Sellar Region Keratinizing and Calcifying Odontogenic Cyst: Case Report

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Introduction

The calcifying odontogenic cysts (COC), first reported by Gorlin et al in 1962 (Gorlin et al. 1235-43) are uncommon and represent about 2% of all odontogenic cysts and tumors (Swan, Houston, and Moore 340-43). Several clinical and histological features have been reported [4, 5]. According to the World Health Organization (WHO) COC has been classified within the group of neoplasms and tumors that originate from odontogenic tissues (Kumamoto, Yoshida, and Ooya 171-76)Normally COCs appear as painless, slow-growing tumors that affect young adults and involved mainly the anterior segment of the maxilla and mandible (Buchner 330-39)

The keratinizing and calcifying odontogenic cyst (KCOC), a lesion that features proliferating ameloblastic epithelium, ghost keratin, calcification and cyst formation may more precisely mimic

the craniophyrgioma. Some believe that keratinizing and calcifying odontogenic cyst and craniopharyngioma are ameloblastoma closely related developmentally (Paulus et al. 172-76).The origin of craniopharyngioma and KCOC is from buccal equivalent of the embryonic enamel organ present in Rathkes pouch. It appears that ghost cells occur in both craniophyrgioma and KCOC, however, stellate cells are found in craniophyarngioma and ameloblastoma but not in keratinizing and calcifying odontogenic cyst (Hong, Ellis, and Hartman 56-64). The differential diagnosis of suprasellar KCOC includes an epidermoid cyst which is lined by stratified squamous cells with keratohyaline granules which are characteristic of epidermal cysts and again not seen in the present case

Case history

A five and a half year old female child presented because of continuing and progressive headache that has lasted for two years. Two months prior to presentation, the child developed visual deterioration that limited her daily activities. This was followed by unsteady gait, mild febrile illness and deteriorating level of consciousness.

On presentation, the child was conscious however depressed. The other higher functions were within normal. Fundal examination showed bilateral dilated pupils with papilloedema. The lower limbs were spastic and hypertonic with brisk ankle and knee jerks. The head circumcuference was 54 cm. Oral examination revealed swelling of the mandible and multiple small and malformed tooth like structures (Fig 1). Radiograph of the mandible showed radiolucent areas with widely scattered tooth-like structures (Fig 2)

Abdominal examination showed distended abdomen, full flanks and everted umbilicus with positive shifting dullness indicating ascites, but no palpable enlarged organs or masses (Fig 3).

MRI brain showed multiloculated cystic lesion in the sellar and parasellar region with heterogeneous intensities (Fig 4). Pituitary hormonal profile was within normal range. T3 3nmol\l, T4 104 nmol\l, TSH 2.3 mu\l.CBC was within normal range and ESR 30mm\h. The renal profiles were normal. Ultrasound abdomen, a part of massive ascites did not reveal any enlarged organs or masses. Ascetic fluid was obtained and sent for cytology and this did not reveal malignant cells.

The patient underwent surgery through Lt pterional approach. A huge cystic lobulated lesion with multiple calcifications and keratin like debris was encountered. The basal major cerebral vessels namely the internal carotid and its bifurcation were found within the cyst. Tiny branches were also found crossing to the wall of the cyst. The keratin debris and calcifications as well as part of the wall were resected and removed. The major vessels were freed and the resected specimen was sent for histological verification.

The patient had uneventful postoperative recovery.

The histopathological result suggested keratinizing and calcifying odontogenic cyst (Fig. 5).

Discussion

Calcifying odontogenic cysts are rare tumors. The tumor is found predominately in the mandible premolar region and was first reported by Gorlin et al in 1962 (Gorlin et al. 1235-43). The presentation of the patient in this report was mainly due to increased intracranial pressure manifested as headache and visual blurring. The behavior of the intracranial tumor is that of Gorlin cyst (calcifying odontogenic cyst) and shows features similar to craniopharyngioma (Hong, Ellis, and Hartman 56-64). The findings of the intra-oral examination showed involvement of the anterior jaw with impaired teeth, root resorption and divergence features suggestive of Pindorg's tumor. Predominance of female gender and

involvement of the anterior mandible region has been reported (Hirshberg, Kaplan, and Buchner 555-58). Association of COCs and odontogenic tumors has been reported (Nagao et al. 174-79). The association of COC and odontoma in this patient could be coincidental (Tomich and Shafer 755-59), or the COCS might have developed secondarily from the odontogenic tumor (Nagao et al. 174-79)or the reverse (Takeda, Suzuki, and Yamamoto 108-13). Furthermore the association of ascites in this case raises a number of postulations. The ascites might have developed secondary to dissemination of malignant cells from the odontogenic tumor. This seems unlikely since neither the histological report of the specimen nor the ascetic analysis was in favour of malignancy. The other possibility is that occurrence of ascites is a result of presumptive neurotransmitter that disrupts the mesenteric vasculature.

In conclusion the calcifying odontogenic cysts, though rare, are to be considered in the differential diagnosis of calcified suprasellar lesions

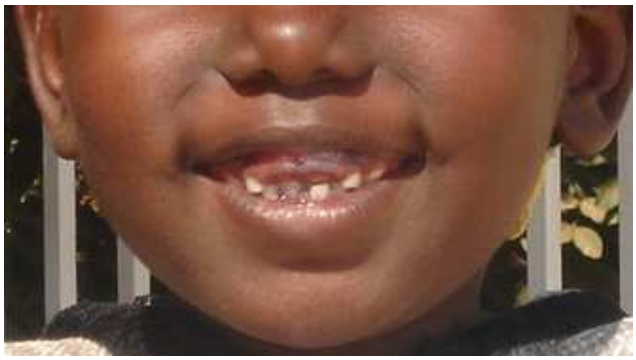


Fig.1
Gingival hypertrophy with tooth-like structures.

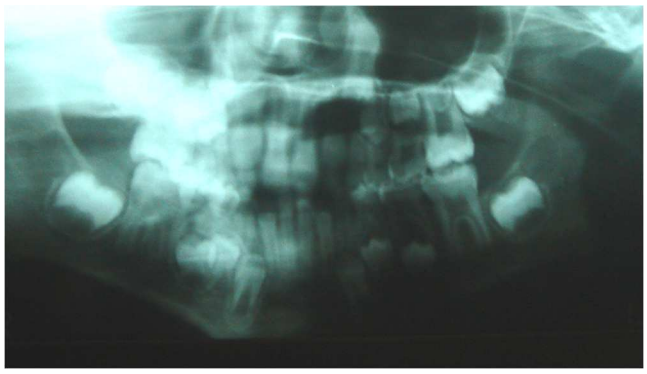


Fig 2
Plain radiograph illustrating the dense organization of the un-erupted and tooth like structures

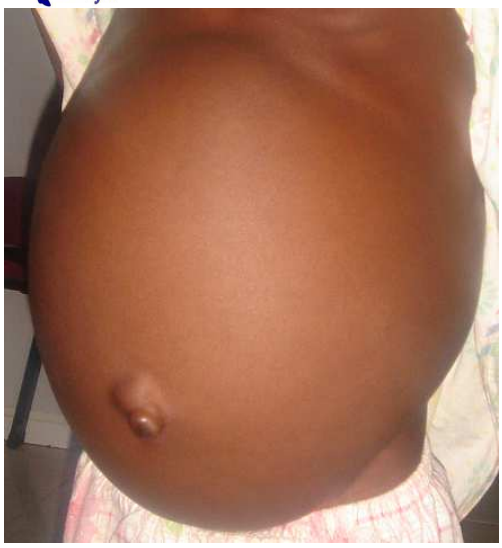


Fig.3
Abdominal distension with full flanks and everted umbilicus

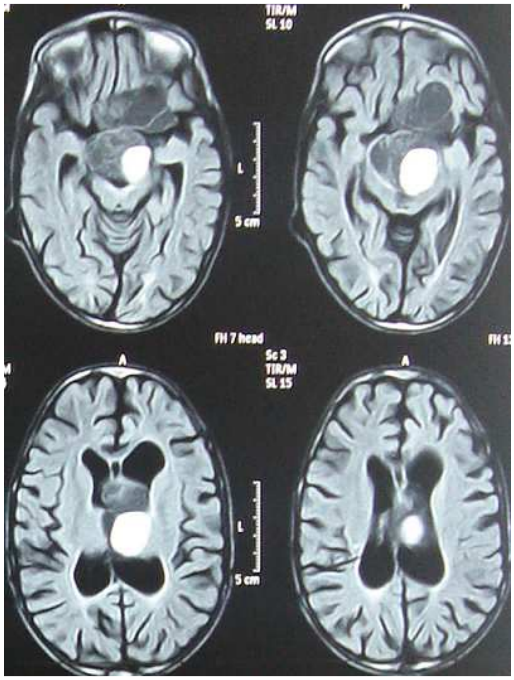
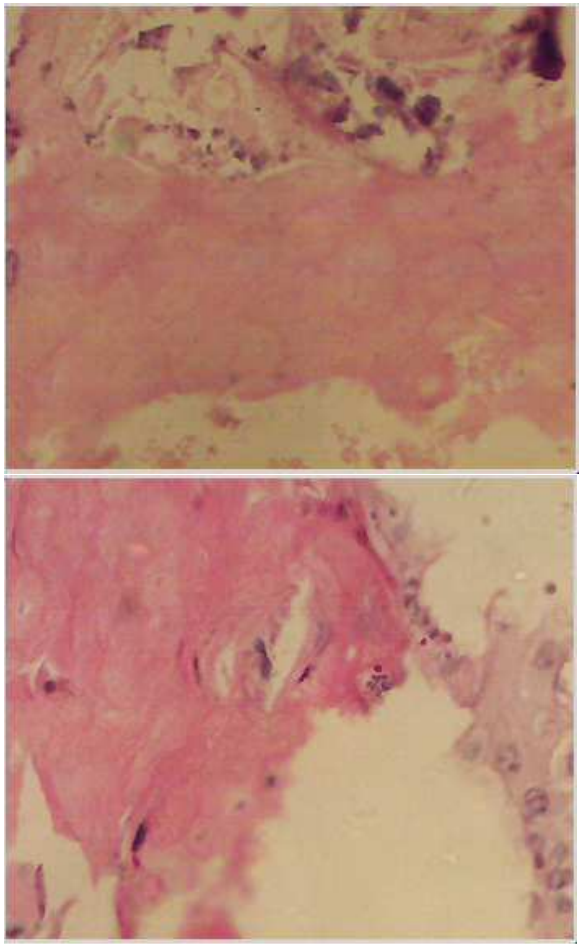


Fig. 4
MRI brain axial images (T1 W) showing multiloculated sellar and suprasellar lesions that extends posterior to the brainstem.



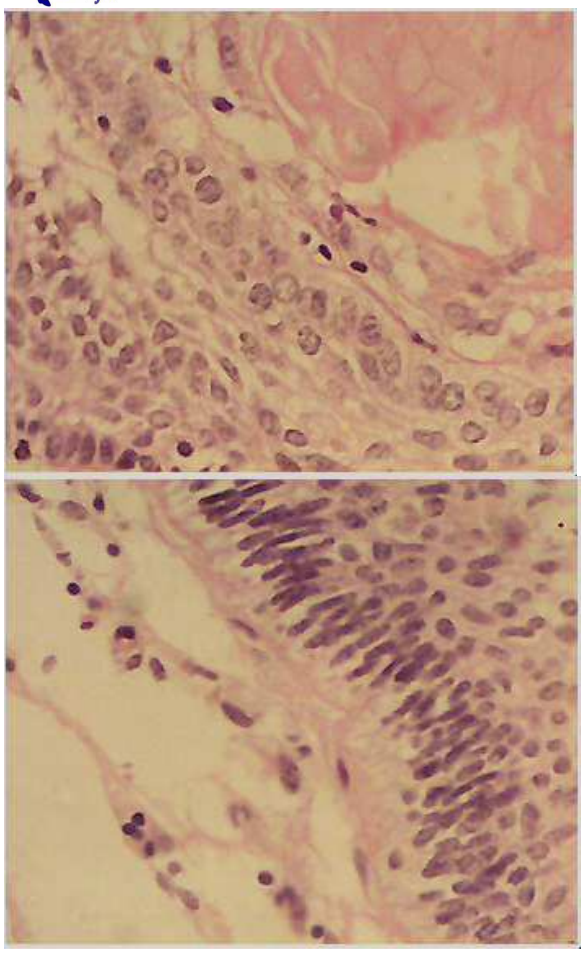


Fig. 5
Sections show a cyst lined by stratified squamous epithelium in which the basal cells are prominent. Some cells show clear cell changes. There are large masses of keratinizing ghost cells, keratin and calcification.

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