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CASE REPORT

How to name it: a rare case of odontogenic cyst

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Odontogenic cysts and tumors are well-recognized entities to the specialist oral pathologist and they seldom pose problems in differential diagnosis. This paper deals with an aggressive cystic lesion in the maxilla of a 65-year-old male that was characterized by a large radiographically multilocular lesion and a multicystic pattern microscopically. The categorization of this lesion was complicated by the presence of features suggestive of both glandular odontogenic cyst and cystic ameloblastoma with aggressive histologic phenotypes.
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Case report

A 65-year-old male presented with the complaints of swelling and pain in the right upper jaw. He stated that the swelling was present without any discomfort for more than a year. Further questioning of the patient showed that he had been evaluated for the swelling by a general dental practitioner 6 months earlier, and was told that the swelling was secondary to an odontogenic infection originating from the maxillary teeth. Subsequently, few maxillary teeth were extracted, after a course of antibiotics and analgesics. He also stated that the remaining maxillary teeth had spontaneously exfoliated and reported that the pain developed only 15 days before his visit here. His medical history and systemic examination was non-contributory.

No unusual features or lympadenopathy were noted on extra oral examination except for the obliteration of the right nasolabial sulcus. Intra-orally, the right upper quadrant was edentulous. A well-defined swelling with buccolingual expansion was seen extending from right maxillary third molar to left maxillary lateral incisor region (Fig. 1). The mucosa over the swelling appeared normal. Palpation indicated a firm swelling with cystic areas in the posterior region. No thrills were palpated, and no bruit was heard. The rest of the examination was not significant.

Radiological examination included occlusal, Waters view of the skull and panoramic radiographs. Occlusal radiograph of the maxilla showed multilocular radiolucency. Two distinct loculi in relation to the right maxillary central and lateral incisor, and in the region of the left maxillary lateral incisor showed thin corticated border. The third locule was large and seen extending from the right maxillary central incisor to the right maxillary first molar region. The lesion had a honeycomb appearance in the region of the left maxillary central incisor with the destruction of the buccal cortical plate and the alveolar bone in the region of the right maxillary second molar to the left maxillary central incisor (Fig. 2A). Waters view of the skull showed haziness in the right maxillary antrum. The lateral wall of the right maxillary antrum and right infra orbital margins were intact. Soft tissue radio opacity was seen in the right alveolar ridge (Fig. 2B). Panoramic radiograph showed the destruction of the alveolar bone extending from the right maxillary third molar to the left maxillary central incisor with multilocular radiolucency extending from the right maxillary central incisor to the left maxillary canine. A large locule was seen extending from the edentulous right maxillary third molar to the canine region with the upper corticated margin of the locule extending up to the maxillary sinus (Fig. 2C).

Radiographically, the lesion can be described as a multilocular radiolucency with a soap bubble appearance posteriorly and honeycomb appearance in the region of the right and left maxillary central incisors. Based on the clinical and radiological information available, our differential diagnosis included ameloblastoma, odontogenic keratocyst and central hemangioma. Aspiration was attempted with a wide bore needle but did not yield any positive result.

Under local anesthesia, an incisional biopsy was performed. Reflection of a full thickness mucoperiosteal flap in the posterior region of the lesion demonstrated a very thin softened cortical bone covering the lesion. The

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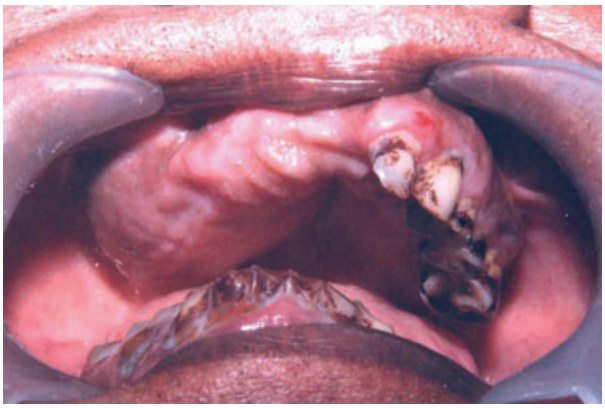

Figure 1 Buccal and palatal expansion.

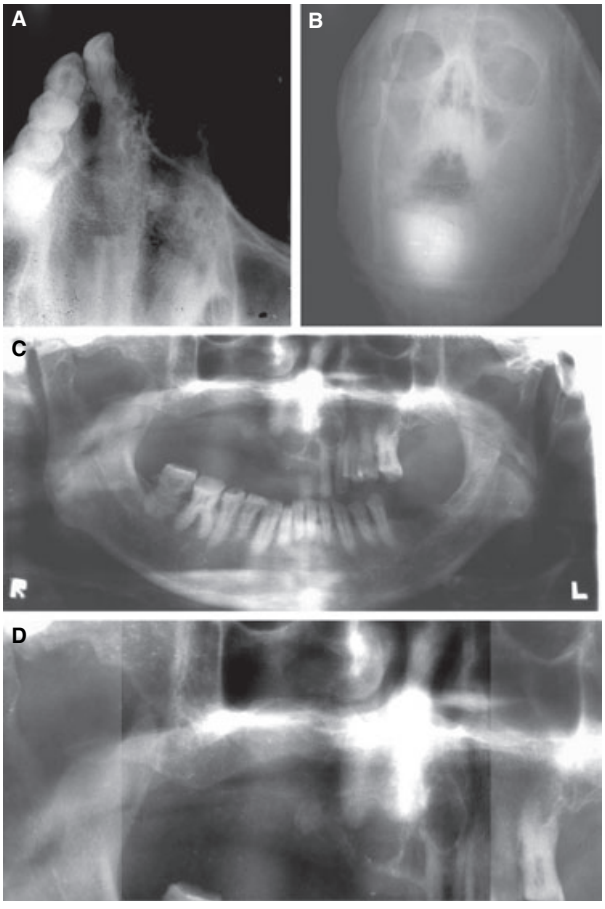

Figure 2 (A) Multilocular radiolucency with honeycomb appearance in relation to left maxillary central incisor with destruction of buccal cortical plate and alveolar bone. (B) Haziness in right maxillary sinus indicates mucosal thickening. (C) Multilocular radiolucency with corticated margin and destruction of alveolar bone. (D) Magnified image of the lesional area in (C).

cortical bone was penetrated and a wedge of cystic lining was obtained and studied microscopically.

Histopathological examination showed a cystic lesion lined in part by a non-keratinized stratified squamous epithelium of variable thickness with hyperchromatic and vacuolated basal cells. The underlying fibrous wall demonstrated odontogenic islands some of which are found associated with peripheral nerve fibres, and closely resembled squamous odontogenic proliferation but were lined by a peripheral layer of columnar cells (Fig. 3). A non-committal diagnosis of 'odontogenic cyst with mural ameloblastoma' was rendered.

As the pathology report was not confirmative of a specific diagnosis, the patient was further evaluated with a computed tomography (CT) scan to determine the extent of the disease and to plan surgery. The computed tomography of maxilla showed a soft tissue homodense lesion of 36–45 Hounsfield unit (HU) in the right maxillary region, multiple loculi in the anterior region with scalloped border on the palatal aspect, and destruction of buccal cortical plate. The CT also revealed destruction of the floor of the maxillary sinus and nasal cavity with an extension of the lesion into the nasal cavity and maxillary sinus (Fig. 4).

Under general anesthesia, partial maxillectomy was performed on the right side, the resected margin extended up to left maxillary canine. The maxillary sinus lining was also thoroughly curetted. During surgery, three to four small loculi were noted in the anterior region corresponding to the right maxillary central incisor to the left maxillary canine, and posteriorly, a large cystic cavity was encountered which had completely eroded the buccal cortex and palatal bone. An acrylic obturator was placed to cover the raw wound.

The post-surgical specimen was studied with blocks made from the anterior and posterior regions of the specimen.

An histopathological examination of the sections made from the posterior part of the lesion showed a multicystic lesion lined in part by a non-specific stratified epithelium with hyperchromatism and vacuolated basal cells. Characteristically, the lining appeared to be fragile and detached with focal thickenings (Fig. 5). In some areas, the epithelium was stratified without any distinguishing feature but lacked polarization. Odontogenic islands showing signs of ameloblastomatous differentiation and dystrophic calcification were found in the densely homogenized connective tissue (Fig. 6). An examination of the sections from the anterior part of the lesion that corresponds to the radiograpically detectable multilocular region (Fig. 7) showed multiple macro- and microcystic cavities lined in part by ameloblast-like cells. Epithelial thickening and downgrowths surrounded by juxtaepithelial hyalinization were also observed (Fig. 8). Neural invasion and encroachment of minor salivary gland were also noted (Fig. 9).

Comments

Odontogenic islands, with or without features of early ameloblastoma as defined by Vickers and Gorlin (1) are not unusual in jaw cysts. Features of early ameloblastoma are often seen in the cystic lining of dentigerous cyst, calcifying odontogenic cyst, and

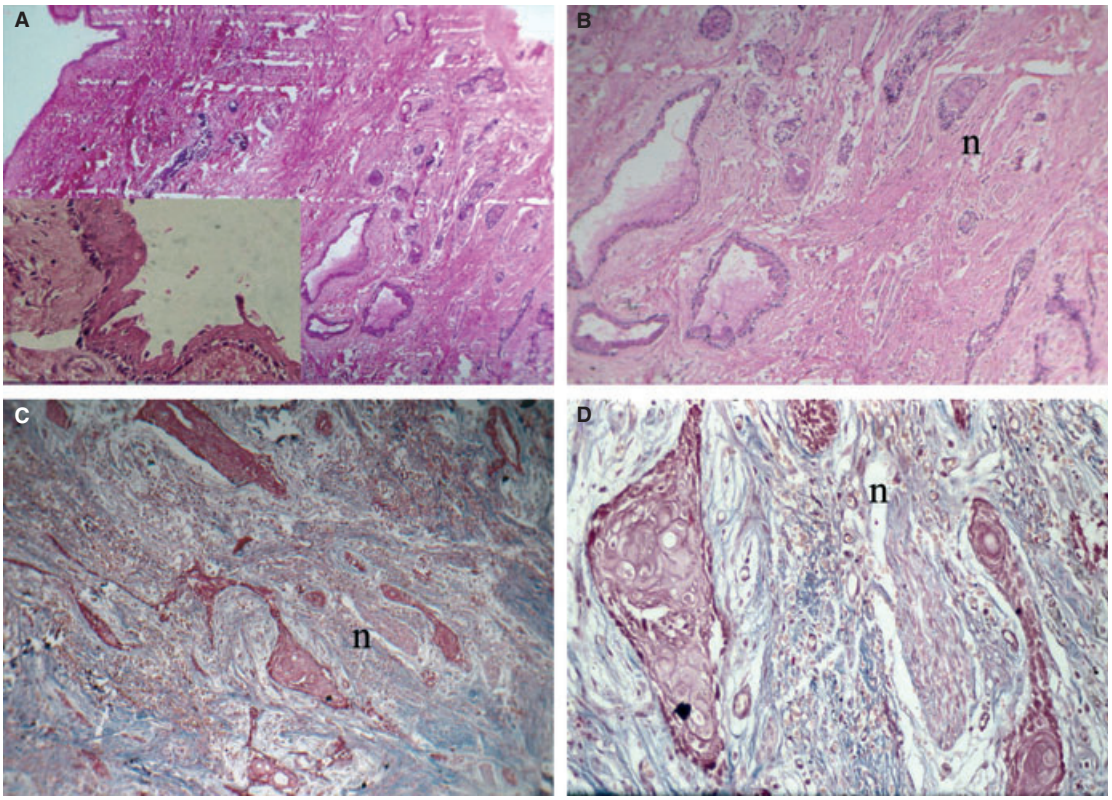

Figure 3 (A,B) Shows microcysts and odontogenic islands. Inset: non-specific stratified epithelium with hyperchromatic and vacuolated basal cells. (C,D) Odontogenic islands with peripheral columnar cells in close proximity to a nerve fiber (n) (Original magnification ×24, ×60, ×60, ×320. H & E, Masson Trichrome).

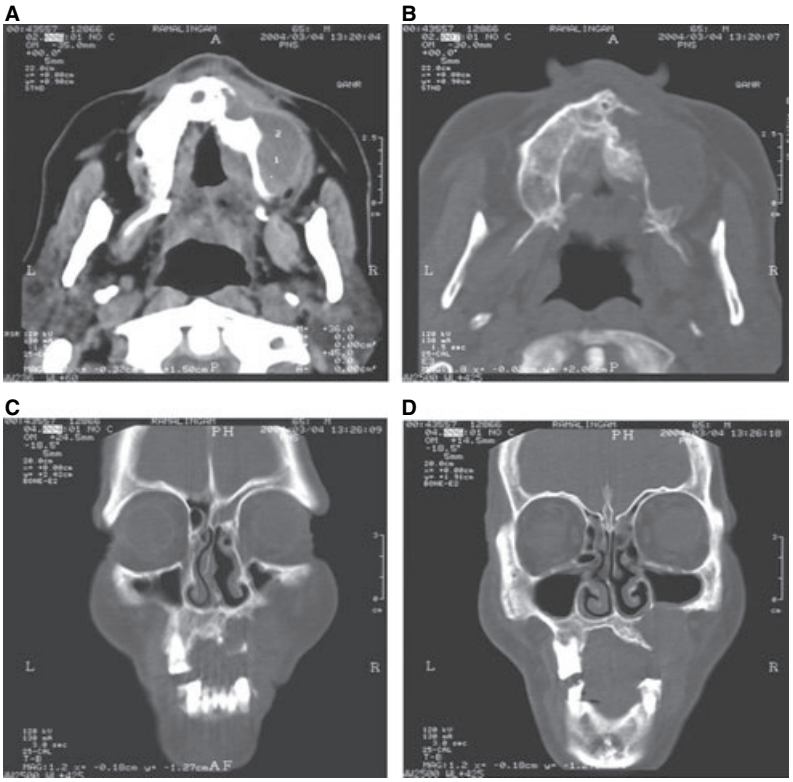

Figure 4 (A) Computed topography scan shows soft tissue lesion in the right maxillary region. (B) Multiple loculi and expansile lesion with thin and scalloped border on the palatal aspect. Note the extensive destruction of buccal cortical plate and alveolar process, and in (C,D) destruction of floor of the maxillary sinus and nasal cavity is evident.

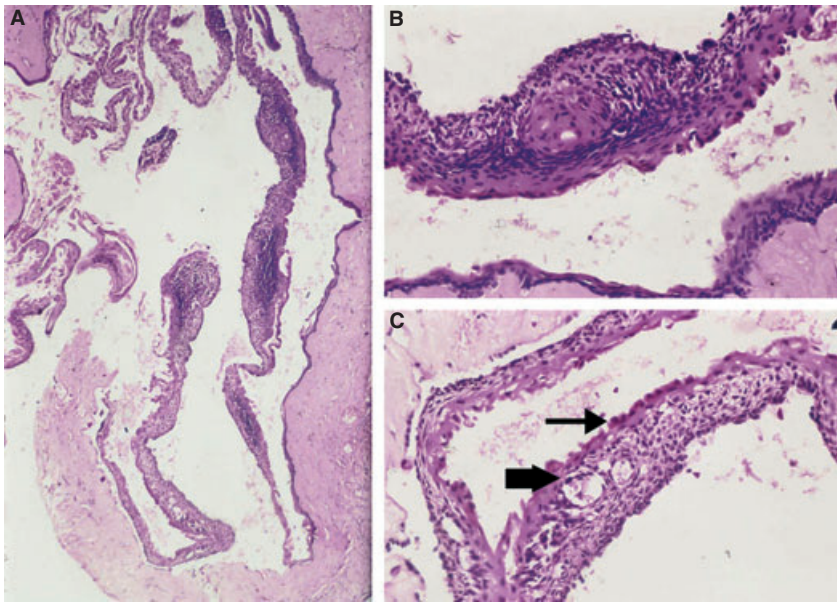

Figure 5 (A) Non-specific lining with epithelial thickening and detachment. (B) Epithelial thickening or whorls. (C) Surface cuboidal cells (arrow) and microcystic spaces (thick arrow) (Original magnification ×80, ×320, ×320, H&E).

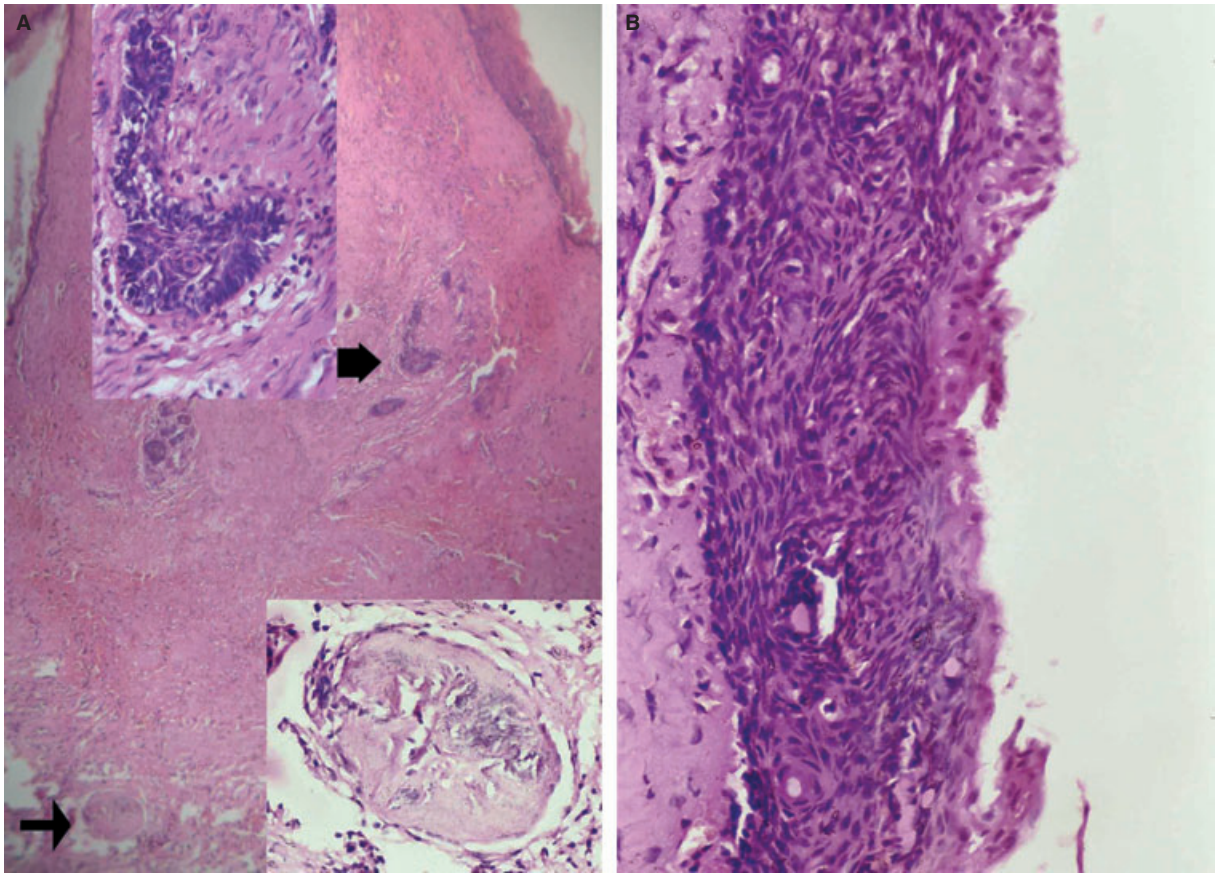

Figure 6 (A) Shows ameloblastoma islands (arrow head) and dystrophic calcification (arrow) in the wall. Inset: higher magnification. (B) Stratified epithelial lining lacking polarization is evident (Original magnification ×80, ×400, H&E).

unicystic ameloblastoma. The lack of ghost cells and pericoronal radiolucency excludes the possibility of calcifying odontogenic cyst and dentigerous cyst from consideration in the present lesion. However, distinction from *de novo* unicystic ameloblastoma and glandular odontogenic cyst is regarded difficult – see below.

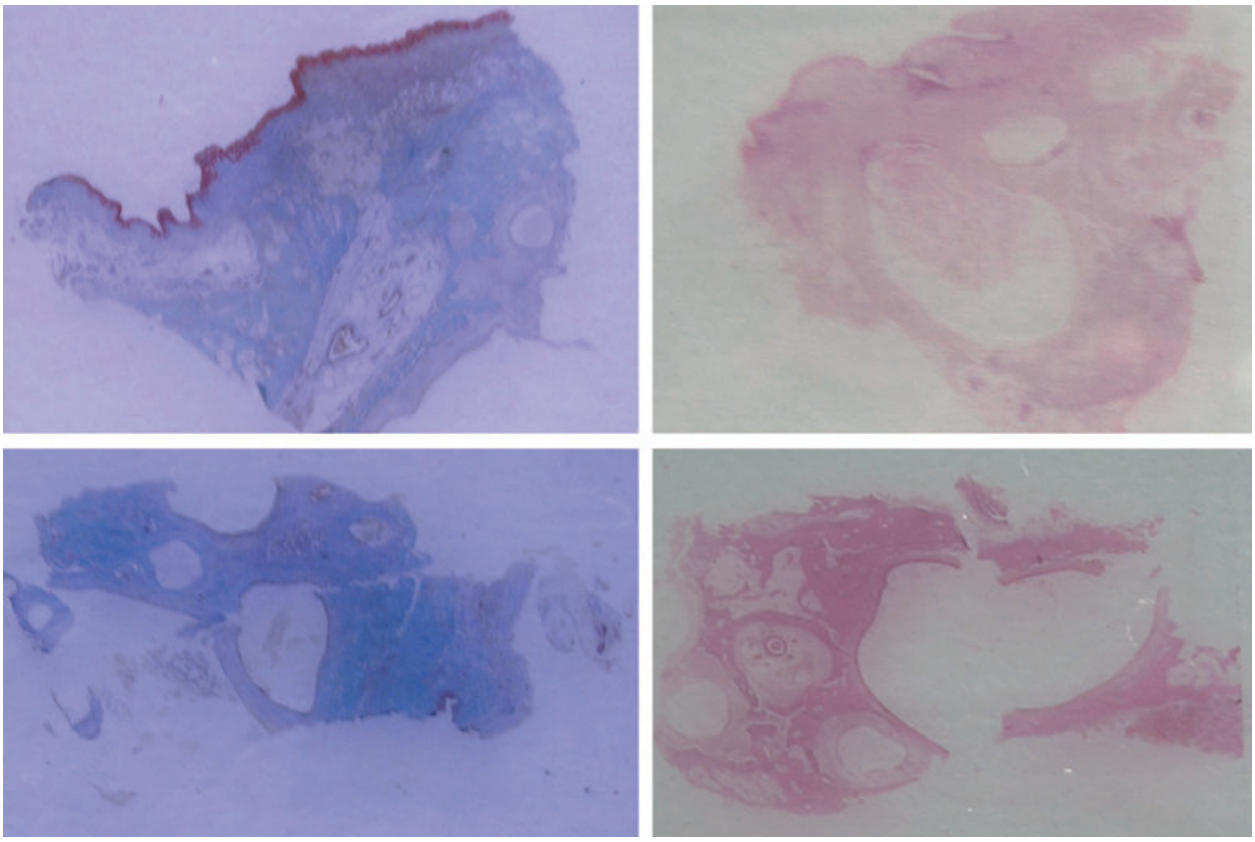

Figure 7 Slide scanner view of macro and microcysts.

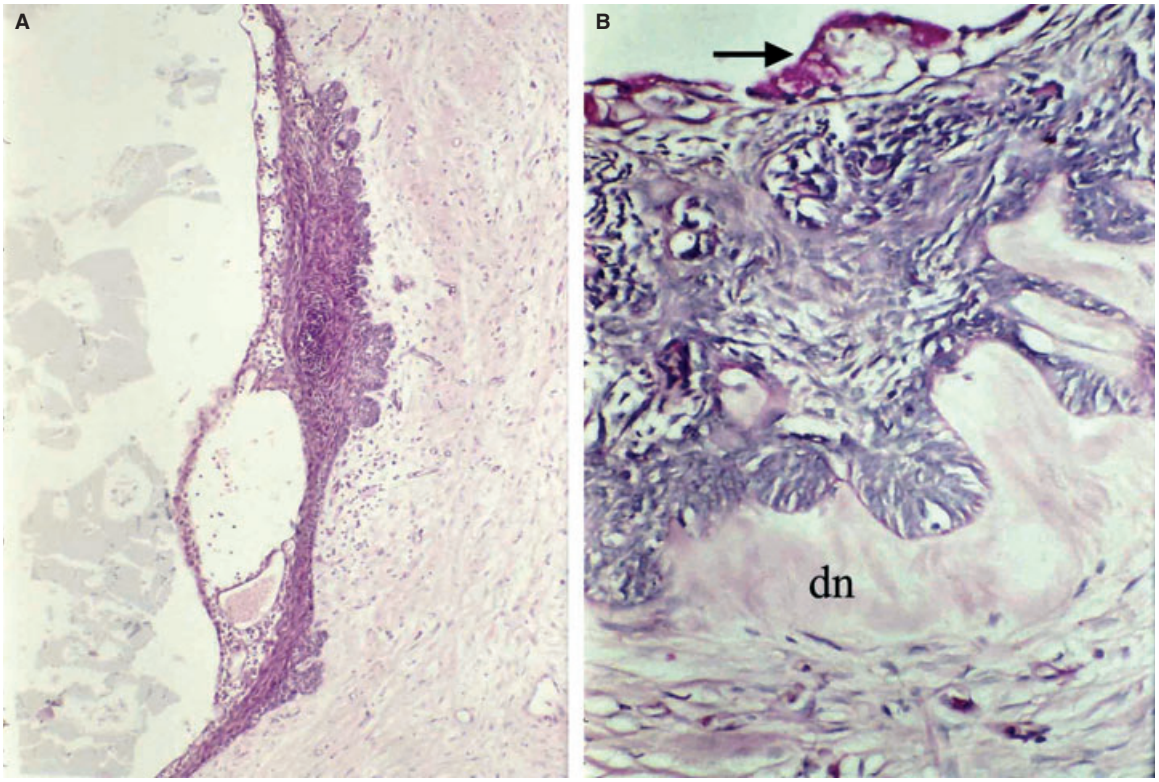

Figure 8 (A) Macrocysts with epithelial thickening lacking polarization and down growths. (B) Lining with surface mucous cells (arrow) and basal ameloblasts-like cells, and juxtaepithelial dentinoid (dn) (Original magnification ×100, ×400. H&E, PAS).

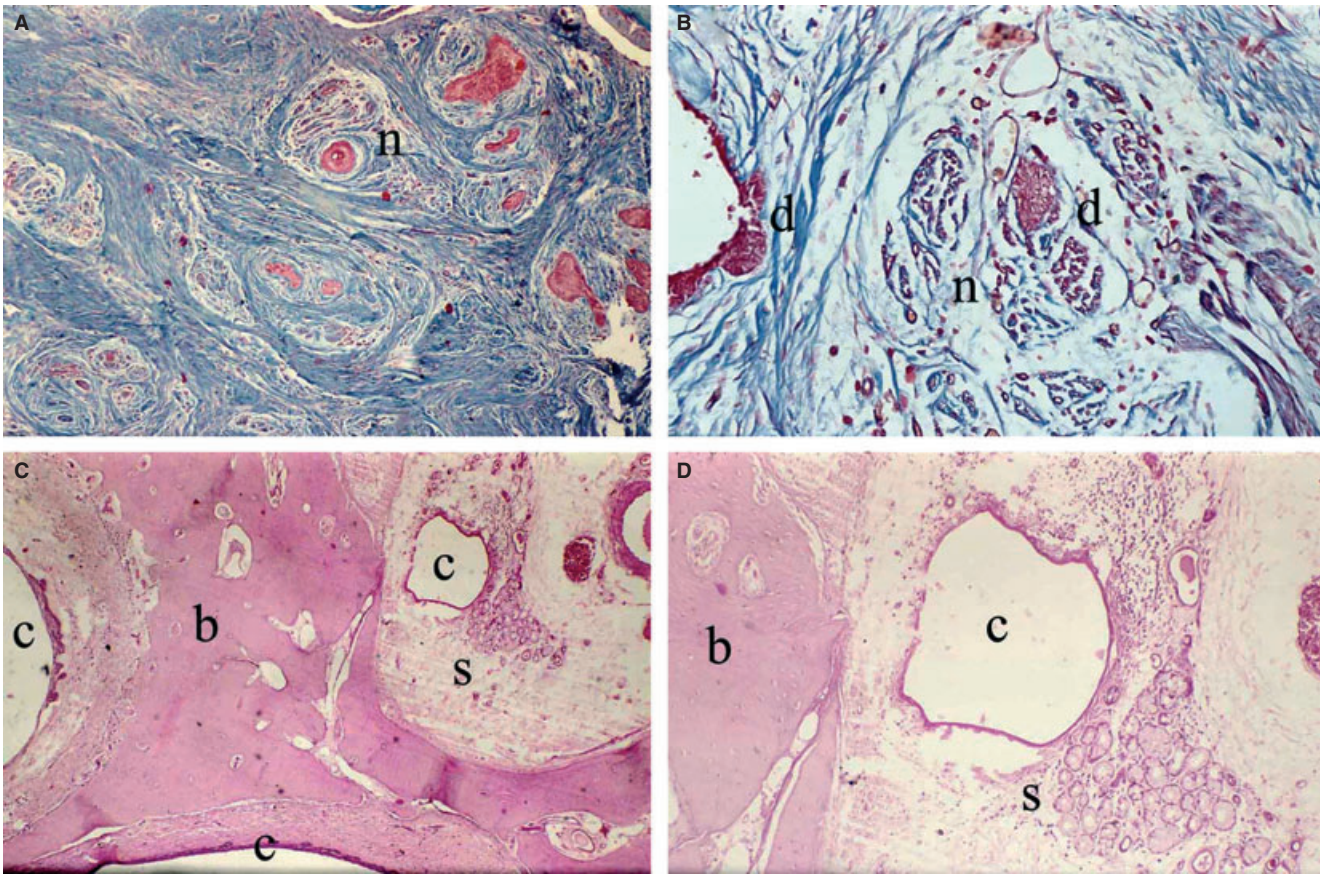

Figure 9 (A) Intraneural and perineural invasion (n). (B) Intraneural invasion of cells similar to the cells in the downgrowth (d) of a microcyst. (C) Encroachment of mucous salivary acini (s) present deeper in the bone (b) by a microcyst (c), also evident in this field are two macrocysts. (D) Higher magnification of (C) (Original magnification ×80, ×320, ×80, ×320).

In the current case, the section from the posterior part of the lesion showed a multicystic lesion characterized by hyperchromatic and vacuolated basal cells with surface cuboidal cells. In addition, there were ameloblastoma islands in the connective tissue wall. While basal cell hyperchromatism and cytoplasmic vacuolation with cuboidal cells at the surface are regarded as characteristic features of the glandular odontogenic cyst (2), there has been no accepted incidence of ameloblastoma in the glandular odontogenic cyst, although a case of glandular odontogenic cyst showed ameloblastoma in the fibrous wall (2). Sections from the anterior part, in contrast, were characterized by multiple macro- and microcystic cavities showing features of early ameloblastomatous changes and juxtaepithelial hyalinization. The common microscopic denominator in both sites is the presence of epithelial thickening lacking polarization and neural invasion.

The varied histomorphology of the present lesion renders specific diagnosis of unicystic ameloblastoma or glandular odontogenic cyst more difficult. As a substantial portion of unicystic ameloblastoma manifests a non-specific lining that may show focal epithelial thickening (3, 4), this lesion could be called unicystic ameloblastoma. But, by definition, it is a single cystic process rather than multicystic. On the other hand, the present lesion in part meets the histological criteria necessary for the diagnosis of glandular odontogenic cyst but lacks other defining features (2). Interestingly, High *et al*. (5) in their 1996 paper preferred the term polymorphous odontogenic cyst to a group of similar cystic lesions that were characterized by diverse histological features. In fact, the photomicrograph of their case 1 closely resembles Fig. 6B. But unlike the case cited here, it was the histology of a recurrent lesion.

The possibility of a collision of two distinct cystic lesions was considered, but the unifying features of epithelial thickening and neural invasion observed in the present lesion indicate, ameloblastomatous transformation of an odontogenic cyst.

This case is interesting not only for its neurotrophism and bone invasion but also for the apparent encroachment of the minor salivary gland. Together, these features indicate the histological aggressiveness of the lesion. With exception (6), there has been no report of neural invasion in odontogenic lesions. It is important that caution should be exercised when interpreting neural invasion, as odontogenic rests are rather common occurrence in the nerves of jaw bones (7).

From the surgeon's point of view, a pathology report is an important pre-operative communication tool that could help to make correct treatment plan. In the current case, a non-specific diagnosis of odontogenic cyst with mural ameloblastoma indicates that the lesion is a cyst but has differentiated towards ameloblastoma. Such interpretation often poses problems in therapeutic decision making, as to treat it as ameloblastoma or cystic ameloblastoma. The former requires radical treatment while the latter can be treated by enucleation and curettage. As maxillary ameloblastoma tend to invade and extend to adjacent vital structures, a second surgical intervention would not be a prudent approach in view of the stated aggressiveness of unicystic ameloblastoma in older individuals (8). The lesion was considered to be aggressive and destructive as evident on imaging studies (9) and therefore, resection was deemed appropriate in view of the post-surgical findings too.

From the foregoing, it appears that the present lesion defies classification because of the wide spectrum of histological changes, nonetheless, is still sufficient enough to warrant a diagnosis of the glandular odontogenic cyst. Furthermore, the aggressive clinical and histological features would suggest the potential for malignancy, perhaps a malignant variant. But there is no local or distant disease during the follow-up of 2 years and 9 months. Therefore, the authors interpret with caution that the extensive bone-destructive features together with the invasion of nerves (peri- and intraneural) and the salivary gland may signify a more aggressive biological behavior rather than malignancy.

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