

Case Report

Hybrid Ameloblastoma: Report of a Rare Case and Review of Literature

Aakriti Gupta, Chhavi Jindal

Abstract

Synchronized existence of desmoplastic ameloblastoma with follicular ameloblastoma is a rare event. Although reported by many authors, it was first described as a separate entity in 1987 by Waldron and El-Mofty under the heading hybrid ameloblastoma. Here we report a case of hybrid ameloblastoma showing unusual squamous metaplasia. The patient, a 35 year old, Indian woman, had an irregular swelling in lower right front jaw region. Radiographically, a mixed radioopaque / radiolucent lesion were seen in the region of right mandibular lateral incisor to second premolar. Histologically, the lesion was characterized by extensive stromal colonization surrounding compressed islands of odontogenic epithelium along with areas of follicular ameloblastoma. Some follicles showed squamous metaplasia. To best of our knowledge, 25 cases of hybrid ameloblastoma have been reported to date, and the present case is one more addition for the better understanding of this variant of odontogenic tumor.

Keywords: Odontogenic Neoplasms; Ameloblastoma; Hybrid; Desmoplastic; Follicular ameloblastoma; Squamous Metaplasia.

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Introduction

Ameloblastoma is a true neoplasm of enamel-type organ tissue that does not undergo differentiation to the point of enamel formation. It was very aptly described as being a tumour that is “usually unicentric, non-functional, intermittent in growth, anatomically benign and clinically persistent”. It arises from dental embryonic remnants (possibly the epithelial lining of the odontogenic cyst), dental lamina or enamel organ, stratified squamous epithelium of the oral cavity, or displaced epithelial remnants.^{1,2}

Histopathologically, ameloblastoma exhibit proliferating odontogenic epithelium within a background of fibrous stroma. It includes several clinical, radiological and histological (follicular, plexiform, acanthomatous, basaloid, granular and desmoplastic) subtypes.² Desmoplastic ameloblastoma was first described in detail by Eversole et al in 1984 and is defined as “a variant of ameloblastoma with specific clinical, imaging and histological features”. Since, it often occurs in the anterior region of jaws, it presents with unique radiographic appearance resembling fibrous lesions and show distinct histopathology characterized by extensive stromal collagenisation or desmoplasia surrounding compressed islands of odontogenic epithelium.¹⁻³ A possible transitional form of

desmoplastic ameloblastoma, showing microscopic features of desmoplastic variant together with areas of classical follicular / plexiform ameloblastoma has been described as a “hybrid lesion”. It is an extremely unusual variant and was first described in detail by Waldron & El-Mofty in 1987.^{1,4}

Here we describe a rare case of hybrid ameloblastoma in a 35 years old woman with details of clinical, radiographic and unusual histologic features suggesting it to be a hybrid ameloblastoma. A comprehensive review of English language literature revealed 25 cases of hybrid ameloblastoma with no recurrence of the lesion in any of the cases. Here, we add an additional case to enhance the knowledge of this interesting tumor.

Case Report

A 35 year old female patient presented with the chief complaint of swelling in lower right front jaw region since 4 months. Initially the swelling was small in size then it gradually increased up to the present size. There was no history of any pain or pus discharge. Past medical and dental history was non-contributory.

Extraoral examination revealed an irregular swelling in relation to the body of mandible on right side extending antero-posteriorly

from midline to angle of mandible and supero-inferiorly from angle of mouth and involving the lower border of mandible. The overlying skin was smooth and normal in colour. The swelling was non tender, firm, non fluctuant, non compressible and fixed to underlying structures. Submandibular lymph nodes of the affected side were non tender and non palpable. Intraoral examination revealed a single bony hard swelling measuring about 6 x 4 cm in size with ill-defined margins, extending antero-posteriorly from lower right lateral incisor to lower right second premolar and supero-inferiorly from attached gingiva of 43, 44, 45 obliterating the right buccal vestibule. The overlying mucosa was normal. Associated teeth were vital, 46 was found to be missing.

Orthopantomogram revealed an ill-defined hazy, radiolucent lesion in the right body of the mandible with flecks of radio-opacity extending from mandibular right lateral incisor to second premolar with displacement of associated roots. There was involvement of the lamina dura and periodontal spaces of associated teeth (Fig-1).

An incisional biopsy under local anesthesia was done and the specimen was received for histopathologic examination. The specimen was creamish white to brown in colour, measuring about 6x3x2 cm in size. The specimen was hard in consistency in some areas while soft in other areas (Fig 2).

Histopathologically, the lesion consisted of numerous ameloblastic islands dispersed in a densely fibrous stroma exhibiting areas of myxoid degeneration. These epithelial islands comprise of a peripheral layer of cuboidal or columnar cells resembling internal dental epithelium or preameloblasts. The central mass of cells comprise of angular cells like stellate reticulum which show squamous metaplasia, at many places. Also cystic degeneration is found in the epithelial islands. The islands appeared small and compressed reminiscent of desmoplastic ameloblastoma. There are areas showing marked hyalinization (desmoplasia) of the connective tissue stroma (Fig 3). The epithelial cells at the periphery of islands are cuboidal, with hyperchromatic nuclei and stellate reticulum or "kite-like" cells in the center showing squamous metaplasia (Fig 3, 4, 5). The overall features were suggestive of a hybrid form of desmoplastic ameloblastoma as both

typical desmoplastic areas with follicular areas were seen. In such cases the management should be done by wide resection followed by artificial reconstruction of the mandible.

Discussion

Ameloblastoma is a rare odontogenic tumor accounting to around 1% of all the cysts and tumors in the jaws. Follicular and plexiform are the commonly encountered variants accounting to 32.5% and 28.2% respectively; followed by the acanthomatous subtype 12.1% while desmoplastic is extremely uncommon with incidence rates ranging from 4-13%.^{1,5} Desmoplastic Ameloblastoma is a benign, locally infiltrative epithelial neoplasm consisting of proliferating, irregular, often bizarrely shaped islands and cords of odontogenic epithelium of varying sizes embedded in a desmoplastic, connective tissue stroma.⁶⁻⁸ Since the first description of desmoplastic ameloblastoma a total of 115 cases have been reported. Waldron and El-Mofty described a hybrid variant of desmoplastic ameloblastoma with unusual features in 1987. To best of our knowledge, a total of 25 cases of hybrid variant of desmoplastic ameloblastoma have been published so far. The present case adds one more case to the literature of hybrid variant of desmoplastic ameloblastoma showing unusual acanthomatous follicles surrounded by desmoplasia.

Desai et al in 2006 reviewed 90 cases of desmoplastic ameloblastoma, the lesion was commonly seen from 3rd - 5th decade of life, males (45.55%) were more affected than females (37.77%).⁶ The reported cases of hybrid desmoplastic ameloblastoma shows slight female predilection with M:F ratio 11:16. These occurred in a wide age range of 17-82 years.^{9,10} The present case is a 35 year old female patient.

In the study by Desai et al tumor location was mentioned in 58 of 90 cases. Tumor was found to be most extensively involving the anterior premolar and molar regions of maxilla as compared to mandible.⁶ Reported cases of hybrid desmoplastic ameloblastoma occur more often in mandible than in maxilla with a ratio of 5:3.^{11,12} In our case, the lesion was present in mandibular anterior region and swelling was the chief complaint of the patient.

Radiologically, desmoplastic ameloblastoma present itself as a mixed radiolucency with mixed radiopacities and demonstrates ill-defined borders. Other radiographic features reported are root resorption and displacement of teeth.^{13,14} The present case showed an ill-defined hazy, radiolucent lesion in the right body of the mandible with flecks of radio-opacity extending from mandibular right lateral incisor to second premolar.



Figure 1: Orthopantomogram revealed an ill-defined hazy, mixed (radiopaque / radiolucent) lesion extending from mandibular right lateral incisor to second premolar with displacement of associated roots.



Figure 2: Cut surface of the gross specimen demonstrating a hard and soft, gritty, creamish white to brown lesion

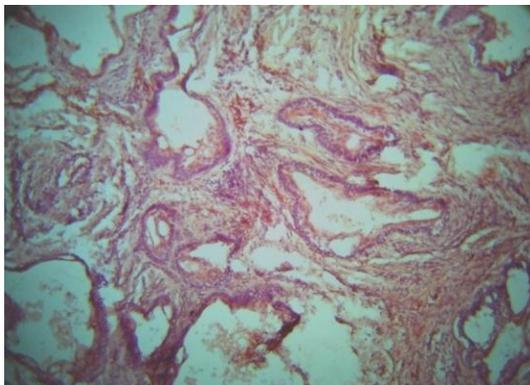


Figure 3: Odontogenic epithelium in the form of follicles and the irregular compressed epithelial islands that are typical of

desmoplastic ameloblastoma (hematoxylin-eosin stain, 4X).

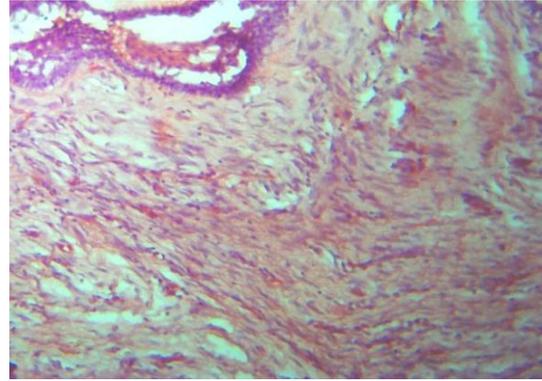


Figure 4: The Photomicrograph shows dense hyalinization in the connective tissue stroma (hematoxylin-eosin stain, 10X).

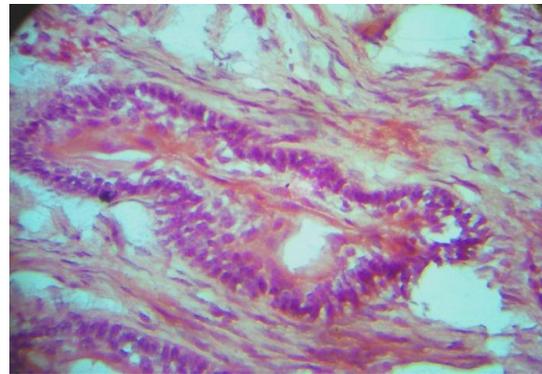


Figure 5: The epithelial cells at the periphery of islands are cuboidal, with hyperchromatic nuclei and stellate reticulum cells in the center showing squamous metaplasia (hematoxylin-eosin stain, 40X).

Histopathologically, hybrid ameloblastoma show areas of compressed odontogenic epithelial islands surrounded by dense desmoplasia along with areas of typical follicular / plexiform / acanthomatous ameloblastoma.^{6,15-17} Some cases of hybrid ameloblastoma also showed granular cell transformation in some of the tumor cells along with areas of follicular and plexiform ameloblastoma while some showed basaloid changes also. Six cases in literature have been reported with extensive osteoplasia.¹⁸ As the present case gives a picture of desmoplastic and follicular ameloblastoma with squamous metaplasia, it can be considered as a case of 'Hybrid Ameloblastoma'.^{7,12}

Whether, the hybrid ameloblastoma should be considered as a hybrid tumor or collision tumor is a matter of debate since its existence. While collision tumors are considered as two lesions arising from

independent topographic sites, a tumor is considered to be hybrid if two or more disparate and well established tumors exhibit obvious differentiation.^{19,20} Hybrid variant of desmoplastic ameloblastoma shares common clinical characteristic features with typical desmoplastic ameloblastoma i.e. no definite gender prevalence, site predilection of anterior region of jaws and mixed radiolucent-radiopaque appearance, except for the histologic presence of areas of conventional ameloblastoma along with areas of desmoplastic ameloblastoma with a striking mandibular predilection pointing towards differentiation concept.^{8,9} But some authors believe that designation of “hybrid tumor” serves no real purpose and if taken, literally, might overstate the significance of finding a desmoplastic ameloblastoma in combination with islands of solid multicystic ameloblastoma (SMA). It might be that areas of conventional ameloblastoma may in fact be a part of histopathologic spectrum of desmoplastic ameloblastoma, hence favoring the collision concept. The debate still continues as more data is required to reach to the final conclusion.^{10,21}

Desmoplastic ameloblastoma should be differentiated from other odontogenic tumors and fibro-osseous lesions. Radiographically, desmoplastic ameloblastoma shows a mixed radiolucent-radiopaque picture which is different from the radiographic view of squamous odontogenic tumor, which comprise of well-defined unilocular and triangular radiolucency between the roots of adjacent teeth with no radiopacity. Ossifying fibroma consists of bone/osseous tissue as the major component histologically which is not similar for desmoplastic ameloblastoma where epithelial islands are found in the connective tissue stroma.¹ The histological picture of Focal cement-osseous dysplasia is also different from desmoplastic ameloblastoma as it is characterized by thick, curved/linear bone trabeculae (ginger root pattern) or irregular cementum like masses that are not seen in desmoplastic ameloblastoma. The odontogenic fibroma is a benign neoplasm composed of cellular connective tissue. It often occurs in fibroblastic strands that are interwoven with less cellular areas in which numerous small blood vessels are present. Foci of calcified collagenous matrix, resembling dysplastic cementum, osteoid or atubular dysplastic dentin often occur which is again not similar to the histopathologic picture of

desmoplastic ameloblastoma.¹¹ Ameloblastic fibroma histologically is composed of proliferating odontogenic epithelium embedded in a cellular ectomesenchymal tissue that resembles the dental papilla, and with varying degrees of inductive change and dental hard tissue formation. This is not similar to the components seen in desmoplastic ameloblastoma. Thus, we may say that a typical mixed radiolucent-radiopaque lesion occurring in anterior region of jaw and with the histologic features of epithelial islands that are small and compressed in the connective tissue stroma may help in concluding it to be a desmoplastic ameloblastoma.^{7,10,11} Taking into account the patient's complaint, clinical features, radiographic and histopathologic findings, diagnosis of hybrid variant of desmoplastic ameloblastoma was made.

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