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CASE REPORT

Non-calcifying variant of calcifying epithelial odontogenic tumor with Langerhans cells

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Calcifying epithelial odontogenic tumor (CEOT) is a rare type of odontogenic tumor. The most characteristic feature of the classical CEOT is the presence of amyloid globules and Liesegang ring calcification in the tumor tissue. Here, we present a non-calcifying variant of intraosseous CEOT with the presence of Langerhans cells within tumor epithelial nests in a 52-year-old Taiwanese woman. The patient was referred from a local dentist to our hospital for treatment of a unilocular radiolucent lesion at the right anterior region of the maxilla. The lesion was excised. Microscopically, the tumor was composed of small nests or strands of odontogenic epithelial cells and amorphous eosinophilic globules of amyloid-like materials in a loose fibrous connective tissue stroma. The tumor epithelial cells were positive for pan-cytokeratins (AE1 and AE3). Langerhans cells demonstrated by anti-CD1a staining were found in nests or strands of tumor epithelial cells. The eosinophilic globules were positive for Congo red and showed green birefringence when subjected to polarized light. Review of the English literature revealed two cases of non-calcifying variant of intraosseous CEOT with Langerhans cells in the anterior and premolar regions of the maxilla. Taken together, we suggest that the non-calcifying, Langerhan cell-rich variant of CEOT may have a distinct predilection for occurrence in the anterior and premolar region of the maxilla in contrast to the classical CEOTs that usually occur in the molar and ascending ramus area of the mandible.
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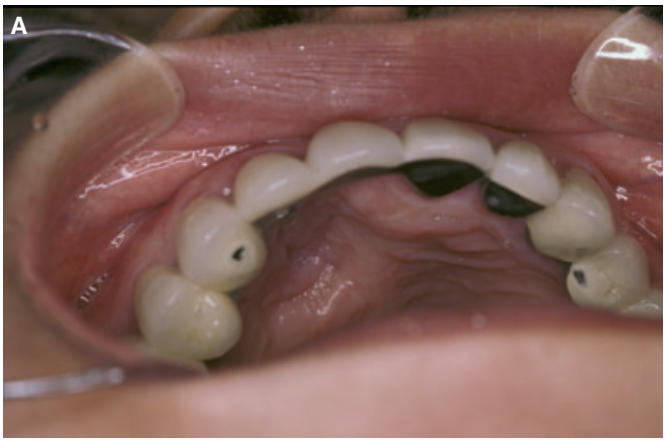
Case report

A 52-year-old Taiwanese woman was referred from a local dentist for evaluation of a radiolucent lesion at the right anterior region of the maxilla in June 2006. There were no active symptoms and signs at the lesional site, as stated by the patient. The patient had a previous history of infiltrating ductal carcinoma of the left breast, and mastectomy and subsequent chemotherapy were performed 3 years ago. Intraoral examination revealed a depression over the right anterior hard palate opposing to the teeth 12 and 13. The overlying mucosa was intact and slightly reddish in color (Fig. 1A). The consistency of the lesion was soft on palpation. The panoramic and occlusal radiographies revealed a unilocular radiolucent lesion without a definite sclerotic border at the right anterior region of the maxilla (Fig. 1B). No evidence of root resorption or displacement of the teeth was found, but involvement of the right nasal floor was discernible.

Incisional biopsy was performed. Microscopically, the lesion was composed of cords of ovoid cells with nuclear atypia and globules of colloid-like materials in a fibrous connective tissue stroma. Immunohistochemical studies showed that the tumor epithelial cells were positive for pan-cytokeratins (AE1 and AE3; Biogenex, San Ramon, CA, USA) and negative for estrogen receptors. Because of the previous history of breast cancer, a metastatic infiltrating ductal carcinoma from the left breast cancer was diagnosed.

The patient was admitted to our ward for surgical excision of the tumor in September 2006. Magnetic resonance imaging showed a mildly irregular nodule about 2.4 cm in diameter at the right anterior area of the maxilla, which compressed upward on the right nasal floor. This lesion showed low signal intensity on T1-weighted image, but high signal intensity on T2-weighted image with strong enhancement. Whole-body positron emission tomography demonstrated a focal area of 2-[Flurorine-18]fluoro-2-deoxy-D-glucose hypermetabolism at the right anterior maxilla (early/delayed standard uptake value = 3.6/4.4). The patient received the partial maxillectomy from tooth 23 to tooth 16 and the right supraomohyoid neck dissection on September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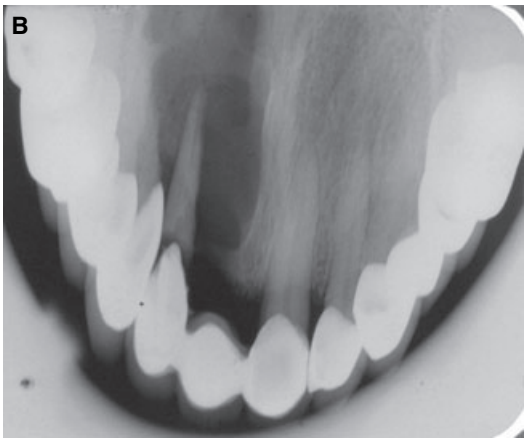
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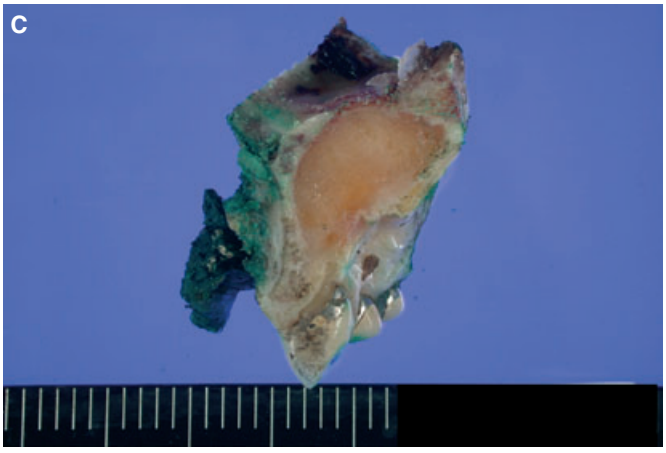
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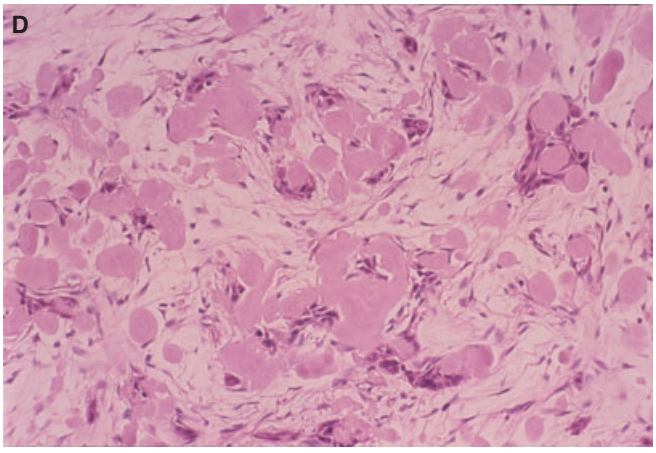
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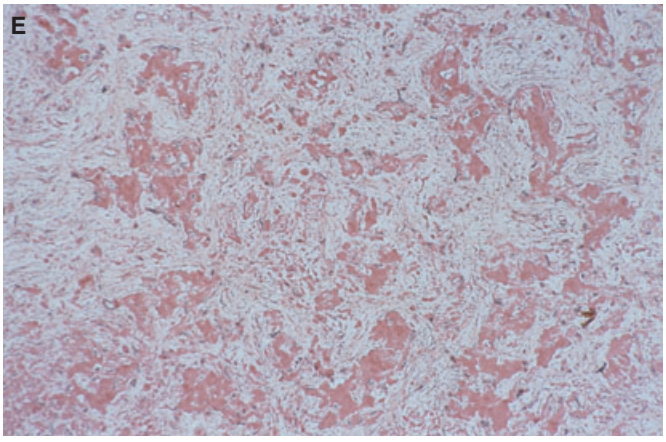
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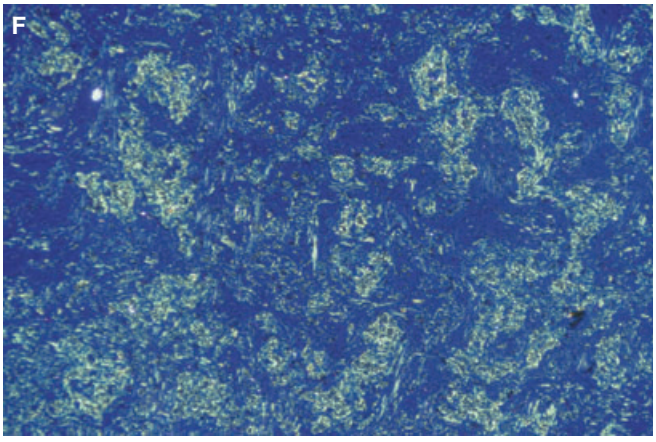
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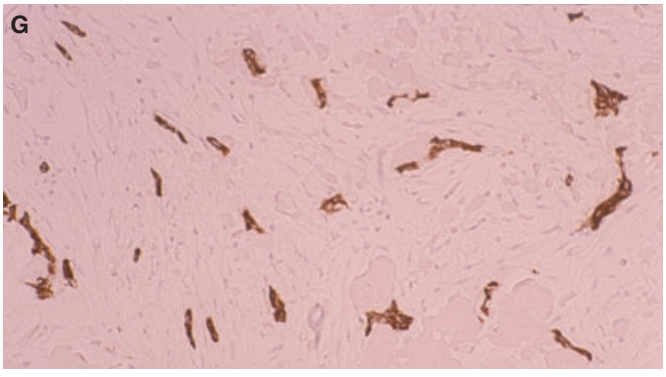
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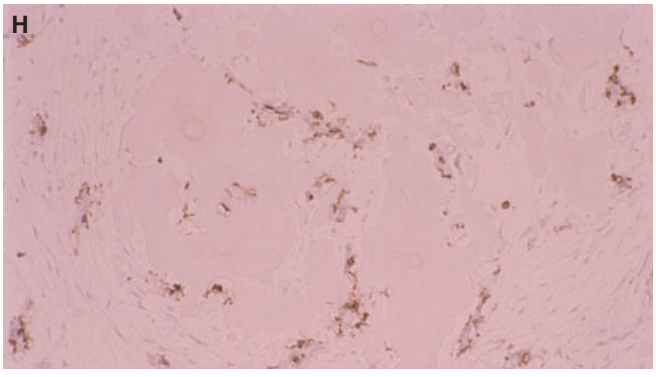
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5, 2006. The superior section margin extended up to the inferior nasal concha level. The patient stood the procedure well and the wound healing was uneventful.

The specimen submitted was fixed in 10% formalin overnight. On serial section, a well-defined orange tumor measuring 2.3 × 2.0 × 2.0 cm in size was found in the maxillary bone at the depression area. No gritty sensation was felt while cutting. Grossly, the tumor perforated the right nasal floor and invaded the palatal mucosa (Fig. 1C). Resorption of the root apices of teeth 12 and 13 was also noted. After decalcification in Plank-Rychlo solution (Muto Pure Chemicals Co., Tokyo, Japan), all specimens were embedded in paraffin and cut in serial sections of 5 μm. The tissue sections were used for routine hematoxylin and eosin (H&E), Congo red, and immunohistochemical stains.

Low-power view of H&E-stained sections showed that the tumor was well defined but lacked a definite capsule. It perforated into the right nasal floor and invaded the palatal mucosa; however, no continuity of tumor epithelial nests with the surface oral epithelium was discernible. The tumor was composed of small nests or strands of odontogenic epithelial cells and amorphous eosinophilic globules of amyloid-like materials in a loose fibrous connective tissue stroma. The epithelial nests or strands were usually distributed at the periphery of the hyalinized globules (Fig. 1D), which were positive for Congo red (Fig. 1E) and showed green birefringence when subjected to polarized light (Fig. 1F). Immunohistochemically, the tumor epithelial cells were positive for pan-cytokeratins (Fig. 1G) and negative for estrogen receptors (1D5; Dako, Glostrup, Denmark) and progesterone recepters (1A6; Novocastra, Newcastle, UK). Langerhans cells demonstrated by anti-CD1a (JPM30; Novocastra) staining were found in nests or strands of tumor epithelial cells (Fig. 1H). No concentric Liesegang ring calcification was observed in all sections. The pathologic diagnosis was a non-calcifying variant of calcifying epithelial odontogenic tumor (CEOT). All section margins were free. The submandibular gland showed minimal histologic change. No evidence of metastasis was detected in the seven lymph nodes dissected from the right neck tissue.

Comments

Calcifying epithelial odontogenic tumor is a locally invasive benign tumor that was first described as an entity by Pindborg. The tumor is typically composed of islands, strands or sheets of polyhedral epithelial cells and amorphous eosinophilic globules of amyloid materials in a fibrous connective tissue stroma. Concentric Liesegang ring calcification often develops within these amyloid globules (1–5). However, a few case reports of intraosseous CEOT with minimal calcification were documented (4, 5).

After reviewing the literature, we found that our case revealed striking histologic resemblance to the two cases reported as non-calcifying variant of CEOT (4, 5). These two reported cases and our case all revealed odontogenic epithelial cells arranged in thin strands or small nests and globules of amyloid materials. Noticeably, no calcifications were presented in contrast to the stereotypic CEOT. Another interesting finding was the demonstration of scattering Langerhans cells within small odontogenic epithelial nests in both of these two reported cases. Anti-CD1a immunostaining also confirmed the presence of Langerhans cells in odontogenic epithelial nests in our case. The pathologic significance of the presence of Langerhans cells in CEOT is still unclear. Langerhans cells are bone marrow-derived cells that migrate into the oral epithelium and serve as antigen-presenting cells. Because both oral and odontogenic epithelia originate from the same oral ectoderm, it is possible that Langerhans cells may also migrate into tumor odontogenic epithelial nests. These Langerhans cells are very close to the amyloid globules. Therefore, they may phagocytize and process the amyloid material and then present the processed antigens to T lymphocytes in the regional lymph nodes. Owing to the paucity of this special variant of CEOT, the true pathologic significance of the presence of Langerhans cells in CEOT needs further study.

Furthermore, the clinical features of the three cases were also comparable. The lesion reported by Asano et al. (4) involved the maxillary bone from the right central incisor to the right second premolar region of a 44-year-old Asian female patient. It presented as a well-defined unilocular radiolucent lesion on the panoramic radiograph. The other lesion presented by Takata et al. (5) was located in the left maxillary canine and first premolar area of a 58-year-old Asian male patient. Radiographically, it was also a well-defined unilocular radiolucent lesion. Taken together, we suggest that the non-calcifying, Langerhan cell-rich variant of CEOT may have a distinct predilection for occurrence in the anterior and premolar region of the maxilla in contrast to the classical CEOTs that usually occur in the molar and ascending ramus area of the mandible. More accumulative data are needed to further confirm this specific finding.

Figure 1 Radiographic, clinical and histological photographs of the patient. (A) Clinical photograph showing a depression over the right anterior hard palate opposing teeth 12 and 13. The overlying mucosa was intact and slightly reddish in color. (B) Occlusal radiograph revealing a unilocular radiolucent lesion at the right anterior region of the maxilla. (C) Sagittal section of the specimen showing an orange tumor which perforated the right nasal floor and invaded the palatal mucosa. (D) Hematoxylin and eosin-stained section exhibiting that the tumor was composed of small nests or strands of odontogenic epithelial cells and amorphous eosinophilic globules of amyloid-like materials in a loose fibrous connective tissue stroma. The epithelial nests or strands were usually distributed at the periphery of the hyalinized globules (original magnification, 50×). (E) Congo red-stained section showing many orange-colored globules of amyloid-like materials in the tumor stroma (original magnification, 10×). (F) These orange-colored globules showed green birefringence when subjected to polarized light (original magnification, 10×). (G) The tumor epithelial cells were positive for pan-cytokeratins (original magnification, 50×). (H) Dendritic Langerhans cells demonstrated by anti-CD1a staining were found in nests or strands of tumor epithelial cells (original magnification, 50×).

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