

12 Year Pediatric Patient Having Pindborg Tumour an Intraosseous Variant: A Rare Case Presentation Affecting the Maxilla

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Abstract

Pindborg tumor is a rare benign epithelial odontogenic tumor occurring most frequently in the posterior part of the lower jaw. I herein report a case of a 12 year old pediatric male patient with a painless swelling of the left maxilla of 5 months duration. Radio graphically, a well circumscribed unilocular radiolucency containing radiopaque foci associated with impacted tooth was seen, along with cortical bone expansion. Histopathology showed sheets of polyhedral epithelial cells in a fibrous stroma. Cells showed distinct cellular outlines with prominent intercellular bridges. Large areas of amorphous, eosinophilic, hyalinized material seen and calcifications within the amyloid-like material (Liesegang ring calcifications) were evident, confirming a diagnosis of Pindborg tumour (Calcifying Epithelial Odontogenic Tumor). The patient was treated by surgical excision of the tumor.

Keywords: Pindborg tumor; CEOT; Liesegang rings; Maxilla

Introduction

Pindborg's tumor-calcifying epithelial odontogenic tumor (CEOT) is a rare tumor. It was first described as a separate pathologic entity by Dutch pathologist Jens Jorgen Pindborg in 1955. The term "Pindborg's tumor" was first used by Shafer et al. [1]. According to the WHO classification, Pindborg's tumor is a benign odontogenic tumor, most frequently presenting as a painless slow growing swelling [2,3] but with local aggressive behavior and a tendency to recurrence [4]. It represents only about 1% of all odontogenic tumours [2]. It has been identified under different denominations, such as, ameloblastoma of unusual type with calcification, calcifying ameloblastoma, malignant odontoma and cystic complex odontoma [5]. Two varieties have been described according to their location: Intraosseous (94% of cases) and extraosseous (6% of cases) [6]. About 65% of all reported cases have occurred in the mandible, most often in the posterior part [3]. I, here in, report a rare case of Pindborg's tumor involving the maxilla of a 12 year old pediatric male patient.

Case Report

A 12 year old pediatric male patient reported to Kailash hospital, Noida, with the complaint of a slow growing painless swelling over the left cheek region since 5 months. Patient revealed in history that the swelling started almost 5 months back and had gradually increased to attain the present size. It was not associated with discharge and numbness. His past medical and dental history were non-contributory. General examination revealed a moderately built and nourished individual of normal gait with vital signs within the normal range. Extra oral inspection revealed an oval-shaped swelling present over the left maxilla, 6 cm 4 cm in size (Figure 1).



Figure 1: Well-defined swelling in the maxillary left vestibular region causing obliteration of the buccal vestibule.

Skin over the swelling appeared normal with no secondary changes. On palpation, the swelling was non-tender, hard in consistency, no localized rise in temperature was noticed, non-fluctuant. Intraoral examination revealed a well-defined swelling in the maxillary left vestibular region (Figure 2), extending from the mesial surface of 23 to the distal surface of 27, causing obliteration of the buccal vestibule. Mucosa overlying the lesion appeared normal and intact and teeth in the vicinity showed no mobility, discoloration, tenderness and responded positive to the vitality tests.



Figure 2: Resected specimens showing a soft tissue mass enclosing crown of an impacted tooth.

Panoramic radiograph revealed a unilocular radiolucency in the left maxillary region, showing a sclerotic border and containing radioopaque foci and an impacted tooth. Differential diagnosis included dentigerous cyst, odontogenic keratocyst, calcifying odontogenic cyst, calcifying epithelial odontogenic tumor, and Adenomatoid odontogenic tumor and ameloblastic fibroma. Surgical excision of the lesion was advised along with removal of the impacted tooth. The lesion was excised in to as a single mass (Figures 2 and 3).

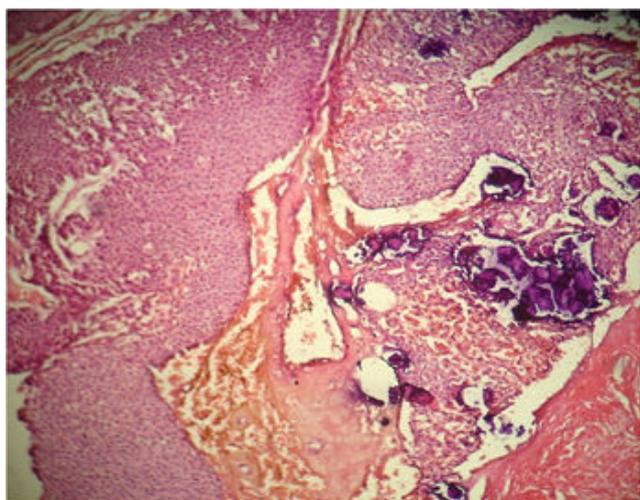


Figure 3: Section showing a fibrous capsule enclosing sheets of polyhedral epithelial cells (H and E, X 10).

Gross pathology showed a soft tissue specimen enclosing crown of a tooth, measuring 6 cm × 3.5 cm in size, brownish red in colour, with

smooth surface and firm in consistency. Apical portion of the root of impacted tooth was seen emerging out of the specimen (Figure 4). Histopathology showed a fibrous capsule enclosing sheets of polyhedral epithelial cells in a scanty fibrous connective tissue stroma (Figure 4).

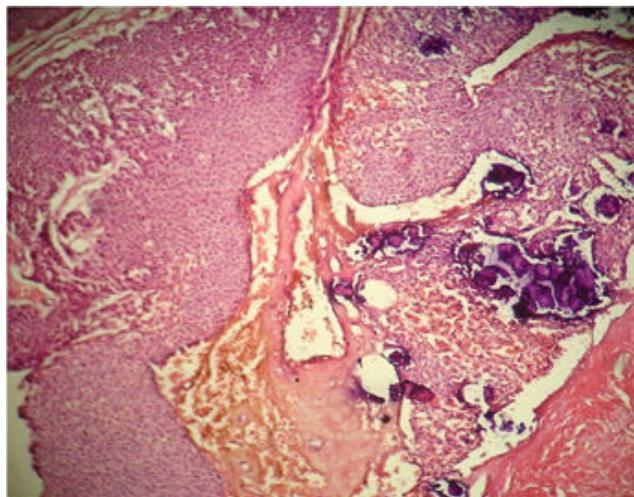


Figure 4: Epithelial cells with distinct cellular outlines showing prominent, hyperchromatic, pleomorphic nuclei (H and E, X 40).

Epithelial cells showed distinct cellular outlines with prominent intercellular bridges and abundant eosinophilic cytoplasm. Nuclei were prominent, hyper chromatic, and large in size and pleomorphic in appearance (Figures 4 and 5).

Large areas of amorphous, eosinophilic, hyalinized material seen and calcifications within the amyloid-like material (Liesegang ring calcifications were seen in between sheets of epithelial cells and also in the connective tissue (Figure 5), confirming the diagnosis of calcifying epithelial odontogenic tumor. Postoperative period of the patient was favorable. His wounds were checked on an outpatient basis. A 24-month follow-up revealed no evidence of recurrence of the lesion.

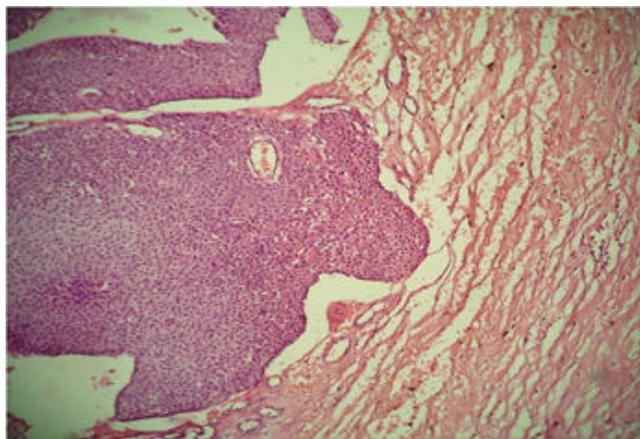


Figure 5: Amorphous, eosinophilic, amyloid-like material and liesegang ring calcifications in the connective tissue (H and E, X 10).

Discussion

Pindborg's tumor (CEOT) is a rare, benign, but locally aggressive tumour, which accounts for less than 1% of all odontogenic tumours [1,2]. The origin of this neoplasm is not clearly known, although it is generally accepted to be derived from reduced enamel epithelium, stratum intermedium or dental lamina remnants [7,4]. For other authors, it would derive from the cellular remnants of the basal sheet or from the basal stratum of the gingival epithelium (what would justify the existence of the peripheral or extraosseous forms) [4].

In reported cases that mentioned race, the great majority of patients were Caucasians. There have only been few reported cases in blacks [8]. Most cases involve the posterior mandible; there have been few reported maxillary cases [2-4,7,9,10]. Although its biological behavior is relatively indolent, maxillary lesions tend to grown rapidly and not be circumscribed [9]. There is no sex predilection [6]. The age distribution is wide (15-75 years) [10] with a peak in the fourth to fifth decade [7]. The rarity of this case is the occurrence of CEOT in the maxilla of a young adult. In our case patient was 12 year old and site was maxilla.

It manifests as asymptomatic swelling that causes slow bone expansion [4,7]; associated with impacted tooth [5], unerupted tooth [5-7,10,11] or odontoma [11]. Bimaxillary and bilateral involvement has also been described [6]. Sedghizadeh has reported a multifocal variant of Pindborg's tumor [12]. The tumor grows by means of infiltration and there can be expansion of the cortical bone, dental movement and root resorption. In some cases there may be pain, nasal obstruction, epistaxis, headaches and even bleeding. In the maxilla, the sinuses may be affected [6]. It is rarely associated with paresthesia [7]. This case manifested as a painless, slow-growing intraosseous swelling associated with an impacted tooth.

It is common to discover this tumor in a routine way starting from an occasional radiological exploration [4]. Radiographically, CEOT generally appears as well delineated areas of radiolucency often associated with an unerupted tooth [13]. Radiologically, the tumor has different stages of development. Initially the tumor is radiolucent, like an odontogenic cyst (especially when it is by an unerupted tooth) or

like a cystic ameloblastoma [6]. In the second phase, small intratumoral calcifications appear which the most characteristic image of the lesion is although never pathognomonic [4]. Finally, an irregular pattern is displayed, with uni- or multilocular areas, giving a honeycomb [4,6] or 'driven-snow' appearance [13], due to osseous destruction and tumor calcifications [4,6]. Locularity and opacity result from 'maturation' of the tumour [2]. The tumor tends to be well-circumscribed radiologically in spite of having sclerotic margins that may not be visible [6]. Panoramic radiograph of this patient revealed a unilocular radiolucency in the left maxillary region, showing a sclerotic border and containing radiopaque foci and an impacted tooth.

The histomorphologic pattern of the CEOT consists of scanty connective tissue stroma [8] that supports sheets, nests and masses of polyhedral epithelial cells [11] with well-defined borders and prominent intercellular bridges [6]. The cytoplasm is abundant and eosinophilic. The cells may show cellular abnormalities including cellular pleomorphic [6] and giant cell formation [11]. The nuclei tend to be prominent, with a great variety of sizes, shapes, numbers [6] and staining quality, producing a pleomorphic appearance [8]. Mitosis tends not to be seen [6,14]. Some cells increase in size and produce a homogeneous, eosinophilic substance which may be liberated as the cells break down [11]. The true nature of this homogeneous, eosinophilic material is still unresolved [11] and has been variously described as amyloid, comparable glycoprotein, or keratin or enamel matrix [8]. This amyloid type material shows birefringence of an apple-green color under polarized light after Congo red staining [6]. A very important finding is concentric calcification in the amyloid material that forms Liesegang rings, which are pathognomonic for this tumor [1,6]. These small, usually round calcifications with Liesegang rings are present among the epithelial cells and also in the connective tissue [8]. All the characteristic histopathological features were present in this case, confirming the diagnosis of calcifying epithelial odontogenic tumor.

Histological variants of CEOT include CEOT with cementum-like components, clear-cell CEOT, CEOT-containing Langerhans' cells, combined epithelial odontogenic tumour (CEOT/AOT) and CEOT with myoepithelial cells [11].

The differential diagnosis should include ameloblastoma, myxoma, aneurismatic bone cyst and odontogenic cysts, depending on the radiographs. Giant cell granuloma should also be ruled out, together with cemento-ossifying fibroma, ameloblastic fibro-odontoma and ameloblastic fibroma. If there is considerable bleeding the differential diagnosis should include central angioma of the mandible. If there are clear cells, the clear cell odontogenic carcinoma should also be considered together with clear cell ameloblastoma, intraosseous carcinoma, salivary gland neoplasms and aggressive osteoblastoma [6].

Treatment of CEOT involves surgery. Surgical management of CEOT varies depending on the size and site of the tumor as well as the extent of bone destruction [3,8]; with removal of the embedded tooth and any associated soft tissue mass [13]. Tumor-free surgical margins should be obtained to reduce the risk of local recurrence [13].

Once correctly identified, mandibular CEOT can be adequately treated by conservative resection, such as enucleation or curettage, including a margin of intact bone, though prolonged follow-up of the patients is advised in view of possible recurrences even after prolonged time intervals [10,11,8]. CEOT involving the maxillary bones or showing a prominent clear cell component should be treated more

aggressively in consideration of their purported more aggressive potential [8-11]. It has been noted that the more amyloid tissue and calcification there is, the less aggressive is the lesion [8].

The prognosis of the CEOT is good with infrequent recurrence [5,10]. The primary CEOT has a recurrence rate of 10-15% [6,13,14], after total excision, and its malignant transformation is a very rare occurrence [5,15]. Although it has not been established in the literature, five years should be the absolute minimum follow-up necessary to assess the healing for this type of odontogenic tumor [5,9,11].

Summary

Pindborg tumor is a rare benign lesion. This report adds to the small number of Pindborg's tumor cases located in the maxilla, since over 2/3 of CEOT case have been described in the posterior portion of the mandible, usually in 3rd to 4th decade of life. Presence of this odontogenic tumor in a young patient, involving a maxillary site is the particularities of this case. Given its unpredictable behavior and its unspecific radiological findings, carrying out an anatomopathologic study is necessary, as is resective surgery in order to minimize the probability of recurrence. Also necessary is a postoperative follow-up of at least five years.

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