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LETTER TO THE EDITOR

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Solid-cystic tumor variant of odontogenic keratocyst: an aggressive but benign lesion simulating keratoameloblastoma

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Sir,

Keratoameloblastoma (KA), which was first described by Siar and Ng [9] under the descriptive term "combined ameloblastoma and odontogenic keratocyst," is a unique tumor within the spectrum of ameloblastoma. To our knowledge, only nine well-documented examples including a rarer papilliferous form have appeared in the English-language literature [1, 2, 5, 7, 9, 10]. We present an 18-year follow-up case of a solid-cystic tumor variant of odontogenic keratocyst (OKC) in the mandible that may be confused with KA. This case defies easy diagnostic and nosologic classification because it demonstrated combined features of KA and OKC, each belonging to a different pathologic entity.

A 49-year-old woman first presented in 1984 with a multilocular radiolucent lesion, measuring 4.5 cm in diameter, in the left mandible. A jaw cyst had been removed from the same site at another institution in 1973, but no further information of the initial biopsy was available. After cystectomy, conservative enucleation or curettage was performed three times (in 1985, 1986, and 1990) because of recurrence. However, in 1998, a honeycombed radiolucency appeared again. Through en-block resection, the lesion was under control for 4 years. During the clinical course, there was no evidence of involvement of the adjacent soft tissues or lymph nodes.

Surgical materials from 1984 to 1990 had essentially similar histological patterns. They showed a benign combined proliferation of solid epithelial follicles with marked central keratinization and multiple keratinizing cysts of varying size (Fig. 1A). Solid nests were in direct continuity with cysts and invaded the cancellous bone. The features diagnostic for ameloblastoma (e.g., peripheral palisading, reversed polarization of nuclei and subnuclear vacuolization) were exceptional (Fig. 1B). Thin squamous epithelial lining of keratinizing cysts had the characteristics of OKC and exhibited extensive proliferation in some areas (Fig. 1C). Notably, the finally resected specimen (in 1998) contained multiple separate OKCs within the marrow spaces (Fig. 2A, B). Some of them showed finger-like proliferations (Fig. 2C). Even on step sections, none involved the solid components.

The present solid-cystic epithelial lesion was originally regarded as KA but proved to be a solid form of OKC with the retrospective review. The patient's significantly excellent prognosis over a period of 18 years ruled out the possibility of highly differentiated squamous cell carcinoma arising in an OKC. There are review articles of clinical and histological observations that indicated that some OKCs behave as aggressive as a true neoplasm [3, 8]. Presumably, the neoplastic OKC family span a broad clinicopathologic spectrum from benign pure cystic form with infiltrative properties on one end [6] to cytologically malignant form with capability of metastasis [4] on the other, with the present solid-cystic form occupying an intermediate position, probably nearer to the benign end, in both morphology and behavior. Very recently, Vered et al. [11] reported a similar solid OKC in the maxilla with the infiltration into the surrounding soft tissues, but it is difficult to compare their lesion to ours because their case was published in the abstract form.

KA is a rare variant of ameloblastoma characterized histologically by an admixture of solid tumor islands resembling follicular ameloblastoma with extensive keratinization, and multiple keratinizing cysts bear characteristics of OKC [5, 7, 9, 10]. Overall, the present case showed architectural characteristics of KA; therefore, the question arises whether this lesion fits the criteria for KA. However, the lack of obvious palisading and reversed nuclear polarity of the peripheral tumor cells could not confirm the diagnosis of ameloblastoma. Interestingly, our case did not change its KA-like appearance for over 6 years, but recurred as an ordinary OKC 8 years after the surgery. Consequently, the present lesion belongs to a rare

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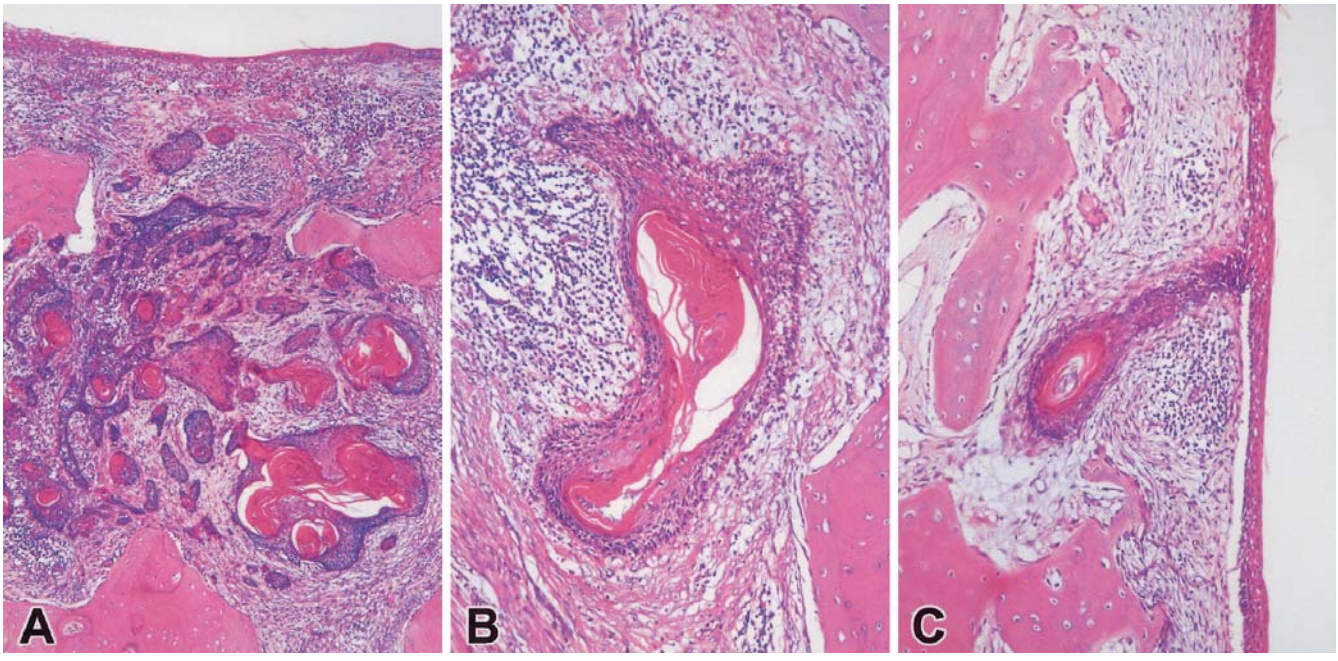

Fig. 1 Solid-cystic epithelial lesion excised in 1984. **A** Solid component invading cancellous bone. **B** Representative solid follicle showing central keratinization. **C** Cystic component lined by thin squamous epithelium of uniform thickness. Note detachment and down-growth of cyst epithelium

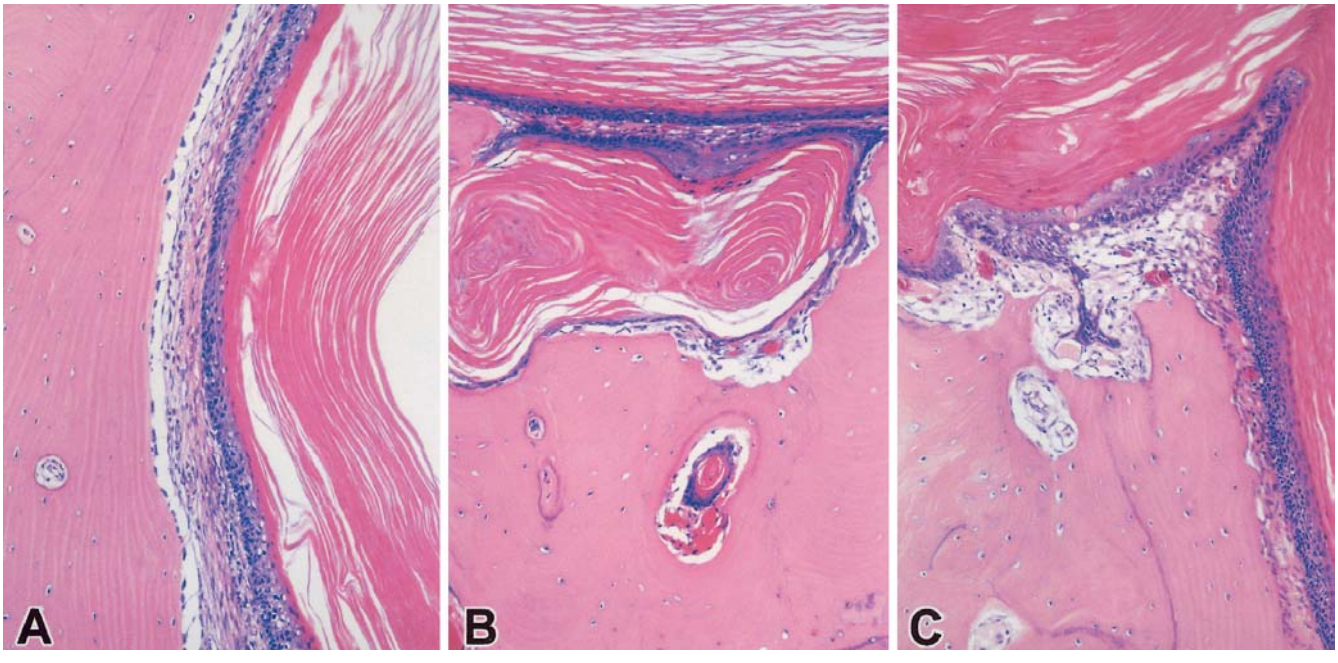

Fig. 2 Multiple keratinizing cysts resected in 1998. **A** Lining epithelium showing pathognomonic features of odontogenic keratocyst. **B** Microcysts within marrow spaces. **C** Finger-like proliferation of cyst epithelium

neoplastic variant of OKC separable from KA. Based on our experience and that in the literature [3, 8], the solid form of OKC behaves as a benign, locally invasive tumor with a propensity for recurrence. It is important to realize that a solid-cystic tumor variant of OKC could be confused with KA.

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