Case Report

Calcifying epithelial odontogenic tumor (Pindborg tumor)

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ABSTRACT

The calcifying epithelial odontogenic tumor (CEOT) is a rare entity and represents less than 1% of all odontogenic tumors. Dr. J J Pindborg (1958) first described four cases of this unusual lesion; subsequently Shafer *et al* coined the term Pindborg tumor. This lesion is a locally aggressive benign odontogenic neoplasm arising from epithelial tissue. It occurs most commonly in 4th-5th-6th decade of life and bears no gender predilection. A case of CEOT in a 50-year-old male arising in the left body region is described.

Key words: CEOT, Pindborg tumor, Hemimandibulectomy

Introduction

The calcifying epithelial odontogenic tumor (CEOT) was first introduced into scientific literature almost 50 years ago by Dr. J J Pindborg.[1,2] World Health Organization in 1992 classified it as a benign odontogenic tumor, which is exclusively epithelial in its tissue of origin. The differential diagnosis for CEOT should include adenomatoid odontogenic tumor (AOT), calcifying odontogenic cyst (COC), ameloblastic fibro odentoma (AFO), odontoma.

Case Report

A 50-year-old male reported to the oral and maxillofacial surgery OPD with a swelling in left body of mandible measuring 8 × 10 cms extending from tooth nos. 32 to 37. The swelling was tender and had progressively increased over a period of two years leading to facial asymmetry. Orthopantomograph [Figure 1] revealed a mixed radiolucent-radiopaque lesion, which was multilocular with coarse trabeculae and scattered foci of calcification extending from left lower canine to first molar region with a radio opaque mass representing embedded premolar. The provisional clinical diagnosis of ossifying fibroma, calcifying epithelial odontogenic tumor, ameloblastoma and odontogenic myxoma was made. An incisional biopsy from the region involved was sent for histopathological examination. Histopathology findings revealed a neoplasm composed of cells arranged as sheets and anastomosing small and large islands. These cells were interspersed by prominent homogeneous hyaline acellular material. Areas of concentric lamellated calcifications were seen. The neoplastic cells have well defined cell borders, abundant eosinophilic cytoplasm and hyperchromatic mildly pleomorphic nuclei, few bizarre nuclei were seen, however no abnormal mitosis was seen. Normal mature lamellar bony trabeculae were seen between tumor islands interspersed with large areas of haemorrhage. The eosinophilic material was confirmed as amyloid upon special staining, diagnosing the lesion to be CEOT.

A presurgical computed tomography (CT) scan [Figure 2] was obtained to ascertain the boundaries of the neoplasm. It [Figure 3] revealed an osteolytic lesion with foci of calcifications. In view of the extensive involvement, resection of the involved portion of the mandible and reconstruction with an AO Unilock 2.4 mm reconstruction plate was planned. The mandible was exposed via an extraoral approach and resection of the left hemimandible from 32 to 37 with safe clinical margins was performed and defect was bridged by 2.4 mm unilock reconstruction plate [Figure 4]. Resected specimen [Figure 5] was dark brown to grey in color surface measuring 7.5×5.5×4 cms. Specimen was submitted for histopathological examination where it was confirmed as CEOT.

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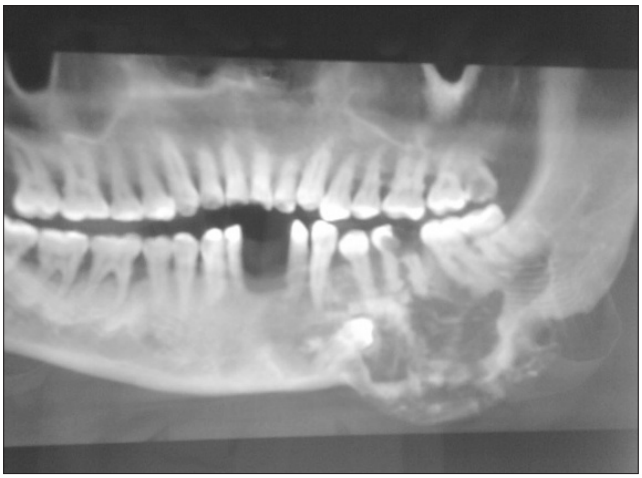

Figure 1: Orthopantamograph revealing large osteolytic lesion

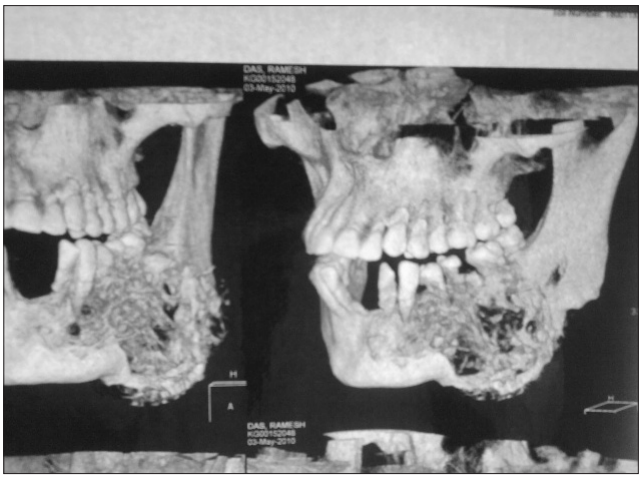

Figure 2: A 3D computed tomography (CT) image of the lesion

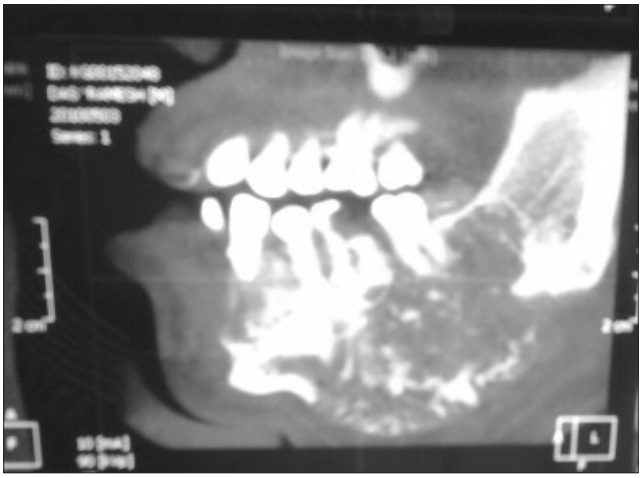

Figure 3: CT scan depicting the large involvement of body of mandible

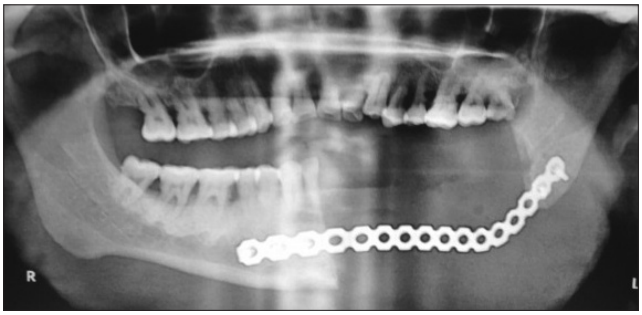

Figure 4: Orthopantamograph revealing reconstructing plate bridging the segmental defect

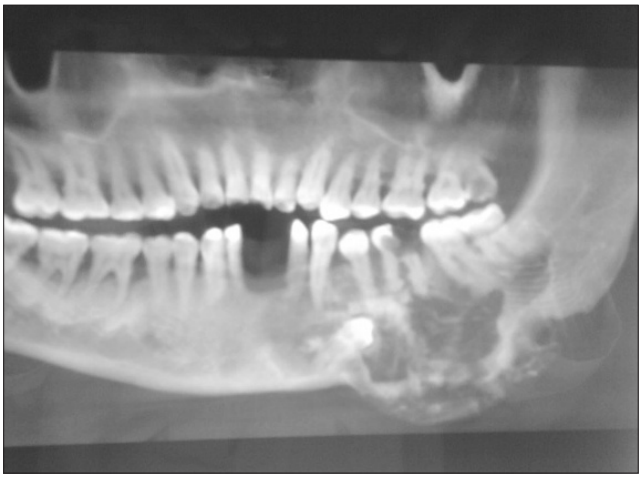

Figure 5: Gross specimen

Discussion

CEOT/ Pindborg tumor is a rare benign but locally aggressive tumor. Neville and colleagues, in their textbook of oral and maxillofacial pathology, assert that the lesion is a distinct entity and probably represents less than 1% of all odontogenic neoplasms.

There are differences of opinion within the oral and maxillofacial pathology community regarding degree of differentiation of the odontogenic epithelium which forms the basis for tumor pathogenesis. Some authors suggest that the epithelial cells of the Pindborg tumor are reminiscent of the sequestered cells in the stratum intermedium layer of the enamel organ.[3-5] This idea is based on the morphological similarity of the tumor cells to the normal cells of stratum intermedium and a high activity of alkaline phosphatase and adenosine triphosphate.[6] It has been suggested that amyloid deposition within Pindborg tumor is an immunologic response to these stratum intermedium cells. Others insist that it arises from remnants of the primitive dental lamina found in the initial stage of odontogenesis, and these epithelial rests are the more likely true progenitor cell.[7] However, the exact etiology is unknown.

CEOT occurs most commonly between 20 to 60 years of age with mean around 40 years. In 113 cases reviewed by Franklin and Pindborg,[8] patients ranged from 8 to 92 years of age with mean at 40 years. In 2004, Cicconetti and colleagues reported that tumor more frequently

affects adults in the age range of 40 to 60 years with peak incidence in the 5th decade with an equal sex distribution.[9]

Ninety-four percent of the lesions are central and intraosseous and 6% are extraosseous. Intraosseous CEOT shows a maxilla-to-mandible site ratio of 1:2 and is mainly located in the premolar/molar region. Half of the cases are associated with an impacted tooth.[7] Fifty-two percent cases have been associated with unerupted or embedded tooth. Similar finding was present in the case reported as premolar was embedded. CEOT may lead to tooth tipping, rotation, migration, and/or mobility secondary to root resorption. This lesion is often symptomless and discovered on routine radiography. Alternatively it may present symptomatically as a slow-growing, painless, expansile, bony swelling with cortical bone resorption and finally perforation, as was seen in the case reported.

In the initial stage, it is totally radiolucent, simulating a dentigerous cyst because of its relation with impacted tooth. Small intratumoral calcification starts appearing in the second phase, which is characteristic but not diagnostic. The final stages are associated with osseous destruction and the tumoral calcification giving it a honeycomb appearance. Quite similarly the radiographic finding in our case showed a loculated/trabeculated honeycomb mixed radiolucent radiopaque finding. The diagnosis of CEOT is also based on histopathological examination revealing polyhedral neoplastic cells, which have abundant eosinophilic finely granular cytoplasm with nuclear pleomorphism and prominent nucleoli. Most of the cells are arranged in anastomosing sheet like masses. An extracellular eosinophilic homogenous material staining like amyloid is characteristic of this tumor with concentric calcific deposits called Liesigany Ring.[5] The case described also depicted calcific foci in abundance with fused amorphous calcified aggregates.

Treatment

Numerous surgical treatment modalities have been suggested, and the treatment plan is dependent on multiple factors such as size and location of neoplasm, general condition of patient and operator skill. Small, intrabony mandibular lesions with well-defined borders are treated by simple enucleation or curettage followed by judicious removal of a thin layer of bone adjacent to the tumor.[6] Large tumors require aggressive approach by segmental resection, hemimandibulectomy and hemimaxillectomy, which causes bone discontinuity requiring reconstruction procedures such as grafting or distraction osteogenesis.[3,4,6] Recurrence rate of 10–20% following conservative treatment is reported.[5,9] Malignant transformation and metastasis is rare.[9] Patient reported here underwent hemimandibulectomy and there has been no recurrence on 6 month follow-up.

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