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Case Report

CT and MR Imaging Appearances of an Extraosseous Calcifying Epithelial Odontogenic Tumor (Pindborg Tumor)

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Summary: We herein report a rare case of extraosseous calcifying epithelial odontogenic tumor with local aggressive behavior. CT and MR imaging showed the distinctive appearances of this histologic entity. We briefly discuss the radiologic features of calcifying epithelial odontogenic tumor and the relevant literature.

The differential diagnosis of a mass in the maxillary antrum is wide and includes such diverse causes as mucous retention cyst, dentigerous cyst, trauma, polyp, carcinoma, and Wegener's granulomatosis (1). Plain radiographs and cross-sectional images obtained by performing CT and MR imaging all may help to determine the diagnosis. We herein present a case of recurrent extraosseous calcifying epithelial odontogenic tumor (CEOT) with progressive local invasion in a young adult. Both CT and MR imaging facilitated surgical planning in this case by delineating the extent of the mass and by revealing local aggressive behavior. We present the imaging findings, the differential causes, and a review of the literature.

Case Report

A 23-year-old Chinese man presented with a 1.5-year history of left facial swelling and tenderness after sustaining a left facial injury in an accident. He had undergone a Caldwell-Luc operation at another institution, at which time a cystic mass involving the left maxilla was noted and a biopsy specimen was obtained intraoperatively. Pathologic evaluation of the lesion revealed a CEOT.

Fifteen months later, this young man presented to the otorhinolaryngology department, complaining of another episode of progressive left facial swelling and double vision. A clinical examination revealed swelling of the left cheek. There was no proptosis. The patient's vision and eye movement were normal. Plain radiographs of the sinuses showed opacities within the left maxilla only, with the other sinuses appearing normal. The maxillary sinus was expanded, and an unerupted tooth was present in its posterior wall (Fig 1A).

A CT scan of the maxillofacial region revealed a rounded heterogeneous mass almost filling the entire left maxillary antrum. The lesion contained a central tooth with surrounding amorphous calcifications and soft tissue elements in the periphery (Fig 1B). There was associated ballooning and remodeling of the maxilla, with resultant medial deviation of the left lateral nasal wall and mild elevation of the left orbital floor. There were foci of bony destruction at the anterior maxillary wall.

MR imaging showed a large heterogeneous mass occupying the expanded left maxilla. There were intralesional areas of low signal on the T1- and T2-weighted sequences, consistent with calcification and the maxillary tooth identified on the CT scan. T1-weighted images obtained after the administration of contrast material (0.1 mmol/kg) showed heterogeneous enhancement, especially on the periphery of the lesion. Anterolaterally, there was a small cortical breakage and extension to the buccinator muscle and subcutaneous tissues of the left cheek (Fig 1C–D). The tumorous lesion extended inferiorly to involve the alveolar ridge. The lower second and third molar teeth were absent.

The tumor was enucleated with the patient under general anesthesia. Intraoperatively, one well-encapsulated mass, with areas of calcification and a central unerupted second molar tooth, was seen. The third molar tooth was embedded behind the posterior wall of the maxillary sinus. The tumor and the third molar tooth were removed with clear margins (Fig 1E). Histologic analysis confirmed a CEOT.

Discussion

CEOT is a benign neoplasm of unknown cause related to the odontogenic apparatus (2). Pindborg, in 1958, first categorized it as a distinct histopathologic entity. It is characterized by three different histologic hallmarks. The first structure comprises solid epithelial layers or islands with variable thickness and prominent borders. The polyhedral epithelial cells exhibit nuclear polymorphism and eosinophilic cytoplasm, but mitoses are rare. The second hallmark consists of an acellular hyalinized stromal bridge, interspersed with foci of an amyloid-like substance. The third hallmark comprises a variable amount of round, conglomerate, or concentric laminar (Liesegang ring) calcifications (3−5). Previously, uncertainty regarding the histologic characteristics of CEOT was reflected in the variety of terms for the disease, including unusual ameloblastoma, cystic odontoma, and adenoid adamantinoma (5). Association with adenomatoid odontogenic tumor and dentigerous cyst had been

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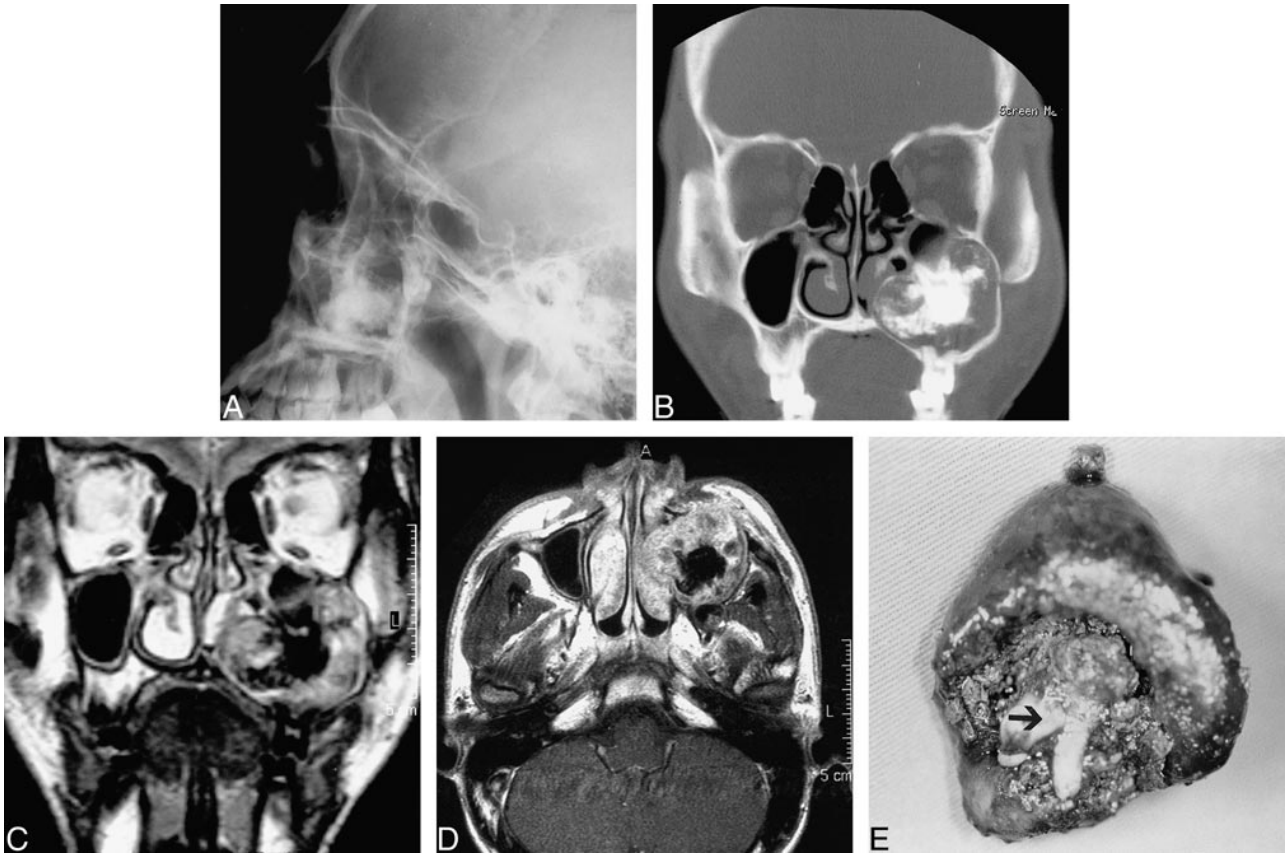

FIG 1. Images from the case of a 23-year-old man with left facial swelling.

A, Lateral radiograph of the sinus shows opacities within the left maxilla and an unerupted tooth in its posterior wall.

B, Coronal CT scan reveals a rounded heterogeneous mass almost filling the entire left maxillary antrum. The lesion contains a central tooth with surrounding amorphous calcifications and soft tissue elements in the periphery.

C, Coronal T2-weighted MR image shows a large heterogeneous mass occupying the expanded left maxilla. There were intralesional areas of low signal in the T2-weighted sequences, consistent with calcification and the maxillary teeth identified on the CT scan.

D, Axial contrast-enhanced T1-weighted image shows a large heterogeneous mass occupying the expanded left maxilla. There were intralesional areas of low signal in the T1-weighted sequences, consistent with calcification and the maxillary teeth identified on the CT scan. Non-homogeneous enhancement can be seen, especially on the periphery of the lesion. Anterolaterally, there is a small cortical breakage and extension to the buccinator muscle and subcutaneous tissues of the left cheek.

E, Gross specimen shows an unerupted second molar (*arrow*) in the calcified substance of the well-encapsulated tumor.

reported and suggests heterogeneity in histopathogenesis (2, 6, 7).

CEOT occurs rarely, with a frequency ranging from 0.17% to 1.8% of all odontogenic tumors (3, 7). Both sexes are equally affected. The disease usually manifests between the ages of 20 and 60 years. Most of the patients are asymptomatic at the time of initial diagnosis. The slowly enlarging mass can result in mechanical effects. Although it is thought to be a benign tumor, local tissue invasion has been documented (8). The reported recurrence rate ranges from 10% to 14% (3, 9, 10).

Two thirds of CEOTs arise in the mandible, whereas one third arise in the maxilla (7). The majority of recorded cases have been centrally located, mainly in the premolar-molar region of the mandible. A recent article by Ng and Siar (3) reported a predilection for the maxilla in Asians, contrary to the higher mandibular prevalence in the West. Although it was predominantly an intraosseous lesion, the extraosseous type is the rarest (5%) of all CEOT. These have been found peripherally in the anterior maxillary or mandibular gingiva, with, to our knowledge, only six reported cases in the literature (3, 4, 7).

The five patterns of radiographic manifestations of CEOT most likely represent sequential stages in a spectrum of disease rather than discrete categories. The most common two appearances of CEOT are of pericoronal lucency and of lucent areas with diffuse opacities. Other appearances, including mixed lucent-opaque lesion not associated with an unerupted tooth, ''driven snow'' appearance, and a solid opacity, account for a minority of cases (7).

Our case showed MR imaging appearances comparable with most sinus tumors described elsewhere, being of predominantly low signal intensity on T1-weighted images and of high signal intensity on T2-weighted images. Heterogeneous contrast enhancement, seen within the mass, is a common finding in cases of sinus tumors. Interestingly, the extensive calcifications and unerupted maxillary tooth situated in the center of the mass were clearly seen on CT scans and MR images in our case. On

CT scans, they were seen as diffuse high attenuation, suggesting calcifications and ossification. On both T1- and T2-weighted MR images, they were seen as areas of low signal intensity. This accompanying unerupted tooth has been mentioned in 52% of CEOT cases (4, 7). In the majority of cases, intralesional calcifications are associated. Local tissue invasion with breakage of the anterior and medial maxillary walls and alveolar process involvement was shown on contrast-enhanced T1-weighted images as replacement of low-signal cortex by high-signal tumor. MR imaging was superior in showing buccinator muscle and subcutaneous tissue invasion in the cheek in our case. In contrast to the general belief of a less active and less calcified extraosseous type of CEOT, our case included gross calcifications and local invasion. As far as we know, there is no report of the MR imaging appearances of an extraosseous CEOT that exhibits local aggressive behavior.

The CT findings were similar to those described with a heterogeneous mass of slightly low-attenuated rim and high-attenuated center. CT was useful in showing calcifications, unerupted tooth, and bony erosion.

Advance imaging CT and MR imaging delineated both size and extent of the CEOT, which is essential for surgical planning. Enucleation of the tumor remains to be the mainstay of treatment. Because there are only a limited number of cases with long-term results available in the literature, long-term follow-up is indicated (10).

Although rare, it is important to include extraosseous CEOT in the differential diagnosis of a complex mass in the maxillary antrum. A careful search should be made for a characteristic unerupted tooth in the center, which would support an odontogenic origin of the tumor. The presence of pressure remodeling of maxillary antrum and exclusively extraosseous location of the lesion makes malignant diseases, such as osteogenic sarcoma or chondrosarcoma, unlikely causes. The differential diagnosis of odontogenic tumors consists of odontoma and benign fibro-osseous lesions, including fibrous dysplasia and ossifying fibroma. Although fibrous dysplasia is essentially an intraosseous lesion, enlargement of the bone with it may simulate a tumor mass in that area. Ossifying fibroma occurring in the maxilla may have gross calcification within the lesion. The typical radiographic appearance of a complex odontoma is of an amorphous opacity with innumerable discrete tooth-like densities (denticles) (11). This feature is absent in our case, and odontoma is histologically distinct from CEOT. CT and MR imaging should streamline the process of interpretation, but ultimately, histologic examination is mandatory for diagnosis.

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