International Journal of Current Medical and Pharmaceutical Research

IJCMPR

ISSN: 2395-6429

Available Online at http://www.journalcmpr.com

DOI: http://dx.doi.org/10.24327/23956429.ijcmpr20170224

CASE STUDY

KERATOCYSTIC ODONTOGENIC TUMOUR: A PRESENTATION CROSSING BOUNDARIES

Sudip Indu* and Priyanka Chakravarty Indu

Air Force Institute of Dental Sciences Agram Post,
Old Airport Road, Bangalore-560007

ARTICLE INFO

Article History:

Received 20th June, 2017
Received in revised form 3rd July, 2017
Accepted 27th August, 2017
Published online 28th September, 2017

Key words:

bony pathology; coronoid process;KCOT; osteolytic lesion

ABSTRACT

The WHO in the year 2005 reformed the nomenclature of odontogenickeratocyst (OKC) to Keratocysticodontogenic tumour (KCOT) as it always has been recognised with alarming interest because of its strange nature, argued expansion phenomenon, distinctive propensity for neoplastic behaviour, recurrence and unclear treatment regimens. These lesions have been the topic of intense study over the last decade. Here, we present a case of a 30 year old man with an extensive osteolytic lesion in the body of the mandible extending right up to the coronoid process. This patient underwent multiple consultations previously, however remained symptomatic. The incisional biopsy was non-contributory. The histological examination of the excised specimen gave a diverse picture of a bony pathology but finally was conclusively established as a KCOT, by the presence of characteristic cystic lining in only one end of a section. The case has been deliberated with the clinical, radiological and histopathological parameters.

INTRODUCTION

Keratocysticodontogenic tumor (KCOT) has been a unique form of a benign neoplasm, resembling an odontogenic cyst but mimicking a sinister lesion because of its intramedullary bony expansion and high recurrence rate.[1] The WHO reformed the nomenclature of odontogenickeratocyst (OKC) to KCOT and defined it as a benign intraosseous neoplasm of odontogenic origin .[2] KCOT commonly occurs in the mandible and generally expands antero-posteriorly. Varied treatment regimens ranging from simple enucleation, curettage using Carnoy's solution, peripheral ostectomies and resection are practised. Long-term follow up with routine radiographs is mandatory for successful management.[3]

CASE REPORT

A 30-year old man reported with complains of pain and swelling on the left side of face and pus discharge from lower left posterior teeth for the past 8 months. Initially the swelling was small in size but gradually increased to the present size of 5x3cm extending from canthomeatal line to lower border of mandible and from premolar region up to the posterior border of ramus.The patient's medical and family history was non-contributory. Extra-oral examination showed mild swelling of face in the pre-auricular region. Skin over the swelling was normal. (Figure 1a) He remained symptomatic even after multiple consultations and reported to our centre for management. Our differential diagnosis ranged from an epidermal cyst to an osteolytic lesion or an odontogenic tumour probably ameloblastoma.

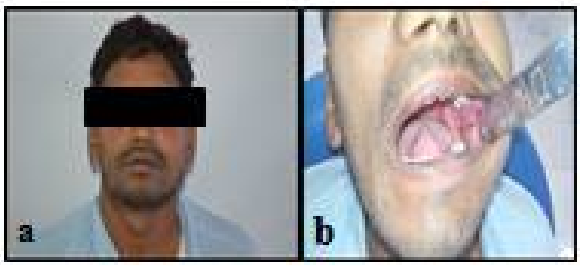

Figure1

Intraorally, a soft tissue defect was noticed in 37, 38 region with ingress of overlying mucosa into the defect. (Figure 1b) Orthopantomography showed well defined radiolucency extending from premolar region to the coronoid process. (Figure 2 a) The 3 D, CBCT image showed an aggressive osteolytic lesion in the mandible with multiple bony perforations in the ramus, coronoid process. (Figure 2 b) The sagittal CBCT image showed bony expansion of ramus of mandible and bone loss near sigmoid notch (Figure 2 c) and coronal view showed bone loss in relation to ramus of mandible. (Figure 2 d)

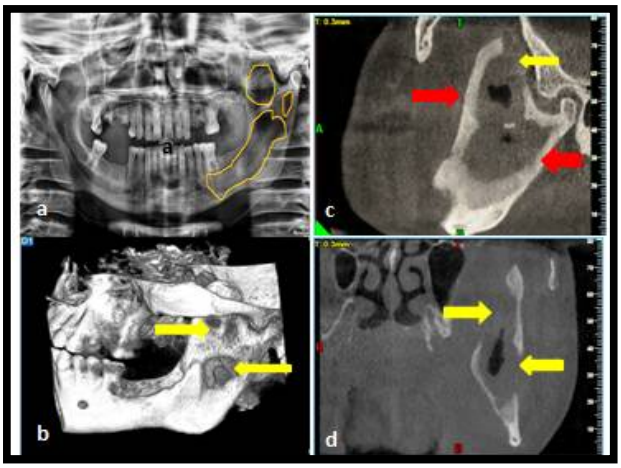

Figure 2

Because of extensive involvement of the body and ramus of mandible and wide expansion of the coronoid process, the surgical intervention was carried out with segmental resection and exarticulation under general anaesthesia. The H&E stained section exhibited a cystic lining epithelium of uniform thickness para keratinised in nature, devoid of any rete ridges. The lining was 6 to8 cell layers thick with basal cell palisading and hyperchromatism, giving a picket fence appearance. (Figure 3 a) The lumen of the cystic cavity showed keratinous flakes. Within the fibrous capsule, few odontogenic epithelial islands probably satellite cysts were noticed. (Figure 3 b) Also, seen were irregular trabeculae of woven bone independent of each other with osteocytes and inconspicuous osteoblastic rimming. (Figure 3 c) Few areas showed stromal cells in close proximity with newly forming trabecular bone. (Figure 3 d) The fibro capsular region was moderately collagenized & minimally vascular but showed absence of any atypical phenomenon. The microscopy was suggestive of a KCOT (Keratocystic Odontogenic Tumour). The patient has been responding well and is asymptomatic for 2 months post-operatively.

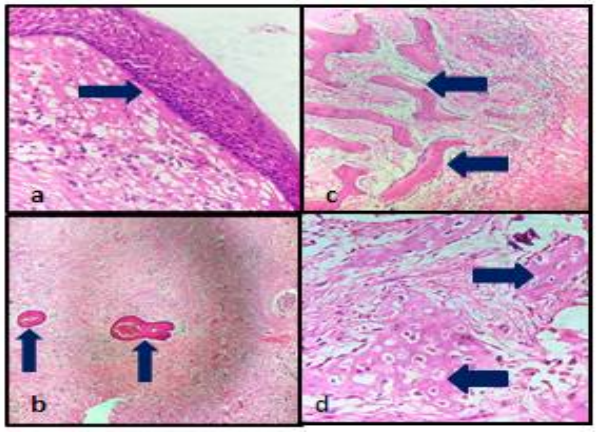

Figure 3

Legends

Figure 1 a- shows mild swelling on the left side of face
Figure 1 b - Soft tissue defect with ingress of overlying mucosa
Figure 2 a- Well defined radiolucency in the left vertical ramus region and body of mandible also involving the coronoid process (Yellow demarcated line)
Figure 2 b- Massive osteolyticlesion in the mandible with multiple bony perforations (Yellow arrows) in the ramus, coronoid process and the region below the neck of condyle.
Figure 2 c-Sagittal view of left mandible showing bony expansion of the ramus of mandible (Red arrows) and bone loss near sigmoid notch (Yellow arrow)
Figure 2 d- Coronal view of left mandible showing bone loss in relation to ramus of mandible (Yellow arrows)
Figure 3 a- Histo-pathological image showed uniformly thickened cystic lining epithelium **w**ith darkly stained basal cell layer. (Blue arrow) (H&E), (100X)
Figure 3 b-Histo-pathological image showed odontogenic epithelial islands probably satellite/daughter cysts in the fibrous cystic capsule/wall.(Blue arrows) (H&E), (100X)
Figure 3 c-Histo-pathological image showed irregular trabecular bonewithin the cystic capsule/wall.(Blue arrows)(H&E), (100X)
Figure 3 d-Histo-pathological image showed newly forming bone within the cystic capsule/wall.(Blue arrows) (H&E), (400X)

DISCUSSION

The KCOT formerly known as the OKC is regarded as one of the most aggressive cysts arising in the odontogenic region because of its long, deceptive &antero-posterior intramedullary bony expansions and high recurrence rate.[1] The WHO in the year 2005 reformed the nomenclature of OKC to KCOT because of these specific features, unique to this group of odontogenic cysts.[2]

An aberrant expression, loss of heterozygosity and inactivity of tumour suppressor gene PTCH is also noted both in syndromic and sporadic OKC's.[3] In the Sonic Hedgehog pathway, the PTCH tumour suppressor gene has an inhibitory control over Smoothened (SMO) Oncogene and the same is lost when PTCH binds with ligand Hedgehog(HH). Numerous studies have shown that activation and overexpression of SMO results in increased intercellular signalling through the GLI zinc transcription factor resulting in various tumours.[4] KCOT's are more common in the body and ramus of mandible but rare presentations in the maxilla has also been reported.[5] Our case represented the classical presentation of a KCOT, with extensive intra-medullary bony antero-posterior involvement and minimal bucco-lingual bony expansion. The marked bony expansion of the coronoid process gave a very unusual picture of a fibro-osseous lesion on radiographic findings. The CT findings brought out the aggressive phenomenon of the lesion where evidence of massive bony osteolytic defect with multiple bony perforations was seen in the ramus of the mandible mimicking a bony malignancy.

Microscopically, KCOT's are commonly occurring in two histopathologic varieties namely, parakeratotic and orthokeratotic types. The orthokeratoticodontogenic cyst type is considered as a discrete less aggressive unit.[6] The reported case was of parakeratotic type, with uniform thickness of cystic epithelium. Our case also showed presence of bone with in the capsular stroma mimicking a histomorphologic picture of an osseous metaplasia. Very few cases in literature have been reported where the capsule of this cystic neoplasm has shown mural bony and cartilaginous metaplasia. [7, 8] However, there has been minimal evidence in regard to the clinical destructive or benevolent nature and final prognosis of these lesions which showed metaplastic changes. Also, whether the calcification within the stroma is a product of known inductive

epithelio-mesenchymal interaction or a simple metaplastic change is something which has to be dwelt upon. Toller had proposed that biochemical protein estimation levels below 3.5 gm/100 ml in aspirated cystic fluids were mostly suggestive of a diagnosis of OKC. This finding was not significant in case of inflamed OKC where the epithelium lost its characteristic parakeratinized lining and in the process became permeable to varied soluble proteins thereby increasing the cystic protein levels somewhere in the range of 5.0-11.0 gm/100ml.[9] However, in our case attempted aspiration of cystic fluid was non- productive.

Diagnosis of KCOT's should therefore be done after careful interpretation of various clinical, radiological, biochemical, histopathological and molecular investigations, as the parakeratotic variant of KCOT are also known to be associated with Nevoid Basal Cell Carcinoma syndrome.[10] A long duration evaluation and review is mandatory, as a diagnosis of KCOT warrants strict follow up because of the aggressive nature of the lesion and its high recurrence rate. The day may not be very far off when newer molecular intervention techniques will be used as valuable adjunct in the management of such lesions which will safeguard patients from undergoing unwanted grave surgical operations.

References

1. Neville BW, Damm DD, Allen CM, Bouquot JE. Oral and maxillofacial pathology. 3rd ed. St. Louis: Saunders; 2009:683-687.
2. Reichart P, Philipsen P.Odontogenic Tumors and Allied Lesions. Quintessence Publishing Co. Ltd; 2004: 22.
3. Rajendran R, Sivapathasundharam R. Shafer's Textbook of Oral Pathology. 7th ed.Elsevier; 2012: 263-267.
4. Ruietal. Smoothened gene alterations inkeratocysticodontogenictumors. *Head & Face Medicine.*2014; 10:36.
5. Chaudhary S, Sinha A, Barua P and Mallikarjuna R.Keratocysticodontogenic tumour (KCOT) misdiagnosed as a dentigerous cyst. *BMJ Case Rep.* 2013;doi: 10.1136/bcr-2013-008741.
6. MacDonald-Jankowski DS. Keratocysticodontogenic tumour: systematic review. *DentomaxillofacRadiol.* 2011; 40:1-23.
7. Gotmare SS, Tamgadge A, Pereira T, Shetty A. Keratocysticodontogenictumor with ossification and calcification: A case report with unusual histological findings. *Indian J Dent Res* 2016;27:441-4
8. Fornatora ML, Reich RF, Chotkowski G, Freedman PD. Odontogenickeratocyst with mural cartilaginous metaplasia: A case report and a review of the literature. *Oral Surg Oral Med Oral Pathol OralRadiolEndod* 2001; 92:430-4.
9. Toller PA. Protein substances in the odontogenic cyst fluids. *British Dental Journal.*1970 ;128(7):317-22.
10. Tarakji B, Baroudi K, Hanouneh S,Azzeghaiby SN and Nassani M. Possible recurrence of keratocyst in nevoid basal cell carcinoma syndrome: A review of literature. *Eur J Dent.* 2013 Sep; 7(Suppl 1): S126-S134.

****