the thinness of bone plates, and lack of a dense cortex as in the mandible. [18] The final bone resection specimen revealed a peripheral ameloblastoma with a desmoplastic pattern, with initial aspects of infiltration in the bone tissue contained within the margins of resection. Peripheral desmoplastic ameloblast-toma is a rare variant of the ameloblastoma. It is a noninvasive, slow-growing soft tissue tumor. No single standard type of therapy should be advocated for patients with ameloblastoma. Rather, each case should be judged on its own merits.

Malignant behaviour by ameloblastoma is rarely encountered. Considering that recurrences have been described in the literature, surgery must be radical. Further reports of new cases are however mandatory for a better knowledge of the origin, biological behavior, and treatment of this particular ameloblastoma odontogenic tumor variant.

DECLARATIONS

Authors’ Contributions:

Andrea Edoardo Bianchi and Luigi Clauser designed the clinical data and research; Luigi Clauser performed surgical treatments and samples collection;Luigi Clauser and Riccardo Luoni Orsi prepared the article.

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Not applicable

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None

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All Authors declared that there are no conflicts of interest

Ethical approval and consent to participate:

Not applicable

Consent for publication:

Written informed consent was obtained for all patient images.

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