A RARE ODONTOGENIC TUMOR; CALCIFYING EPITHELIAL ODONTOGENIC TUMOR (PINDBORG TUMOR)

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SUMMARY

Calcifying epithelial odontogenic tumor, which is locally aggressive in nature, is a rare benign tumor of jaws. It usually locates in the posterior mandibular region with the mandible/maxilla involvement rate of 2/1. Since its first description by Pindborg in 1955, several reports with different histopathological forms, clinical features and treatment modalities have been reported. In this report, three cases of calcifying epithelial odontogenic tumor, two located in the maxilla, one located in the mandible are presented. We would like to remind this rare odontogenic tumor with review of the literature and discuss management in plastic surgery.

Key words: Odontogenic tumor, Pindborg tumor

INTRODUCTION

Calcifying epithelial odontogenic tumor (CEOT) is a rare benign tumor of the jaws. In 1955 Pindborg first reported three benign odontogenic tumors arising from the mandible [13]. Since then nearly 200 cases have been reported [2]. There are two main types. Central or intraosseous type is the most common type (95%) that is usually located in the premolar and molar regions and presents as a slow growing intraoral mass. Mandibular to maxillary involvement rate is 2/1 [2]. Extraosseous or peripheral type occurs less than 5% of the cases and usually presents as a painless gingival mass similar to the clinical appearance of fibrous hyperplasia or epulis [2,8].

CEOT accounts for 1% of the odontogenic tumor and it is considered benign but locally aggressive in nature. 10% to 15% recurrence rates were reported in the literature [12]. In this case report we present three cases of CEOT, two of them arising from maxilla and one of them arising from mandible. We would like to remind this rare odontogenic tumor with review of the literature and discuss management in plastic surgery.

CASE REPORT

Case I:

A 32 years old woman was referred to us with a progressive enlargement of her right molar region. The patient also had complaints of nasal stiffness. Clinical examination revealed a firm, painless mass causing facial asymmetry. There was an expansive lesion located at the right hard palate obliterating the maxillary buccal vestibule. Computed tomography (CT) showed a mass occupying most of the maxillary sinus with compression of the left nasal wall and roof of the maxillary sinus. The incisional biopsy revealed CEOT (Figure 1). Our choice of the treatment was right maxillectomy. The patient stayed free of the disease after 8 years of follow up.

Case II:

A 45 years old woman presented with a complaint of swelling extending from the

Received for publication: February 06, 2004

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Figure 1. Sheets of epithelial cells with uniform nuclei and distinct cell borders containing homogeneous globoid material (HE x 310).

right maxillary molar region to the left maxillary anterior incisive region (Figure 2). The swelling had been present for three years on the right premolar area and had gradually increased in size and crossed the midline in several months. CT examination showed a mass covering the right inferior maxillary region, which extended to the left inferior medial maxillary area. Multilocular radiolucent area containing radiopaque masses of varying size and opacity, which was the typical lesion of the CEOT, was seen (Figure 3). After the incisional biopsy of the mass revealed CEOT (Figure 4), we performed right maxillectomy, which was continuous with resection of the left anterior incisive region. The patient stayed free of the disease after 3 years of follow up.

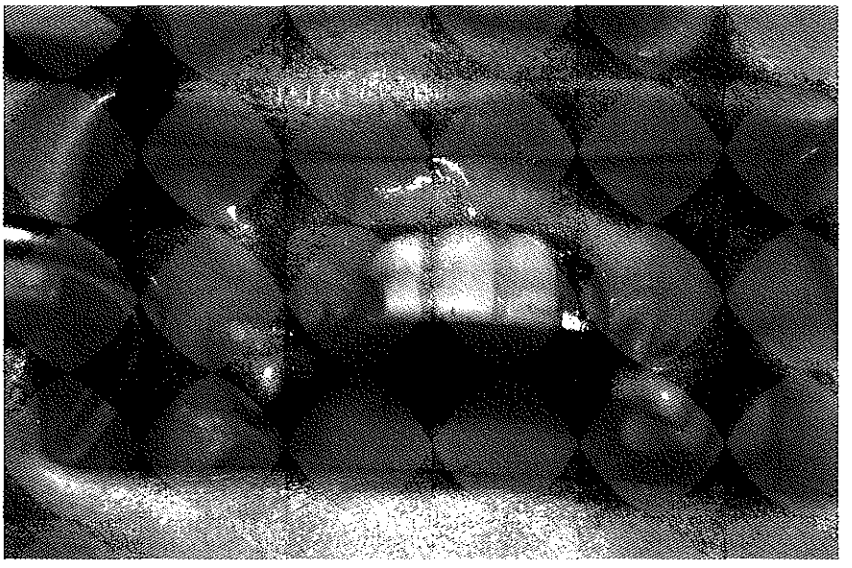
Case III:

A 46 years old man attended to our clinic with a 3x3 cm. painless mass on the right side of his mandible. About 2 years ago he discovered a swelling on his gingiva adjacent to his molar teeth. He received medical treatment for several months until a dentist performed the incisional biopsy (Figure 5). The tumor was located on the right mandibuiar corpus (Figure 6). We performed segmental tnandibulectomy with tumor free margins and reconstructed the defect with an iliac bone graft. The patient stayed free of the disease after 3 years of follow up.

Histopathological Findings:

Ali cases had similar histopathological appearance with minor morphological differences. Solid islands of polygonal cells with eosinophilic cytoplasms and distinct celi membran were seen (Figure 1). Nuclear pleomorphism was minimal. Homogeneous pink material, resembling amyloid or hyaline was a charactcristic feature observed in ali cases (Figure 2). Small foci of calcifications were seen (Figure 3). In the third case dentinoid like material was an additional finding. Amyloid like material stained positive with Congo-red and crezyl violet in two

Figure 2. CEOT located on the right maxillary premolar region.



cases and showed apple-green birefringence under polarized light, which is characteristic for amyloid. Epithelial islands were positive for pancytokeratin.

DISCUSSION

Several reviews about Pindborg tumor have been performed since 1966 [12]. Philipsen et al examined 181 cases of CEOT in 2000 and reported that the mean age of the patients were 36.9 varying between 8 to 92 years. The male/female ratio of this large series was very near even distribution. One third of the cases were known to have association with unerrupted tooth [11].

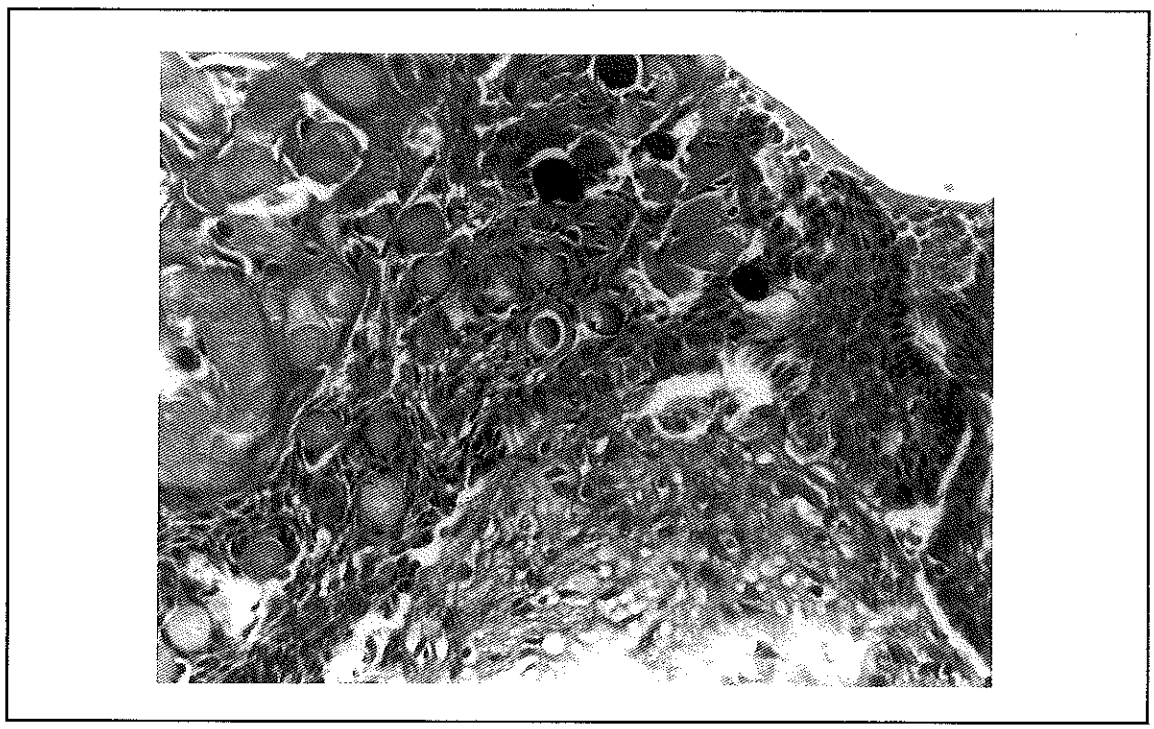
In recent years variants of CEOT, which may have different prognosis, have been described. Noncalcifying CEOT with Langerhans cells, the CEOT displaying cementum like and bone like material and clear cell CEOT are the histopathologic variants [5]. Especially clear celi type shows an aggressive behaviour and indicates a more radical surgical approach. It has a high recurrence rate of 22% and considered to be a low grade odontogenic carcinoma [2]. CEOT has also been found to be associated or combined with other odontogenic lesions such as adenomatoid odontogenic tumor and dentigenous cyst [5,16].

The CEOT shows considerable radiographic variation. Its appearance may range from a diffuse or well-circumscribed unilocular radiolucency to a combined pattern of radiolucency and radiopacity with small intralesional bony septa producing multiocular pattern. A classic lesion exhibits scattered flecks of calcification in the radiolucency, which has been described as a "driven snow" appearance. Kaplan et al studied radiological and clinical aspects of CEOT and concluded that "driven snow" pattern was seen in only a small percentage of cases [10]. CT is superior for investigating bony margins of the tumor. However with increasing experience Magnetic resonance imaging is usually preferred to show involvement of the inferior alveolar nerve [6].

Histological pattern may show subtle changes in cellular pattern. An amorphous homogenous eosinophilic, amyloid like material is observed. Many cells can be seen to be filled

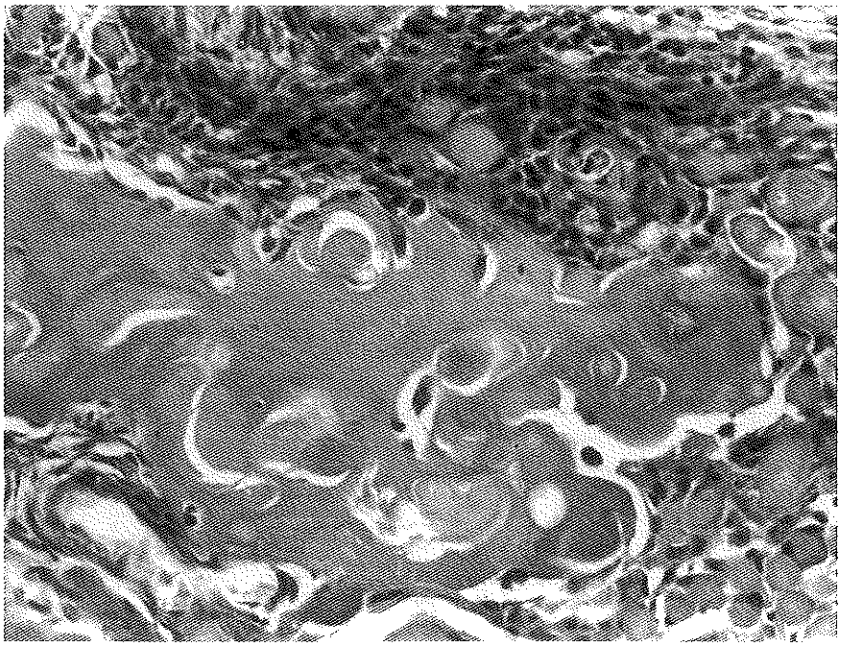
Figure 3. Computed tomography of Case 2 showing multilocular radiolucent area containing radiopaque masses of varying size and opacity.

Figure 4. Abundant homogenous amyloid like material with minor epithelial component (HE x 125).



by calcifying material in the form of concentric Liesegang's rings. Occasionally the lesional cells may exhibit a clear, vacuolated cytoplasm (clear celi variant). It is also considered to be the product of degeneration of celi basal lamina and/or the product similar to the dentine or cement of the tumor cells. A rare case of CEOT devoid of calcification with Langerhans cells is reported. It is suggested that although the majority of the cal-

Figure 5. Small psammomatous calcifications in the epithelial islands (HE x 310).



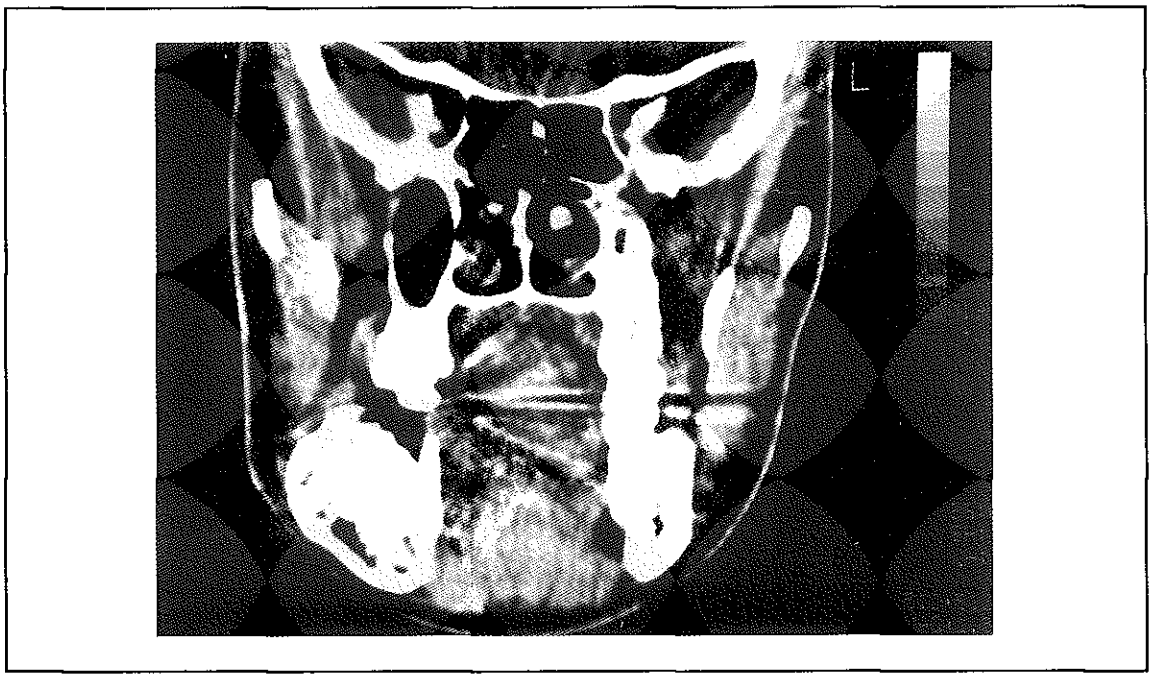
cified lamellar bodies in this tumor represents calcified amyloid, cementum like material may also contribute a part [15].

The clinical presentation of the jaw tumors is very similar. The histopathological examination with an incisional biopsy is essential to decide the treatment and the follow up modality. Adenomatoid odontogenic tumor, calciyfing odontogenic cysts, ameloblastic fibro-odontoma, odontoma and malign tumors of the jaws are the examples for the tumors that should be included in histopathological differential diagnosis [15].

The treatment should be individualized for each lesion because of radiographic and histologic differences from one lesion to another. Surgical management depends on the site and size of the lesion. Franklin and Pindborg reported a recurrence rate of 14%, which was mostly due to the inadequate treatment modality. CEOT's clinical process is similar to solid ameloblastoma however its growth pattern may be slower. Some believe that the two should be treated in similar manner [7]. With its typical pattern containing sheets of eosinophilic cells and amyloid-like material and islands of epithelial cells can be misdiagonosed as ameloblastoma in small biopsy specimen. In the mandible the recommended surgical approach is enucleation with vigorous curettage in the early stages. With more advanced bone infiltration, wide marginal resection should be considered. However CEOT of the maxilla should be treated more aggressively because they grow more rapidly and can impinge on vital structures. Therefore the mean therapy should be maxillectomy. CEOT arising in the maxillar sinus as we presented (Case 1) is extremely rare [3,11].

Reconstruction of the maxilla and mandible is the same as the other jaw tumors. The reconstruction of the maxillary defect without orbital floor and overlying skin is performed by prosthetic devices. Custom made obturators should be prepared by the orthodontist following the operation. In the young pa-

Figure 6. CEOT located on the right mandibular corpus.



tients with high esthetic expectancy and in the patients with extensive tumors crossing the midline of the maxilla or the orbital floor, reconstruction can be achieved by vascularized or non-vascularized bone grafts and soft tissue flaps. The marginal resections in the mandible do not necessarily require reconstruction. The segmental mandibuiar resections require reconstruction. Vascularised or non- vascularized bone grafts can be preferred depending on the defect size, the patient's general health conditions and the patient's age [4,9].

According to Philipsen et al 5 years should be the minimum follow up period to determine the cure rate for CEOT [11]. Basu et al reported a malignant CEOT that showed local tissue invasion and lymph node metastasis [1]. Veness et al also reported CEOT with malignant transformation and metastatic spread. They recommended high dose adjuvant radiotherapy in the postoperative period because of the possibility of multiple recurrences, malignant transformation and metastatic spread [14].

In the literature, CEOT cases have been mostly presented by dental surgeons. Locally aggressive nature and possible malignant transformation of the tumor may require reconstruction following wide excision. Plastic surgeons should be aware of the high recurrent rate of this benign tumor and the follow up period should be longer than the other benign tumors of the jaws.

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