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Case Report

Peripheral and central aggressive form of Pindborg tumor of mandible – A rare case report

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ABSTRACT

Introduction: The calcifying epithelial odontogenic tumor (CEOT), or Pindborg tumor, is a rare and benign odontogenic neoplasm that affects the jaw. Calcifying epithelial odontogenic tumor can be divided into 2 clinico-topographic variants: Intraosseous (central) or Extraosseous (peripheral), with an incidence of 94% and 6%, respectively. The intraosseous tumors have a predilection for the mandible, and most tumors arise in the molar and premolar region of the mandible. When present, extraosseous tumors are often located in the anterior jaw and involve the gingiva.

Purpose: In this article we present a unique rare variety of Pindborg tumor of mandible with both intraosseous & extraosseous presentation and also locally aggressive behavior.

Case report: A 35 year old man presented with Pindborg tumor (CEOT) of mandible with both intraosseous and extraosseous variant in a single lesion, extending from right premolar region to the left molar region.

Discussion: CEOT is an uncommon neoplasm accounting for less than 1% of all odontogenic tumors. Although CEOT is considered less aggressive clinically than typical infiltrating ameloblastoma, but our case is a rarity which was aggressive and also rapidly progressive in short duration, with intra and extraosseous presentation. Hence it was managed surgically similar to a malignant lesion.

Conclusion: Presentation of two different variants of an odontogenic tumor in a single lesion and aggressive behavior of rather slow growing tumor, mimicking malignant lesion definitely shows possibility of rare variations in different magnitude. Hence proper diagnosis for management with better prognosis of these kinds of tumors is very much essential.

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1. Introduction

In 1955, Pindborg¹ reported three cases of a benign but locally aggressive tumor, which he named as calcifying epithelial odontogenic tumor (CEOT), today known as the intraosseous variant of CEOT. The short communication was later followed by two publications,^{2,3} where the author reported 24 cases of intraosseous CEOT and the first two cases of an extraosseous variant. The tumor had, however, been reported prior to 1955 under different names, such as ameloblastoma of unusual type with calcification, calcifying ameloblastoma, malignant odontoma, adenoid adamantoblastoma, cystic complex odontoma and as a variant of the simple ameloblastoma. The term ‘calcifying epithelial odontogenic tumor’ has been generally accepted and adopted by the WHO in the first edition of “Histological Typing of Odontogenic Tumours, Jaw Cysts, and Allied Lesions”, where it was recognized as a distinct entity. For more than 30 years the CEOT has been known popularly as the “Pindborg tumor”.

Although most of these tumors are primarily intraosseous (central), an extraosseous (peripheral) tumor is also known to occur. This tumor was first observed by Pindborg in 1966. Additional extraosseous lesions have been reported; however,

only four of the nine reported cases have exhibited the unique microscopic features of a prominent clear cell component.

Here we present a unique and rare case, reported with both central and peripheral variety of Pindborg tumor in a single lesion of the mandible.

2. Case report

A 35 year old male patient reported to the dept. of oral and maxillofacial surgery, Government dental college, Thiruvananthapuram, Kerala, India, on April 2007, with the chief complaint of a rapidly growing mass in the oral cavity, causing inconvenience while eating, difficulty in speech, and foul smell due to inability to maintain oral hygiene.

Even though pain was not his chief complaint, lesion used to become painful if traumatized, as the lesion was fragile and bleeding was noted sometimes while brushing or while having food.

The lesion was small and asymptomatic to start initially, when patient noticed it 16 months back. In the last 5 months rapid growth was noticed, causing nuisance and discomfort to the patient (Fig. 1a).

Fig. 1 – a. Tumor mass expanding the mandibular labial cortex and exophytic soft tissue proliferation on lingual aspect, with pathological migration of associated teeth. **b.** CT scan axial view of mandible showing the lesion in sections. **c.** Panoramic view of mandible showing the radiolucent lesion with irregular radio-opaque masses inside. **d.** Histopathological view of soft tissue lesion (Hematoxylin-eosin stain; magnification * 400). **e.** Histopathological view of bony lesion (Hematoxylin-eosin stain; magnification * 200).

Clinical examination revealed intraoral pedunculated lesion on lingual aspect of mandible of size $6 \times 4 \times 4$ cm extending from lingual aspect of left second molar to right second premolar, confined to the attached gingiva of alveolus which was fragile and soft with whitish slough on surface. Labial cortex of mandible expanded but was hard to palpate and non-tender, pathological migration of lower central and lateral incisors with slight mobility were seen. There was no parasthesia or altered sensation in the chin or lower lip area.

A differential diagnosis of Pindborg tumor, peripheral giant cell lesion, Ameloblastoma, and Adenomatoid odontogenic tumor was given.

Radiographic examination by an Orthopantomogram (Fig. 1c) and CT scan Axial (Fig. 1b) and Coronal view showed radiolucent lesion extending from distal root of left first molar to mesial side of right first premolar superior-inferiorly extending up to the lower border of mandible. Radiopaque irregular masses were seen in the anterior aspect apical to incisors. Slight resorption of apical third of lower anteriors was also noted.

An incisional biopsy was planned and performed under local anesthesia from the soft pedunculous mass lesion as well as from the bony lesion through the labial cortex of mandible.

Histo-pathological examination of the biopsy specimen of soft tissue (Fig. 1d) and bony lesion (Fig. 1e) revealed characteristic features of calcifying epithelial odontogenic tumor (Pindborg tumor). Hence final diagnosis was made as “peripheral and central variety of Pindborg tumor of mandible”.

Accordingly surgery was planned under general anesthesia for resection of the lesion with minimum of 1 cm margin and

reconstruction using reconstruction plate. Surgery was carried out (Fig. 2a), with lip split incision for a better access and lesion was resected (Fig. 2b) with 1 cm of normal margin, from mesial side of left third molar to mesial side of right second premolar.

Reconstruction with mandibular reconstruction plate, stabilized with 4 screws on right side and 3 screws on left side on lower border of mandible was done, only after achieving occlusion by maxilla-mandibular fixation, by direct wiring. Post-operative recovery was uneventful and patient was discharged on 6th post-operative day (Fig. 2c and 2d).

3. Discussion

CEOT is an uncommon neoplasm accounting for less than 1% of all odontogenic tumors. It is classified between the benign epithelial odontogenic tumors. After being described by Pindborg in 1955, during the next 20 years only approximately 100 cases were reported, until now around 350–400 cases are reported. But occurrence of peripheral and central variety of Pindborg tumor, together in the same lesion is not reported till now.

A histo-chemical study of the CEOT has shown intense alkaline phosphatase activity in the epithelial cells.⁴ The histo-chemical pattern of this activity resembles the alkaline phosphatase appearance that occurs in the stratum intermedium of tooth germs, a finding later confirmed. The clinical differences among the CEOT types may be because of their origin.⁵ Hicks⁶ distinguishes the histogenetic origin of

Fig. 2 – a. Intra operative view. b. Excised mass lesion. c. Post-operative Panoramic radiographic view of mandible showing the reconstruction plate in position with pre-operative occlusion relationship maintained. d. Post-operative intraoral view after 2 months.

Pindborg tumor according to its clinical presentation, that is, central (intraosseous, 87.8%), peripheral (extraosseous, 6.1%), or as a hybrid tumor in combination with adenomatoid odontogenic tumor (AOT). In his opinion, central tumors are derived from the stratum intermedium of the enamel organ. In contrast, the extraosseous form arises from dental lamina epithelial rests in gingival and/or basal cells of the gingival surface epithelium.

CEOT is considered a tumor of adults, with most patients in the age range of 30–50 years. The mean age is slightly greater in central CEOT (40.3 years, 31.8 years in peripheral CEOT). However, there is a wide age range reported from 8 to 92 years.³ There are no differences according to gender in central CEOT. With regard to localization, most cases (68%) occur in the mandible, around the premolar molar region.⁶ Our patient presented with fairly uncommon localizations, in the symphysis. Fifty-two percent of the cases of CEOT are associated with an unerupted or embedded tooth.⁵

The typical clinical presentation is a slowly enlarging intraosseous mass that causes expansion of the affected mandible and is asymptomatic.⁶ Although the CEOT is a benign neoplasm, it has a variable biologic behavior ranging from very mild to moderate invasiveness.⁸ The tumor grows by infiltration and may produce cortical expansion, tooth movement, and root resorption. Peripheral CEOTs are typically smaller than 2 cm. They grow from the gum or from the edentulous alveolar mucosa, generally in the anterior region (incisor-premolar). They are painless and firm, without osseous invasiveness of the maxilla. Nevertheless, the bone adjacent to the tumor typically shows a superficial erosive pattern.⁶ Its behavior is less aggressive clinically than the central one, and therefore the treatment could be more conservative.

Histopathology is probably the most variable aspect of this tumor. CEOTs of central variety are unencapsulated, infiltrating tumors. The characteristic epithelial component consists of sheets and nests of polyhedral epithelial cells with well-defined cell borders, often displaying intercellular bridges.⁶ These neoplastic cells show a moderate degree of pleomorphism, but only rare typical mitoses. Nuclei are centrally located and often contain a large nucleolus. The nuclei may be pleomorphic, hyperchromatic, and bizarre in appearance.⁸ Binucleated cells may be frequent. The most characteristic findings are the presence of amyloid-like substance and calcified concentric Liesegang rings.⁶ Rounded, pale eosinophilic masses (amyloid-like) may be found within the sheets of tumor cells and, because of its affinity with mineral salts, it can undergo calcification, often having the concentric appearance of lamellar bodies or Liesegang rings.⁹ The surrounding tissue may also contain large clumps of this homogeneous eosinophilic material. Calcium salts are often diffusely deposited within these areas. The eosinophilic material has an apple-green birefringence under polarized light after staining with Congo red.⁷ It also stains positive for crystal violet and thioflavine T.⁸ As the globules mineralize, the amyloid-like material loses its positivity for Congo red⁹ and changes from PAS-negative to strongly PAS-positive. Peripheral variety of CEOTs shows an epithelial neoplasm composed of cords and nests of polyhedral cells with an abundant, eosinophilic, granular cytoplasm in certain areas

admixed with regions of cells exhibiting a clear, vacuolated cytoplasm.

The CEOT is generally managed surgically. Because even small CEOTs are infiltrating tumors, treatment should include removal with a border of clinically and radiographic normal bone. A margin of about 1 cm is appropriate. Margins placed too close are likely to be compromised.⁸ Peripheral tumors may be treated with a smaller margin of 0.5 cm, because of its lesser aggressiveness. Although CEOT is considered less aggressive clinically than typical infiltrating ameloblastoma, but our case is a rarity which was aggressive and also rapidly progressive in short duration, hence a minimum of 1.0–1.5 cm normal margin in bone was maintained, and the soft tissue lesion was confined within the boundary of bony lesion, hence no separate margin was required for soft tissue lesion.

It seems that those treated with enucleation and curettage procedures show a recurrence rate ranging from 15% to 30% after just 2–4 years.⁸

An overall recurrence rate of 14% was recorded.⁴ Those treated with resection approaches have few if any recurrences. Therefore, the CEOT is best treated with a resection using 1.0–1.5 cm margins in bone.⁹ A minimum of 5 years and as many as 10 years of follow up may be necessary because of the very slow growth rate of this tumor. Recently, Veness et al¹⁰ reported a case of mandibular Pindborg tumor with malignant transformation and metastatic spread. After initial excision, the surgical margins were involved. The tumor was diagnosed as a CEOT. Afterward, the tumor recurred, and it was re-excised, and the tumor reached the surgical margin again. The histopathologic analysis showed a tumoral evolution, with an increase in cellularity and mitoses and with loss of amyloid-like material and calcification. Invasion of small blood vessels was a new finding. A further re-excision was undertaken, with a supraomohyoid neck dissection. Histopathologic examination confirmed the diagnosis of odontogenic carcinoma and revealed infiltration into skeletal muscle and metastatic spread to 1 level-3 lymph node. In the literature there is only 1 other case of Pindborg tumor with lymphatic metastatic spread before,¹⁰ but in that case there is not a previous diagnosis of benignity.

4. Conclusion

Pindborg tumor shows a characteristic variation in its histopathologic and clinical presentation. The clinical presentation, as can be seen in our case, is related to the location as well as to the size and degree of local invasion. The histopathology presents a wide variety of subtypes, something surprising coming from a tumor so unusual. These circumstances make the diagnosis more difficult. Nevertheless, achieving a correct diagnosis and adequate resection with accurate tumor-free margins for a benign neoplasm with an invasive behavior, is extremely important.

Conflicts of interest

All authors have none to declare.

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