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Case Report

An atypical presentation of Pindborg tumor in anterior maxilla

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ABSTRACT

Pindborg tumor, the eponymous counterpart of calcifying epithelial odontogenic tumor (CEOT), is a rare benign odontogenic neoplasm that was first described by a Dutch pathologist Jens Jorgen Pindborg in 1955 and accounts for approximately 1% of all odontogenic tumors. Its origin as well as its malignant potential is controversial. This neoplasm is possibly of stratum intermedium origin and occurring predominantly in the mandible of adults. We hereby report a case of Pindborg tumor arising in the premaxilla which is seldom (9 out of 200 cases) documented in the scientific literature.

Key Words: Amyloid, anterior maxilla, Pindborg tumor

INTRODUCTION

The calcifying epithelial odontogenic tumor (CEOT) for more than 30 years has been known eponymously as the Pindborg tumor. This is a rare benign odontogenic neoplasm of the jaws. Many names have been given to this lesion.[1] Pindborg was the first to describe this lesion as a separate clinicopathological entity, and he named it as CEOT.[2] Today, nearly 200 cases of this neoplasm have been reported in the literature. The origin of this neoplasm is controversial, though it is generally accepted to have derived from the oral epithelium, reduced enamel epithelium, or stratum intermedium.[3] Pindborg's proposed origin, from reduced enamel epithelium, is plausible because more than half are associated with unerupted or impacted teeth.[2] Chaudry's histochemical studies support the theory that the CEOT arises from the stratum intermedium.[4]

Clinically, CEOT manifests as an intraosseous lesion in the majority of cases, associated with impacted or unerupted teeth.[1,2] There is an almost equal distribution between men and women. The most common radiographic finding is a well-defined unilocular radiolucency, but sometimes the neoplasm may also appear as a multilocular lesion.[1] The histologic criteria for the diagnosis of CEOT are sheets of polyhedral epithelial cells that have well-defined borders and often show prominent intercellular bridges. There is usually pleomorphism of the epithelial cells, and the nuclei and nucleoli are often prominent. A characteristic feature within the sheets of epithelial cells is the presence of amyloid-like material.[5] Some of these cells may also show concentric calcifications which are pathognomonic of this tumor.[3,6] The purpose of this article is to report an unusual CEOT arising in the anterior region of the maxilla of which only around 9 cases have been documented in the English literature.

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CASE REPORT

A 43-years-old male patient reported to the Department of Oral Medicine and Radiology, Bapuji Dental College and Hospital, Davangere with the chief complaint of swelling in the gums in right upper front teeth region for fifteen days. The growth pattern of the tumor was slow and gradual without any associated symptoms. Patient is a known tobacco chewer and an alcoholic since ten years. Extraoral

examination revealed a diffused swelling in the right side of the upper lip extending from the midline to the corner of the upper lip below the ala of the nose and above the upper line of the lip. There was mild obliteration of the nasolabial sulcus. The skin over the swelling was normal with no rise in temperature and non-tender on palpation. Intraoral examination revealed a solitary swelling seen on the upper labial mucosa which is 3 × 2 cms in size extending from right maxillary lateral incisor to right maxillary first premolar involving the attached and the marginal gingiva. The swelling was roughly oval in shape, superoinferiorly involving the mucogingival line and free marginal gingiva. The surface is lobulated, mildly erythematous with well-defined borders [Figure 1]. The radiographic evaluation with an orthopantogram (OPG) showed an ill-defined radiolucency extending from the right upper central incisor to the right upper premolar [Figure 2]. Intraoral periapical radiograph (IOPA) also revealed radiolucency in the periapical region of the right lateral incisor and canine [Figure 3]. A differential diagnosis of central giant cell granuloma was given and an incisional biopsy was sent for histopathological examination. Histopathological examination revealed an epithelial neoplasm composed of discrete sheets, islands and strands of polyhedral epithelial cells in a fibrous stroma [Figure 4]. Cellular outlines were distinct and prominent intercellular bridges were noted. Considerable nuclear pleomorphism was a frequent finding [Figure 5]. Areas of amorphous eosinophilic hyalinized (amyloid like) extracellular material are also present. The tumor islands frequently enclose masses of this hyaline material. Specks of calcifications were present in few areas. Congo red stain revealed intense eosinophilic staining to aggregates of amyloid deposited in fibrous connective tissue stroma.

Based on the clinical, radiographic and histopathological observations, the lesion was diagnosed as CEOT or Pindborg tumor.

DISCUSSION

Pindborg, in 1955, first categorized this tumor as a distinct histopathological entity. Previously, the uncertainty regarding the histological characteristics of CEOT was reflected in the variety of terms for the disease, including unusual ameloblastoma, cystic odontoma, and adenoid adamantinoma. Pindborg tumor is a rare, benign, but locally aggressive odontogenic tumor[1] which accounts for less than 1% of all odontogenic tumors.[2,3] Most investigators believe that the tumor cells originate from the stratum intermedium of the normal dental germ.[4] This idea is based on the morphological similarity of the tumor cells to the normal cells of the stratum intermedium, and high activity of alkaline phosphatase and adenosine triphosphate.[5] Clinically, CEOT manifests as an intraosseous lesion (central type) in the majority of cases (95%). Extraosseous or peripheral lesions account for less than 5% of cases. The latter are usually in the anterior region of the jaws. Most investigators agree that the central type is usually located in the premolar and molar regions, with a mandibular to maxillary ratio of 2 to 1. The majority of CEOTs (52%) are associated with impacted or unerupted teeth. There is an almost equal distribution between men and women. The age of the patients affected by this tumor ranges from 8 to 92 years, with a mean age of 40. This neoplasm occurs in different population groups, with a slight predilection for whites.[1] However, this may merely reflect a reporting bias. The clinical features are usually those of a painless, slow-growing intraosseous mass. However, a tumor in the maxilla may cause pain, nasal obstruction, epistaxis, headache, and proptosis. Our patient gave no history of any of the above symptoms.The most common radiographic finding is a well-defined unilocular radiolucency, which resembles a dentigerous cyst. Our case also mimicked the same unilocular appearance. The neoplasm may also appear as a multilocular lesion mimicking an ameloblastoma. Classically, areas of scattered flecks of calcifications in the central radiolucency may be seen; however, calcifications may sometimes not be evident on radiographs.[1] The histologic criteria listed by Franklin and Pindborg[1] for the diagnosis of CEOT are sheets of polyhedral epithelial cells that have well-defined borders and often show prominent intercellular bridges. There is usually pleomorphism of the epithelial cells, and the nuclei and nucleoli are often prominent. Mitotic figures are rarely seen. A characteristic feature within the sheets of epithelial cells is circular areas filled with a homogeneous substance resembling amyloid-like material, which stains positively with Congo red. Some of these cells are also filled by calcified material in the form of concentric Liesegang rings, which are pathognomonic of this tumor.[3] The histopathologic features in our case also matched the above criteria.

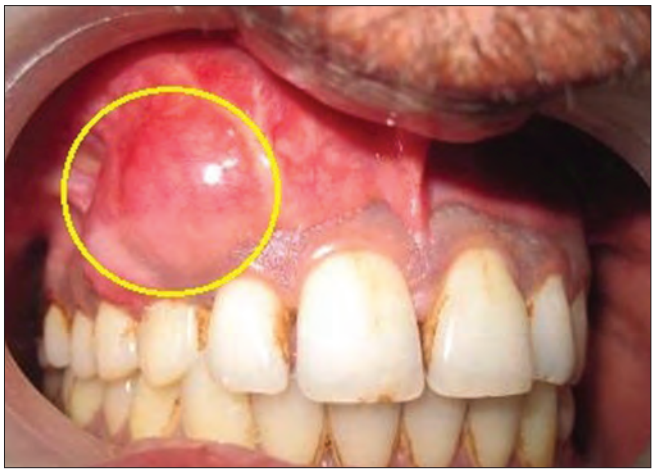

Figure 1: Solitary swelling seen in the right anterior maxilla, obliterating the labial vestibule

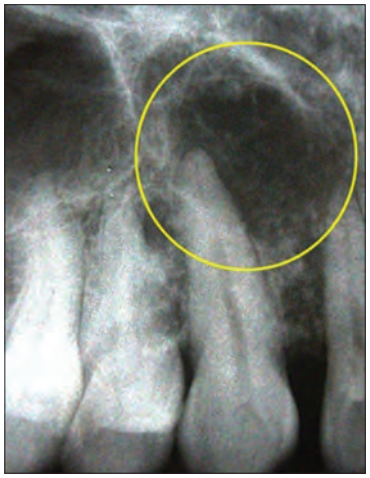

Figure 3: Intraoral periapical radiograph showing radiolucency in the maxillary right lateral incisor and canine region

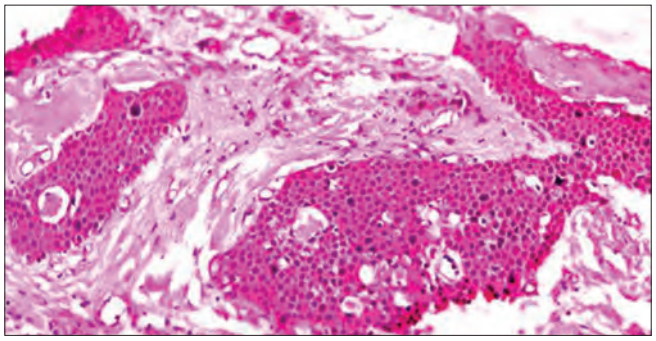

Figure 5: Higher magnification demonstrating neoplastic epithelial cells with prominent intercellular bridges and nuclear pleomorphism. (Hematoxylin-eosin stain; original magnification ×40)

Many variants of CEOT, which may have a bearing on the prognosis and management, have been described. Three such variants reported in the

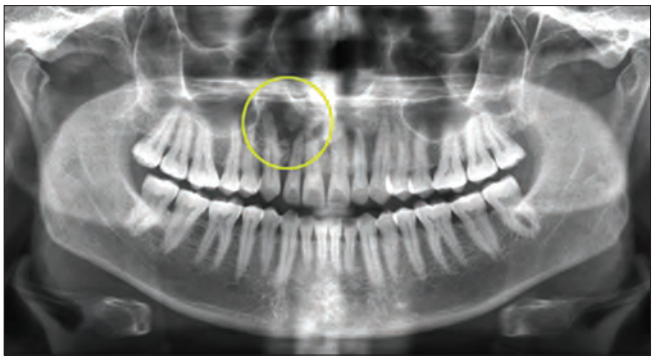

Figure 2: A panoramic radiograph showing a poorly demarcated lytic lesion between right maxillary lateral incisor and canine

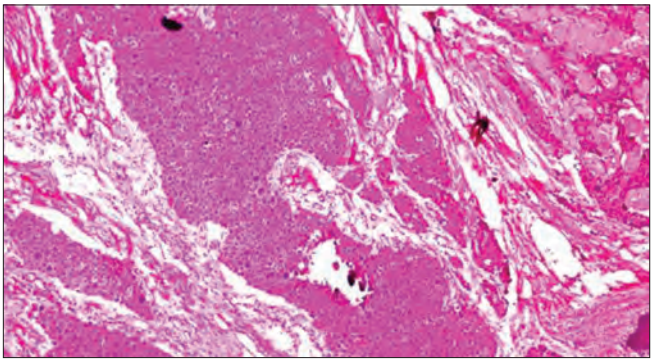

Figure 4: Pindborg tumor exhibiting sheets of polyhedral epithelial cells with evidence of calcification and amyloid. (Hematoxylin-eosin stain; original magnification ×10)

English literature are the noncalcifying CEOT with Langerhans cells, the CEOT displaying cementum-like and bone-like material, and the clear-cell CEOT.[7] The former variant of CEOT is histologically similar to the peripheral type of CEOT but is devoid of calcifications, and one may speculate that this variant's clinical behavior would be less aggressive than the peripheral lesion.[7] It has been proposed that CEOTs with more amyloid and calcifications could be treated less aggressively.[6]

CEOTs with large amounts of bone-like or cementum-like material probably indicate a higher level of differentiation and thus may account for their more self-limiting behavior, unlike the ameloblastoma.[8] The clear-cell CEOT variant is more aggressive with a higher recurrence rate (22%), and some would consider this form to be a low-grade odontogenic carcinoma.[9] In 1984, Basu *et al*. reported a malignant CEOT that showed evidence of local tissue invasion and regional lymph node metastasis.[6] There have also been reports of various other odontogenic lesions occurring in association or in combination with CEOT. These include dentigerous cysts and adenomatoid odontogenic

tumors as well as presentation of CEOT as an intramural lesion of the dental sac.[6]

CONCLUSION

Thus, we conclude that this was an atypical presentation of CEOT as it was seen in the anterior maxilla. Hence oral pathologists should add this neoplasm in the list of differential diagnosis for tumors in the anterior maxillary region as CEOT of the maxilla should be treated more aggressively, as maxillary tumors tend to grow more rapidly than their mandibular counterpart and do not usually remain well confined.

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