Case Report

Case Report and Review of Keratoameloblastoma – A Rare Type of Keratin Producing Odontogenic Neoplasm

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ABSTRACT

Ameloblastoma is a slow-growing, locally invasive odontogenic tumour having odontogenic epithelial origin. Some histological variants of ameloblastoma show keratinization such as - Acanthomatous ameloblastoma, Keratoameloblastoma and Papilliferous Keratoameloblastoma. Among these, Keratoameloblastoma is a very rare tumour. Here, we present a case of Keratoameloblastoma in a 45-year-old female patient. Clinically, there was an extra-oral bony hard, tender swelling involving symphyseal and right para-symphyseal region of the mandible. Intraorally, there was a tender swelling in the dentoalveolar region with buccal cortical plate expansion which extended from 32 to 45 tooth region. Focal area of ulceration was also seen intraorally. Based on the clinical, radiological and histopathological features, Keratoameloblastoma was diagnosed. So, each and every case of Ameloblastoma should be judiciously examined for any histopathological metaplastic changes and also to be treated accordingly to prevent recurrence.

Key words: *Keratoameloblastoma, Keratinized Odontogenic Rare Tumour, Paccinian corpuscle*

Ameloblastoma is a slow-growing but locally invasive odontogenic tumour involving the jaws. It is of odontogenic epithelial origin, which lacks differentiation towards hard tissue formation. Ameloblastoma usually manifests during fourth and fifth decades of life [1], having no sex predilection but has a good recurrence rate if not operated and treated adequately. Histologically, the common subtypes of ameloblastoma are the follicular and plexiform types [2]. Some histological variants of ameloblastoma show keratinization such as - Acanthomatous ameloblastoma, keratoameloblastoma and papilliferous keratoamelo-blastoma. Of these, Kerato and papilliferous keratoameloblastoma are very rare tumours, having distinct histomorphology [3]. The unique histopathological feature of keratoameloblastoma is the presence of large amounts of keratin within the neoplastic odontogenic epithelium [4]. The term "Keratoameloblastoma" was first coined by Pindborg [5] in the year 1970, where he described an odontogenic tumour which comprised of keratinizing cysts as well as islands with papilliferous appearance. Hence, he suggested the term papilliferous keratoameloblastoma. After thorough study and review of literature, it was found that only few cases of ameloblastoma revealed extensive keratinization in their parenchyma and those all cases have been included in the group named keratoameloblastoma. Here, we present a case of Keratoameloblastoma involving the mandible in a 45-year-old female patient.

CASE DESCRIPTION

A 45-year-old female patient reported to a tertiary health care centre in Kolkata, West Bengal with a

complaint of a slow growing, slightly painful swelling in the anterior and right-side region of lower jaw since last eight months. Medical history of the patient was nil of note. Patient had a habit of betel leaf and areca nut chewing since last few years. Extra-orally, there was a bony hard, tender swelling involving symphyseal and right para-symphyseal region of mandible with a discharging sinus [Figure 1]. Regional lymph nodes were palpable. Intraorally, there was a tender swelling in the dentoalveolar region with buccal cortical plate expansion extending from 32 to 45 region. Focal area of ulceration was also noted intraorally [Figure 2].

Orthopantomograph revealed the presence of a large unilocular radiolucent lesion extending from 32 to 45 [Figure 3]. On the basis of clinical and radiological findings, a provisional diagnosis of odontogenic tumour (Ameloblastoma) was made. Differential diagnoses were calcifying cystic odontogenic tumour or any kind of intraosseous neoplasm.

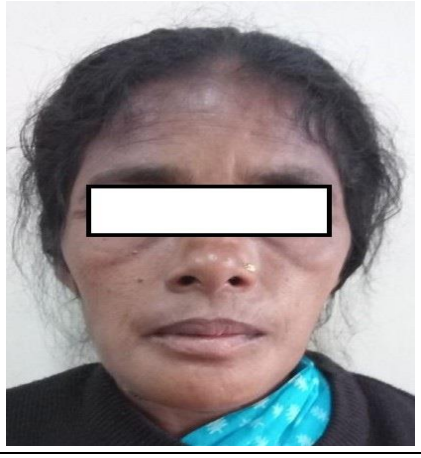

Figure 1: Extraoral view showing enlargement of symphyseal & right para-symphyseal region of mandible

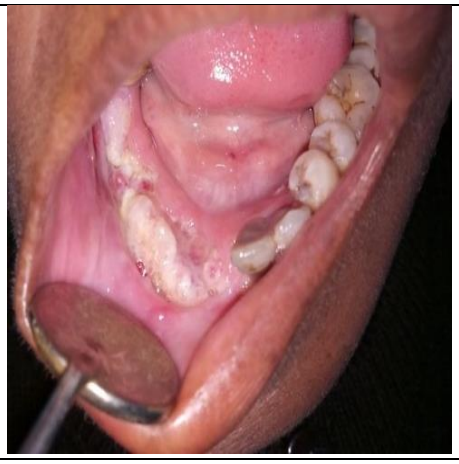

Figure 2: Intraoral view showing ulcerated area over the alveolus

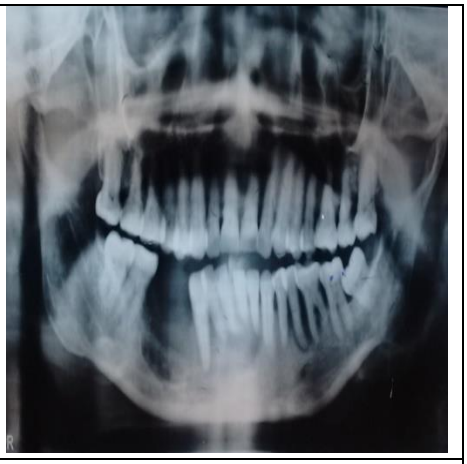

Figure 3: Orthopantomograph showing unilocular radiolucent lesion extending from 32 to 45 tooth region

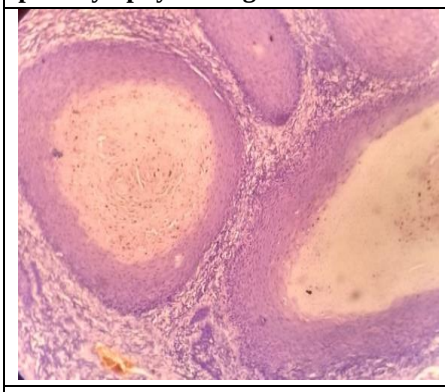

Figure 4: H&E-stained section (10X) showing ameloblastic follicles

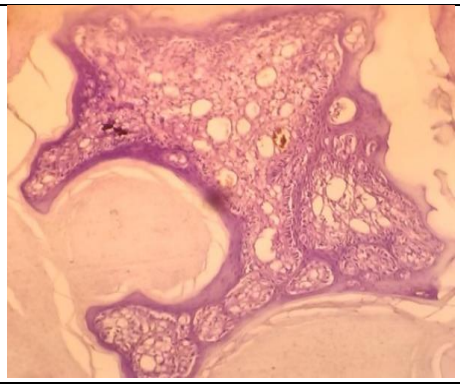

Figure 5: H&E-stained section (40X) showing ameloblastic follicles with peripheral ameloblast like cell and central stellate reticulum like cells

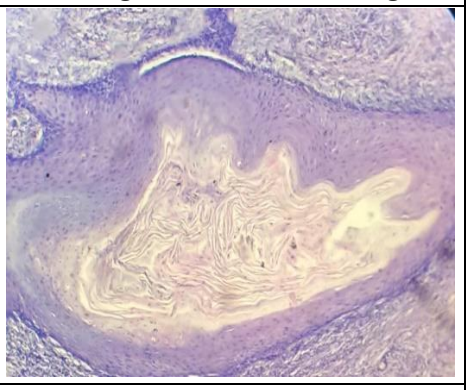

Figure 6: H&E-stained section (10X) showing ameloblastic follicles with central keratin formation

After routine pre-operative investigations and getting informed consent from the patient, an incisional biopsy from the lesion was done for histopathological confirmation. Tissue sections from the biopsied sample, when stained with Haematoxylin & Eosin and examined under microscope, revealed the presence of islands and strands of odontogenic epithelium [Figure 4] with central para and orthokeratin plugging. The periphery of the islands showed cuboidal to low columnar palisaded basal cell layer intermixed with multiple variable sized cysts. Some odontogenic follicles with peripheral layer of tall columnar ameloblast like cells with reverse polarity

and central stellate reticulum like in appearance were also noted [Figure 5]. Lamellated parakeratin is scattered throughout the tissue forming Paccinian corpuscle like structures [Figure 6]. The surrounding stroma was loose and oedematous with dispersed chronic inflammatory cells. The overall histopathological features were suggestive of Keratoameloblastoma.

DISCUSSION

According to World Health Organization (WHO), "Ameloblastoma is a benign intraosseous progressively growing epithelial odontogenic neoplasm characterised by expansion and a tendency for local recurrence if not adequately removed" [6]. Mutations in genes that belong to MAPK pathway results in the pathogenesis of ameloblastomas. The common mutations noted in ameloblastomas are *BRAF* (v-raf murine sarcoma viral oncogene homolog B1), *V600E* (activating missense mutation in codon 600 of exon 15), *KRAS* (gene Kirsten rat sarcoma viral oncogene homolog), *NRAS* (neuroblastoma RAS viral oncogene homolog), *HRAS* (Harvey Rat sarcoma virus) and *FGFR2* (Fibroblast growth factor receptor 2) [6].

Due to the multipotentiality of odontogenic epithelium, ameloblastomas often undergo certain types of metaplastic changes thereby giving rise to various histo-logic subtypes like granular cell, acanthomatous, basal cell, desmoplastic, papilliferous, clear-cell, keratoamelo-blastoma, haemangiomatous ameloblastoma [7, 8].

The term keratoameloblastoma refers to a group of ameloblastomatous lesions where the odontogenic ameloblastic epithelium produces a considerable amount of keratin. Whitt *et al*. [9] classified these keratoameloblastomatous lesions into four histological groups - Papilliferous; Simple histology; Simple histology with OKC like features and Complex histology. However, the common thing is the presence of excess keratin. Parakeratinization is mostly seen, but orthokeratinization may also be noted. The chief histopathological feature is the proliferation of odontogenic epithelium in a follicular pattern and the centre of such follicles often shows keratinization with or without cystic degeneration [10].

The common differential diagnoses were excluded based on the histopathological feature. Odontogenic keratocyst (OKC) was excluded because OKC exhibits a cystic lining with palisaded picket fence appearance of odontogenic epithelial cells and parakeratin lining. Acanthomatous variant was excluded as extensive keratin formation is unlikely in it. Treatment of Keratoamelo-blastoma is wide local excision of the lesion. A regular follow is needed to prevent or diagnose any recurrence.

CONCLUSION

Ameloblastoma may be considered as the second most common odontogenic neoplasm after the odontome. Ameloblastoma usually have numerous histologic variants. The main problem with histological diagnosis of is excessive keratinization which may mask the normal histological feature. So, a proper diagnosis, thorough treatment and a regular post-operative follow up is needed for proper management of Keratoameloblastoma.

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