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DESMOPLASTIC AMELOBLASTOMA IN THE MANDIBLE: A CASE REPORT

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MANDİBULADA DESMOPLASTİK AMELOBLASTOMA: VAKA RAPORU

ÖZET

Ameloblastomalar çenelerin en sık gözlenen neoplazmlarıdır. Desmoplastik Ameloblastoma (DA) tüm ameloblastomaların % 4-13'ünü oluşturur.

53 yaşında bir erkek hasta, sol mandibulasında ağrısız bir şişlik şikayeti ile kliniğimize başvurdu. Hastanın ağız içi klinik muayenesinde sol mandibular birinci premolar hizasından arkaya doğru uzanan, hem bukkal hem de linguali kaplayan sert bir şişlik saptandı. Radyografik muayenede irregüler radyoopak ve radyolusent alanlar gözlendi. Çıkarılan spesmen, histopatolojik olarak desmoplastik ameloblastoma olarak yorumlandı.

Sunulan bu vaka son zamanlarda tanımlanması ve yayınlanan vakaların sayılarının nispeten az olması nedeniyle varolan literatür eşliğinde tartışılmıştır.

Anahtar Kelimeler: Desmoplastik Ameloblastoma, Odontogenik tümör, Maksilla, Mandibula.

ABSTRACT

Ameloblastomas are the most common neoplasms of the jaws. Desmoplastic Ameloblastoma (DA) comprises from 4 to 13 % of all ameloblastomas.

A 53 year-old man was referred to our clinic with a complaint of painless swelling on the left mandible. Intraoral clinical examination revealed a bony hard swelling extending posteriorly from the left mandibular first premolar. The lesion extended both buccaly and lingually. Irregular radiopaque and radiolucent areas were seen on radiographic examination. The removed specimen was diagnosed as desmoplastic ameloblastoma on histopathological grounds.

Because this variant has only recently been recognized and the number of published cases is still relatively few, the case is presented and discussed with the available literature.

Keywords: Desmoplastic Ameloblastoma, Odontogenic tumour, Maxilla, Mandible.

INTRODUCTION

Ameloblastomas are the most common neoplasms of the jaws. They are usually first recognised between the ages of 30 and 50. They are rare in children and old people. Eighty per cent form in the mandible; of these, 70% develop in the posterior molar region, and often involve the ramus. They are symptomless until the swelling becomes obtrusive.[1]

Ameloblastomas are separated into solid or multicystic (the classic intraosseous ameloblastoma), unicystic, and peripheral types.[2,3] In general, the histopathology of the classic intraosseous ameloblastoma is well known. The numerous histologic variants that have been described in the literature have similar clinical characteristics.[4] There are, however, two recent developments that warrant discussion, namely the recognition of the desmoplastic ameloblastoma (DA) and of the

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clear cell ameloblastoma. DA comprises from 4 to 13 % [5-7] of all ameloblastomas. Ng and Siar[7] reported a 3% rate for DA, when based on all odontogenic tumours. The histologic variant now referred to as DA is not new, but little attention was paid to it until Eversole et al.[8] described it in detail in 1984.

A case of mandibular DA is presented as a rare variant of ameloblastoma in this paper.

REPORT OF THE CASE

A 53 year-old man was referred to the Department of Oral and Maxillofacial Surgery, Dental Faculty of Ondokuz Mayıs University, Samsun, because of painless swelling on the left mandible in the end of December 1997. Intraoral clinical examination revealed a bony hard swelling extending posteriorly from the left first premolar. The lesion extended both buccally and lingually. Irregular radiopaque and radiolucent areas were seen on radiographic examination (Figure 1). The lesion was 2 cm at the greatest diameter. An incisional biopsy led to the diagnosis of desmoplastic ameloblastoma. The tumour was removed with large margins under local anesthesia after six months than incisional biopsy. The specimen was examined histopathologically, confirming the first diagnosis. The tumour consisted predominantly of dense collagenous fibrous tissue in which there were small, irregular islands of neoplastic epithelium. There was little cyst formation, and ameloblast like cells were present in small foci on the periphery of some islands of epitelium. There was scarce stellate reticulum like tissue in the interior of the epitelial islands. Squamous metaplasia and foci of keratinization were seen centrally within the ameloblastic epithelial islands. (Figure 2 a,b) No calcification in the fibrous stroma was seen. But also there was unresolved bone spicule. This finding was confirmed irregular radiopaque area on radiographic illustration.

Bony healing was observed on radiographic examination postoperatively (Figure 3). No recurrence was noted during the postoperative long term period.

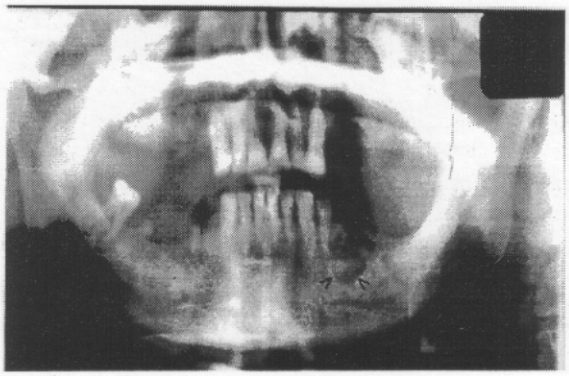

Figure 1. Irregular radiopac and reduolucent areas (arrows) were seen on preoperative radiographic examination

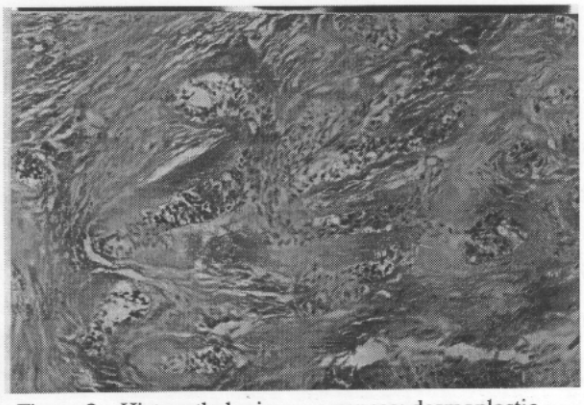

Figure 2a. Histopathologic appearances; desmoplastic stroma are seen.

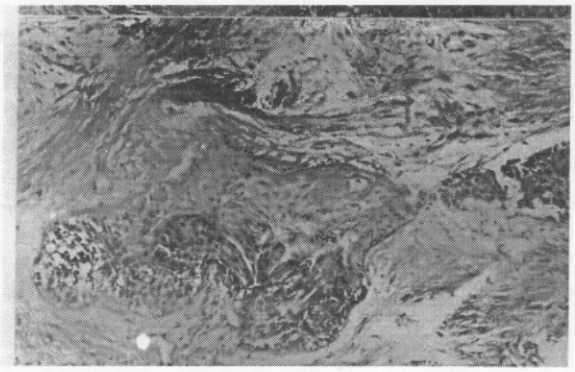

Figure 2b. Histopathologic appearances; together with ameloblastic islands are seen

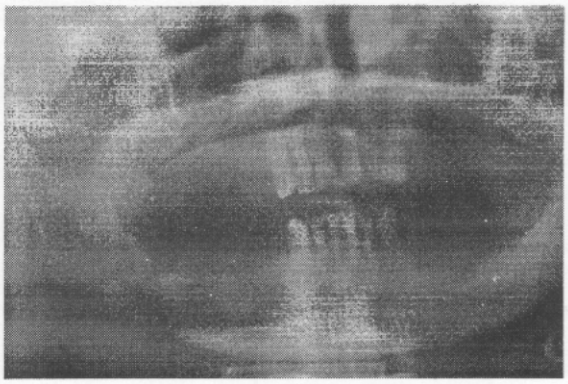

Figure 3. Bony healing was seen on radiographic examination after postoperative 5 month.

DISCUSSION

Two reports from Japan, the first in 1981[9] and the second in the 1983,[10] first called attention to an unusual variant of the intraosseous ameloblastoma. The tumour was reported to be characterized by an unusual histomorphology, including extensive stromal collagenization or desmoplasia.[6,11,12] DA was recognized as a benign, locally invasive variant of the intraosseous, infiltrative ameloblastoma (IA).[13] Literature review revealed that DA accounted for 1.4 and 13% of ameloblastomas.[5-7,13,14]

DA consists predominantly of dense collagenous fibrous tissue in which there are small, irregular islands of neoplastic epithelium. These islands are rounded or angular, and may have slender straggling extensions. There is little or no cyst formation, and ameloblast like cells are typically only present in small foci on the periphery of some islands of epitelium.[1,15,16] The center of the epithelial islands may appear hypercellular with spindle shaped or squamatoid epithelial cells.[15] Stellate reticulum like tissue is also absent from the interior of the epitelial islands, which consist of densely packed spindle shaped or polygonal cells.[1,13,16] However, The occasional large tumour islands are often very irregular in shape with a pointed, stellate or kite like appearance.[15] Squamous metaplasia or foci of keratinization may occasionally be seen centrally, and this variant may be difficult to recognize as an ameloblastoma.[1] Microcysts containing eosinophilic amorphous deposits or appearing empty are commonly found within the tumour island.[13] Calcification in the fibrous stroma and, occasionally, bone formation is seen1. True glandular differentiation with mucus cells has been described in tumour nests.[17] In keeping with the literature in the case presented characteristic features of DA variant of ameloblastoma were found to be dense collagenous desmoplastic stroma with squamous metaplasia and foci of keratinization located centrally within the ameloblastic epithelial islands, with little cyst formation.

A comparative immunohistochemical study showed that the connective tissue stroma in DA contrary to that of IA, exhibited a strong positive reaction for collagen type VI.[13] This was interpreted as indicating an active de novo

synthesis of extracellular matrix protein. In other words, the desmoplastic stroma of DA was interpreted as not a scar tissue, but newly produced connective tissue. In contrast to IA, marked immunoexpression of transforming growth factor (TGF-≤3) was observed in six out of seven cases of DA.[17] It was suggested that TGF-≤3 produced by DA tumour cells played a part in the prominent desmoplastic matrix formation.[13] However, the mechanism of desmoplasia is far from being understood. Myxoid changes of the stroma may be observed surrounding the odontogenic epithelium. Oxytalan fibres have been identified in the stromal tissue of one case reported by Kawai et al.[15] This finding was interpreted as indicating a tumour derivation from the epithelial rests of Malassez in the periodontal membrane of a related tooth[13,15] Although formation of metaplastic bone trabeculae (osteoplasia) rimmed by active osteoblasts has been described in several cases,[13] no bone formation was observed but unresolved bone spicule was seen in the present case.

As the predominant structure was desmoplastic, the presence of little cyst formation with scarce stellate reticulum like tissue observed in the present case was not regarded as characteristic of either follicular or plexiform ameloblastoma or a hybrid form of the either. A possible "transitional" form of the DA, showing microscopic features of the desmoplastic variant together with areas typical of classical follicular or plexiform ameloblastoma, were described as a "hybrid" lesion of ameloblastoma.[5,11,12] The hybrid lesion of ameloblastoma (HLA) was first described by Waldron and El-Mofty[5] and is a tumour variant where histologically, areas of follicular or plexiform ameloblastoma coexists with areas characteristic of DA. It has been inconclusive to speculate whether desmoplastic changes occur secondary in the stroma of a pre-existing IA, or whether areas of primary DA transform into an IA.

The DA involves the anterior maxilla predominantly, which is an unusual location for ameloblastoma although it also involves the anterior mandible quite commonly, and presents a radiographic appearance that is more typical of a benign fibroosseous lesion than of ameloblastoma.[18] In the case presented, however, the tumour located in the premolar region of the mandible. Further research is needed on the etiology of DA.

Although the biological behaviour of the DA still remains unresolved, it is generally agreed that the tumour is a variant of the intraosseously located ameloblastoma. A painless swelling represents the chief initial complaint in most cases,[13] including the present case. The size of the tumour varies between 1.0 and 8.5 cm at the greatest diameter,[13] as the case with ours. A true peripheral variant of the DA without bone involvement and, thus, similar to the peripheral ameloblastoma has not been reported so far.[13] DA in the present case is an intraosseous tumour, in keeping with the literature.

The radiographic features of DA differ in most cases from those of IA.The radiographic features of the conventional, intraosseous ameloblastoma are classically described as uni-or multilocular radiolucencies with relatively well defined borders including 7 % of DA cases.[13] The content of the lesion is mixed radiolucent/ radiopaque in half of the cases (53%)[13] as was the case with ours. Thus, in many cases the

preoperative radiographic diagnosis alone could be misleading, for a fibro osseous lesion.[13] The fact that new bone formation has been reported in several cases of DA may explain the mixed radiolucent/radiopaque appearance.[11,19] On the other and, Takata et al.[17] explained that the mixed radiological appearance expressed the infiltrative pattern of the tumour, for when DA infiltrated the bone marrow space, remnants of the original, nonmetaplastic or nonneoplastic bone could remain in the tumour tissue. In conclusion, the infiltrative behaviour of DA seems to explain one of the characteristic features of this tumour, the ill defined border. An association between DA and an unerupted or impacted tooth has been found in only three cases (3.4%) so far 6 as against 8.7% among IA.[14] In the case presented there was no tooth impaction associated with the tumour.

The age range of patients with DA varies between 17 and 72 years.[13] Similarly, the presented patient was 53 years old.

No recurrence was seen in the present case after three years and seven months' follow-up. The biological behaviour of the DA including recurrence rate is difficult to estimate due to the relatively few reported cases with sufficiently long follow-up periods. According to the WHO classification it is stated that unicystic, peripheral and possibly desmoplastic ameloblastoma have lower recurrence rates than other ameloblastomas. The radiological and histological findings of poor encapsulation or total lack of a capsule requires a long term follow-up of the potential for recurrence of the DA variant. The authors agree with the general literature[1,2,13,18] in that same radical treatment modality for the DA should be followed as that of the classical ameloblastoma (IA). Many more cases with detailed clinical, radiological data and corresponding histopathologic analysis are needed to clarify the biological behaviour of the DA variant.

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