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# Desmoplasic Ameloblastoma in Maxilla - Report of Case and Review of the Literature

## Ameloblastoma Desmoplásico en la Maxila - Relato de un Caso y Revisión de la Literatura

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**SUMMARY:** The ameloblastoma is a benign neoplasm of epithelial odontogenic origin, slowly growing, locally invasive and it is the most common odontogenic tumor in the jaws. Histologically, various types have been described in literature. The desmoplastic variant is rare and characterized by difference in the typical findings of ameloblastoma, including localization, radiographic and histological features. The purpose of this article is to report a case of desmoplastic ameloblastoma in the left maxilla, and review of relevant literature, emphasizing peculiar aspects of this unusual lesion.

**KEY WORDS: Ameloblastoma; Desmoplastic Ameloblastoma; Odontogenic tumours.**

## INTRODUCTION

The ameloblastoma is a benign odontogenic tumor of epithelial origin that exhibits a locally aggressive behavior with a high level of recurrence, being believed to theoretically come from dental lamina remains, the enamel organ in development, epithelial cover of odontogenic cysts or from the cells of the basal layer of the oral mucosa (Güngüm & Hos¸gören, 2005; Adebiyi *et al*., 2006).

The ameloblastoma is the second most common odontogenic tumor of epithelial origin (25%) that happens in the oral cavity, often attacking the molar regions and ascending ramus of the mandible (Fregnani *et al*., 2007), and only about 20% happens in the posterior maxilla region. The lesion presents prevalence between the third and the fourth life decade, equally affecting both genders and having no predilection for race (Zwahlen & Gräntz, 2002; Ledesma-Montes *et al.*, 2007). The tumor is usually asymptomatic, grows slowly and small lesions can be detected only by routine radiographic exams. The lesion can cause increase in volume, pain, bad occlusion and paresthesia of the affected area (Becelli *et al*., 2002). Radiographically, the neoplasia can be presented as a unilocular or multilocular radiolucent image in the shape of "soap bubbles" or "honeycomb". The re-absorption of the adjacent teeth roots is not found to be uncommon and the presence of a tooth inside, usually the inferior third molar, can be associated to the tumoral mass (Hollows *et al.*, 2000; Neville *et al*., 2004).

Various histological patterns of the ameloblastoma are described on the literature, including the follicular, plexiform and acantomatosous types, of granular cells, basal cells and the desmoplasic (Dos Santos *et al*., 2006). Recently, the World Health Organization (WHO) considered the desmoplasic ameloblastoma not only a histological variant, but a clinic variant of this tumor itself.

The desmoplasic ameloblastoma is a rare variant, initially described in 1984 by Eversole *et al.* (Hirota *et al*., 2005; Desai *et al*., 2006), which presents accentuated predilection for the anterior region of the maxilla and the mandible, preferably for the maxilla (Neville *et al*.). The tumor usually happens between the third and fifth life decade, with an equal attack rate between men and women. It represents from 4 to 5% of all ameloblastomas (Manuel *et al*., 2002). In relation to the radiographic aspects, this type of lesion rarely suggests the diagnosis of an ameloblastoma

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and, many times, it is similar to a fibro-osseous lesion due to its mixed radiopaque-radiolucent appearance of loose outline (Iida *et al.,* 2002; Kaffe *et al*., 1993). Histologically, the desmoplasic ameloblastoma is characterized by a stroma with intense desmoplasia, densely collagenized, comprising little odontogenic epithelium islands and cords of various sizes (Iida *et al*.; Manuel *et al*.; Dos Santos *et al.*).

This article aims to report a case on a desmoplasic ameloblastoma of rare location, emphasizing the clinic and histopathological aspects that are relevant for the diagnosis and treatment of this pathology.

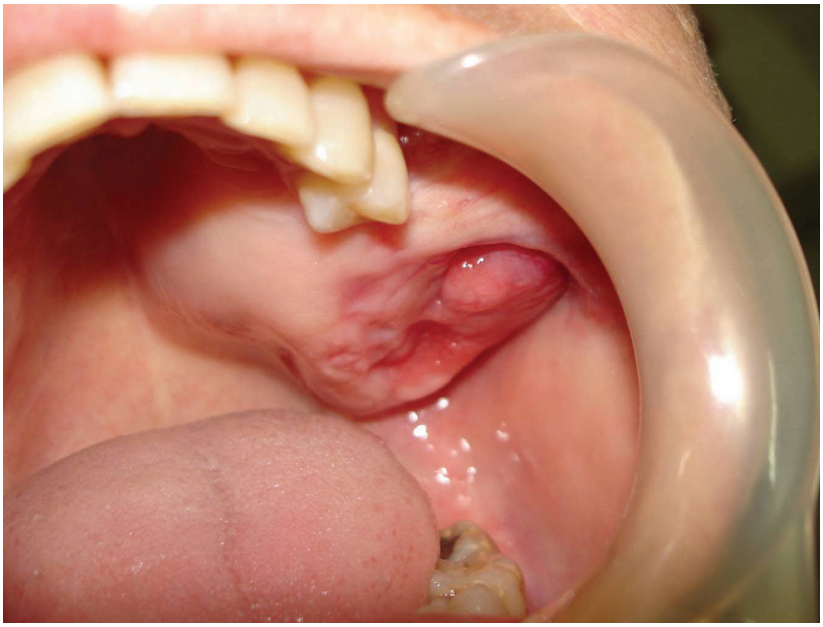

Fig. 1. Tumoral lesion with areas of ulceration in the left maxilla.

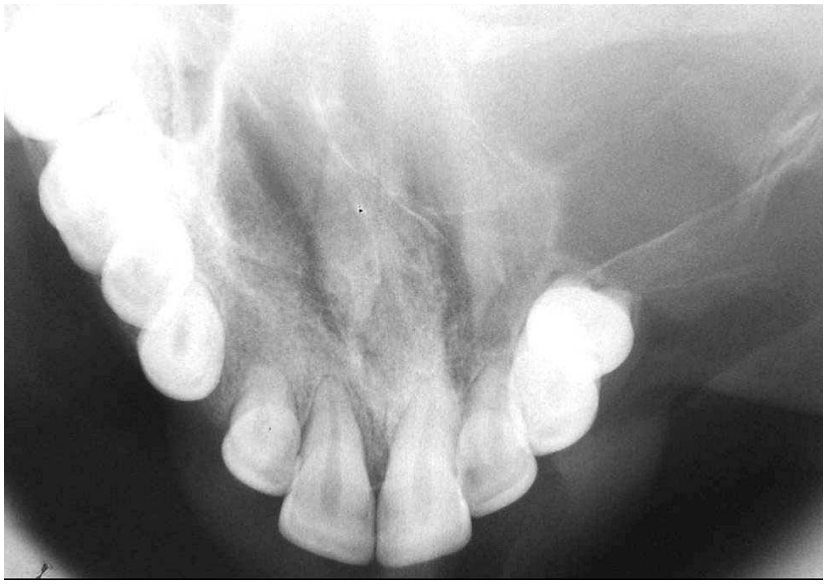

Fig. 2 Radiographies showed the presence of a radiolucent image of irregular edges with discreet radiopaque focus in its interior.

## CASE REPORT

Female patient, 45 years old, white, looked for odontological assistance complaining about a lesion in the left maxilla, in the region that corresponds to the molar one. It has had for 5 years. Through the intra-oral exam, it was observed a tumoral lesion, ulcerated, symptomatic, pinkish-reddish coloration, irregular surface of imprecise outlines, firm consistency, exophytic, sessile implantation and measuring on its biggest diameter 2.5 cm (Fig. 1). Through the ectoscopic exam, there could be seen a volumetric increase of the left side of the face, causing facial asymmetry, involving the ocular globe as well. The patient also complained about nasal obstruction and lachrymation. The occlusal and panoramic radiographies showed the presence of a radiolucent image of irregular edges, with discreet radiopaque focus in its interior (Figs. 2 and 3). Because of this, an incisional biopsy was done and the fragment removed was taken to the Serviço de Anatomia Patológica da Disciplina de Patologia Oral da Faculdade de Odontologia - UFRN. The incisional biopsy reveled fragments of a benign neoplasia of odontogenic origin, characterized by the proliferation of small odontogenic epithelium islands and cords interlarded by a dense fibrous stroma, exhibiting intense collagenized areas (Fig. 4). It can also be seen in some of these islands the presence of squamous metaplasia, peripheral cells arranged in palisade exhibiting hyperchromatism and cores of reverse polarity. In the center of the islands, loosely organized cells were found, resembling the stellate reticulum of the enamel organ (Fig. 5). From these findings, a histopathological diagnosis was issued as of being a desmoplasic ameloblastoma. Then, the patient was sent to surgical treatment in order to have the lesion totally removed. The patient was kept for follow up for 18 months, with no signs of recurrence.

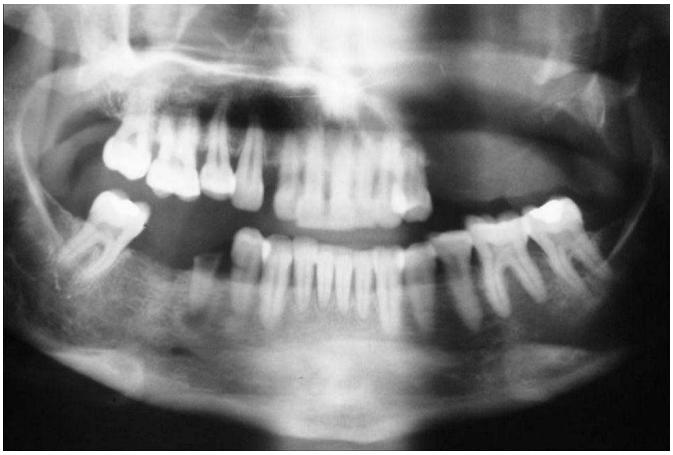

Fig. 3. Radiographies showed the presence of a radiolucent image of irregular edges with discreet radiopaque focus in its interior.

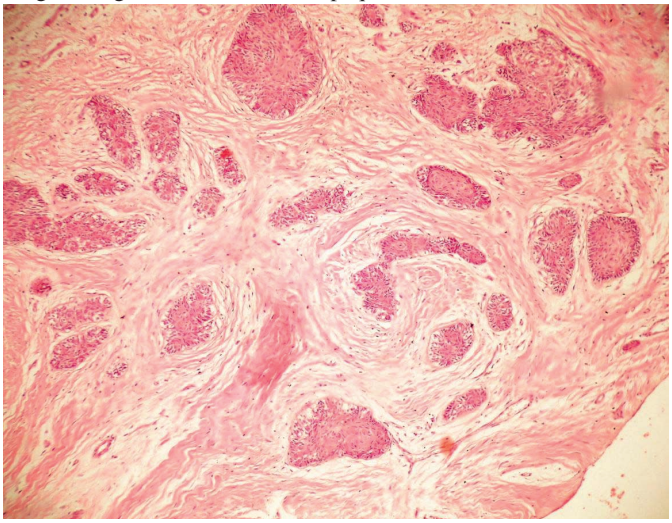

Fig. 4. Proliferation of small odontogenic epithelium islands and cords interlarded by a dense fibrous stroma, exhibiting intense collagenized areas (HE 100x).

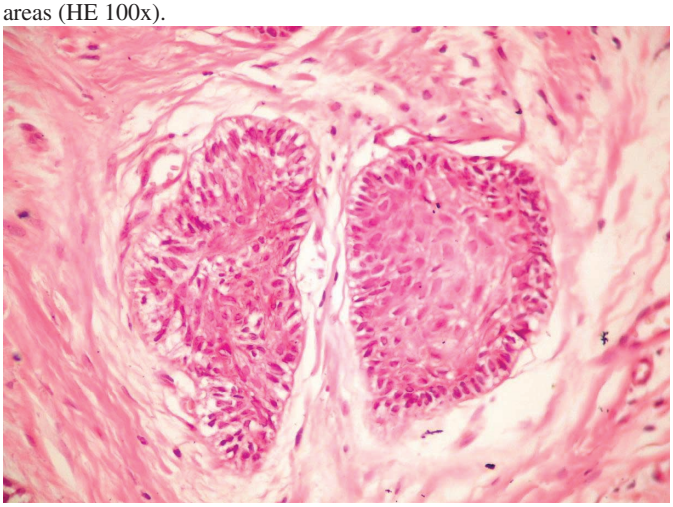

Fig. 5. Islands of odontogenic epithelium exhibiting peripheral cells arranged in palisade with hyperchromatism and cores of reverse polarity. In the center of the islands, loosely organized cells were found, resembling the stellate reticulum of the enamel organ (HE 400 x).

## DISCUSSION

The desmoplasic ameloblastoma was firstly described by Eversole *et al*. (1984), who described three cases (Hirota *et al.*) It is a rare entity that exhibits important differences in its anatomical, histological and radiographic distribution when compared to other kinds of ameloblastoma. Thus, by presenting its own clinic and radiographic characteristics, the desmoplasic ameloblastoma has been considered by some authors as a distinct clinic-pathological entity. This was recently confirmed by the World Health Organization, in 2005, whose classification organized the ameloblastomas in solid, extra-osseous, desmoplasic and unicystic (Phillipsen *et al.*, 1992; Dos Santos *et al.*).

The literature shows the desmoplasic ameloblastoma as the least frequent odontogenic tumor. Among 89 ameloblastoma cases studied by Takata *et al*.(1999), seven (7.9%) were diagnosed as desmoplasic, and in another study of 163 ameloblastoma cases done by Ledesma-Montes *et al.* in the Latin American region, two were desmoplasic. According to Adebiyi *et al*., from 77 cases of ameloblastomas diagnosed within the Nigerian population, four were the cases diagnosed as desmoplasic.

More than 70% of the desmoplasic ameloblastoma cases happen in the anterior region of the maxilla (Kaffe *et al.*, Manuel *et al*.), the opposite from the conventional ameloblastomas, which are usually found in the posterior region of the mandible. In 15 cases of desmoplasic ameloblastoma analyzed by Kaffe *et al*., 11 lesions were located in the maxilla (73%) and only four in the mandible (27%), with most of the cases reaching the anterior region. These findings partially corroborate with the clinical case presented here, since the lesion was located in the posterior region of the maxilla. However, in another study of ten desmoplasic ameloblastoma cases reported by Kishino *et al.* (2001), 40% involved the anterior region of the maxilla and 60%, the posterior region of the mandible. On a literature review done by Desai *et al.,* from the 89 cases studied, the desmoplasic ameloblastoma's occurrence predilection is in the anterior region of the jaw bones, being the mandible the most common one (57.3%) when compared to the maxilla (42.7%).

The clinical aspects presented in this case are in agreement to the characteristic reports presented on the literature, with a proportional attack happening

between men and women, specially within the third and fifth life decade (Manuel *et al.*). To Kaffe *et al.*, the lesions usually attack the female gender most in a 2:1 proportion. However, in the 89 cases reviewed by Desai *et al.*, 54.66% involved the male gender and 45.33% the female one. Frequently, the tumor is asymptomatic, the tumefaction is painless or the maxilla expansion are commonly observed clinical manifestations (Neville *et al*.) In the case reported here, the patient had a painful symptomatology and areas of ulceration, probably because of the size of the lesion (Becelli *et al*.) and the trauma during mastication.

The radiographic findings, observed in the case mentioned above, are typical on the desmoplasic ameloblastoma, presenting itself as a unilocular radiolucent lesion of indefinite outlines. Discreet radiopaque focuses could be observed inside the lesion through an occlusal and panoramic radiography, characteristic observed in more than 50% of the cases, imitating many times a fibro-osseous lesion due to its radiolucent-radiopaque mixed appearance (Kawai *et al.*, 1999). The differential diagnosis according to the radiographic aspects include fiber-osseous lesions, fiber dysplasia, chronic osteomyelitis and ossifying fibroma (Iida *et al*.)

The histopathological characteristics described for this case are in accordance to the diagnosis criteria established for the desmoplasic ameloblastoma according to Kishino *et al.* This way, such lesion is characterized by small nests and cords of odontogenic tumoral epithelium, organized in an abundant stroma densely collagenized, which makes these tumoral islands seem like they are comprised (Dos Santos *et al.*, Hirota *et al*.) The peripheral cells can lose, sometimes, those morphologic characteristics similar to the ameloblastomas such as the polarity inversion. In addition to this, the central portion of the tumoral islands can present loosely organized cells resembling the stellate reticulum of the enamel organ, with occasional squamous metaplasia and central focuses of keratination (Takata *et al*.) In the desmoplasic variation, usually, there can be verified an osseous neo-formation that according to Thompson *et al*.(1996), represents an attempt to fix the damages caused by the osseous re-absorption caused by the tumoral expansion. In the case above, such osseous formations were not evidenced in the epithelial proliferation.

The most important characteristic of the desmoplasic ameloblastoma is the extensive collagenization present in the stroma of the lesion, also called desmoplasia. It has been proposed that this phenomenon has its origin from a new protein synthesis that comes from the extracellular matrix where such components play a fundamental role in supporting, adhering, proliferating, migrating and differing tumoral cells, interfering in the behavior and modulation of the tumoral cells (Desai *et al*.; Phillipsen *et al.*, 2001).

According to Philippsen *et al.* (1992), radiographically, the badly-defined lesion borders suggest an infiltrated process that tends to be recurrent. The significant stromal reaction can be seen as a reply from the host trying to make the lesion stop growing. The recurrence level of the desmoplasic variant is still not known due to the low number of reported cases in the literature. However, similar recurrence has been seen with other types of ameloblastomas. Due to the lack of capsule and precise limits, a radical treatment is recommended for most of the cases (Ashman *et al.,* 1993) However, because of the limited understanding on the biological and prognostic behavior, the therapeutical approach strategy is still not completely clear.

## CONCLUSION

The desmoplasic ameloblastoma is a rare entity that presents a radiographic appearance that resembles a fibro-osseous lesion that needs a diagnosis based not only on clinical and radiographic aspects, but also on those histopathological ones. However, the importance of knowing the ameloblastomas' histological and radiographic variants in relation to possible differential diagnosis and prognosis to confirm and, consequently, apply proper therapy is highlighted.

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**RESUMEN:** El ameloblastoma es una neoplasia benigna de origen epitelial odontogénica de crecimiento lento, localmente invasiva y constituye el tumor odontogénico más común en los maxilares. Histológicamente, existen varios tipos descritos en la literatura. La variante desmoplástica es rara y se caracteriza por diferencias en los hallazgos típicos del ameloblastoma, incluso la localización y aspectos radiográficos e histológicos. Los objetivos de este artículo son relatar un caso de ameloblastoma desmoplásico en el lado izquierdo del maxilar superior y presentar una revisión de la literatura relevante, enfatizando aspectos peculiares de esta rara lesión.

**PALABRAS CLAVE: Ameloblastoma; Ameloblastoma desmoplásico; Tumores odontogénicos.**

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