Pindborg Tumour: A Rare Odontogenic Neoplasm

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Citation

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Abstract

Calcifying epithelial odontogenic (Pindborg) tumour is a rare benign odontogenic neoplasm of unknown etiology. These are slow growing but locally aggressive in nature. These tumours have a characteristic histological appearance revealing amyloid like substance present as eosinophilic globules. The true nature of this substance is still under investigation. We briefly describe two cases of Pindborg tumour diagnosed on routine histopathological examination.

INTRODUCTION

Pindborg (calcifying epithelial odontogenic) tumour is a benign neoplasm which was first described by Pindborg (1955) as a distinct clinicopathological entity. These are uncommon neoplasms accounting for approximately 1% of all intraosseous odontogenic neoplasms. These tumours are thought to arise from stratum intermedium of the normal dental germ ([1,2]). Calcifying epithelial odontogenic tumour (CEOT) are generally considered benign but can be locally aggressive in nature. Occasional case reports of its malignant transformation and metastatic spread had also been reported([2]). Most often, they arise centrally in the mandible around the premolar-molar region of posterior mandible but 10% may occur at extraosseous site. These tumours are characterized by the presence of squamous cell proliferation, calcified rings and notably amyloid or amyloid like deposits ([1]). We present two cases of Pindborg tumour diagnosed on routine hisotpathological examination.

CASE REPORTS

CASE 1

A 22 years female presented with swelling right side of face since last 2 years which was gradually increasing in the size. She had a history of tooth extraction. Clinical examination revealed a diffuse, irregular hard swelling on right lower mandible. On radiological examination, orthopantomogram (OPG) showed a mixed radiolucent and radioopaque areas in right body of mandible in relation to 6th molar teeth. Roots of 5th and 7th molar teeth were displaced with lesion extending from alveolar crest sparing lower border of mandible (Fig. 1). Computed Tomography (CT) revealed a mixed lytic expansile lesion with few calcified areas within the body of right mandible in relation to root of 6th molar teeth. The patient was subjected to excisional biopsy. The histopathological examination revealed strands and sheets of polyhedral epithelial cells with eosinophilic cytoplasm and pleomorphic nucleus having variable degree of hyperchromasia(Fig.2). Numerous homogenous spheroidal globules of varying sizes surrounded by calcified rings (i.e. Liesesang ring) were seen within the tumour epithelium and intervening stroma (Fig.3). On Congo-red stain, these structures gave apple green birefrigerance under polarized light. A histopathological diagnosis of calcifying epithelial odontogenic tumour (Pindborg Tumour) was made. The patient is under followup for any recurrence.

CASE 2

A 40 year male presented with swelling and pain in left mandible around the molar region since last eighteen months which was of gradual onset. On examination, there was an irregular firm to hard, mild tender, pinkish mass in upper molar area which bled on touch. The radiological examination revealed a lobulated radiolucent area with multiple radioopacities. CT revealed an osteolytic lesion in the left maxilla opposite 1st, 2nd and 3rd molar teeth with few flecks of calcification. There was expansion of overlying cortex with cortical erosion at places (Fig. 4). A clinical diagnosis of giant cell granuloma was made. An incisional biopsy was performed and sent for the histopathological examination. Microscopic examination revealed polyhedral epithelial cells arranged in sheets and strands exhibiting nuclear pleomorphism and variable hyperchromasia but no mitoses. There were numerous homogenous, calcified bodies (Liesesang ring) which stained positive for congo-red. A histopathological diagnosis of

calcifying epithelial odontogenic (Pindborg) tumour was rendered. Later, the lesion was completely excised and the diagnosis was confirmed on histopathology. The patient is on regular follow up since then without any signs of recurrence.

Figure 1

Figure 1 : OPG showing a mixed radiolucent and radioopaque areas in body of mandible

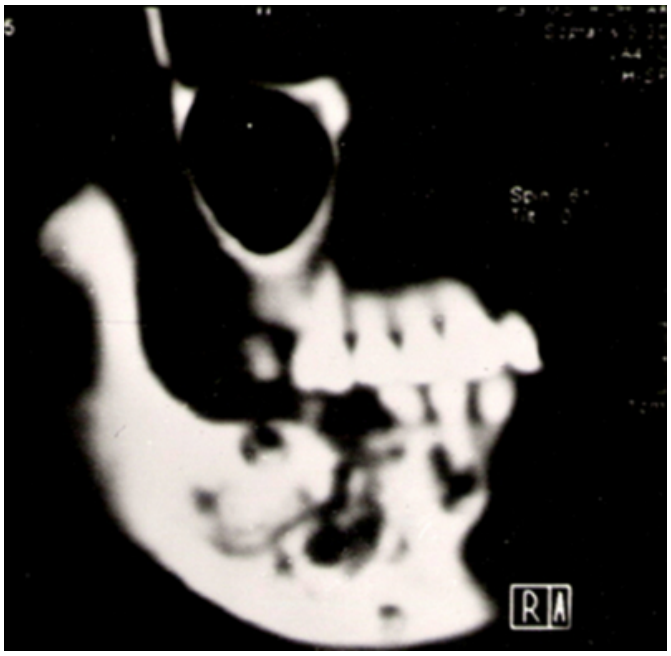


Figure 2

Figure 2 : Photomicrograph showing sheets and strands of tumour cells with calcified laminated globules (H&E 100x)

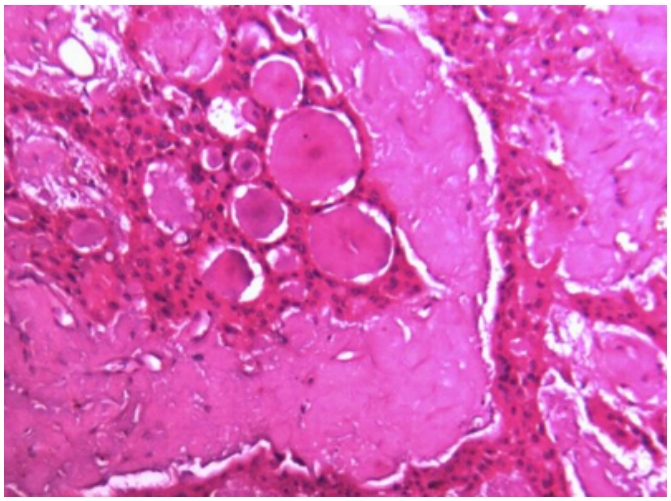


Figure 3

Figure 3: Photomicrograpgh showing Liesesang ring (H&E 400x)

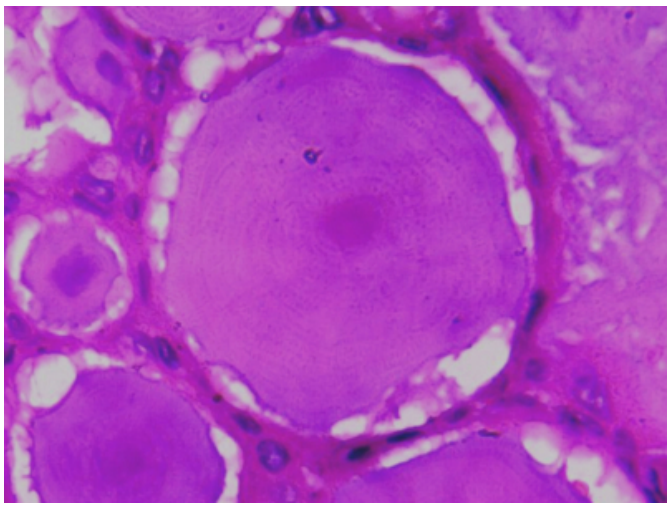
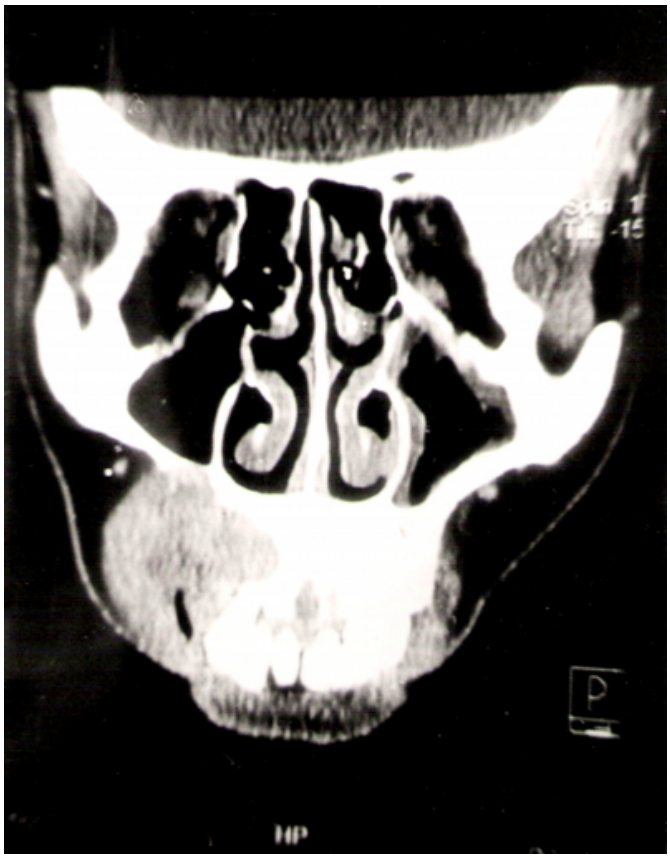


Figure 4

Figure 4: CT revealing an osteolytic lesion in the left maxilla.



DISCUSSION

Pindborg tumour is an uncommon odontogenic neoplasm of unknown etiology related to the odontogenic apparatus. However, most of the cases of intraosseous variant are

associated with an unerrupted or embedded tooth (odontomes). The lesions commonly manifests in an age group of 20-60 years with a peak incidence in the fourth and fifth decade of life with a mean age 40.3 year for intraosseous and 31.8 years for extraosseous variants. The intraosseous variants (94% cases) of CEOT have an almost 1:1 gender ratio, while the extraosseous (6% cases) tumours are twice as common in women ([1,2]). The patient may be aware of a gradually increasing painless mass at the time of diagnosis. Symptoms like nasal stuffiness, epistaxis and headache may be evident in some of the patients ([3]). Although these tumours are thought to be benign, local tissue infiltration had been documented with a recurrence rate ranging from 10% to 15% ([2]). A few cases of CEOT with malignant transformation and metastatic spread had also been reported in the literature ([2]). Variable radiographic features which are characteristic of this lesion appears as a diffuse or well circumscribed unilocular/multilocular radiolucent area with honeycomb pattern. Multiple radioopacities of varying size with areas of calcification may also be evident. However, Magnetic resonance imaging and CT are not of primary diagnostic importance as a histopathological diagnosis is more precise and easy. Radiological findings had been beneficial in the surgical planning of such cases ([5,6]). Treatment in most of the cases is enucleation of the mass with tumour free surgical margins to prevent recurrence. Adjuvant radiotherapy had also been recommended in the cases with malignant transformation and metastatic spread ([2]).

The histological characteristics of CEOT are well delineated. Variation in nuclear size, hyperchromasia, occasional multinucleate forms of epithelial cells, intracellular bridges are commonly encountered in this neoplasm. However, the most characteristic feature is the presence of homogenous eosinophilic globules with liesesang ring phenomenon ([2]). These globules react with conventional stains of amyloid but differs from amyloid at the ultrastructural level. Immunohistochemically this amyloid is positive for a cocktail cytokeratins([1,5,6,8,13,16]) and cytokeratins AE1 and AE3 but negative for basement membrane components ([3]). On aminoacid sequencing and mass spectrometry these fibrils of amyloid like substance were found to be composed of polypeptide of approximately 46 mer ([7]). However, others are of opinion that this substance is derived from filamentous degeneration of keratin filaments that originate from the tumour squamous epithelium ([3]). A proportion of amyloid negative and non-calciying variants of Pindborg tumour had also been reported. Other types of odontogenic tumour like ameloblastomas, ademato id odontogenic tumours and ameloblastic odontomas may also contain amyloid or amyloid like substances and needs to be differentiated from these tumours ([8]). Several cellular variants like clear cell, pigmented, Langerhans cell containing, bone and cementum forming, and non-calcifying subtypes of CEOT too have been identified. The clinical signifincance of these cellular variants is still unclear ([4]).

We describe two classical cases of Pindborg tumour diagnosed on routine histopathological examination, one of which presenting at an quite early age with the review of the clinicopathological aspects of this rare entity.

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