Malignant Squamous Changes in Unicystic Ameloblastoma: A Case Report and Review of Literature

Ankita Tandon*

Faculty of Dentistry, Jamia Millia Islamia, New Delhi, India

*Corresponding author: Ankita Tandon, Faculty of Dentistry, Jamia Millia Islamia, New Delhi, India, Tel: 9891987040; E-mail: drankitatandon7@gmail.com

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Abstract

Unicystic ameloblastoma, a variant of solid or multicystic ameloblastoma is commonly reported to occur in third decade and encountered in the posterior mandible with a biologically low-grade course and with limited recurrence potential. Clinically, it frequently manifests as a painless swelling, which can be accompanied by facial edema, malocclusion and paresthesia of the affected area. Histologically, the minimum criterion for diagnosing a lesion as unicystic ameloblastoma is the demonstration of a single cystic sac lined by odontogenic (ameloblastomatous) epithelium often seen only in focal areas whereas radiographically they represent a unilocular radiolucency. This report however, is first to show malignant squamous conversion of unicystic ameloblastoma lining in a 35 year old female patient.

Keywords: Unicystic ameloblastoma; Carcinoma; Enucleation

Introduction

The concept of unicystic ameloblastoma was first introduced by Robinson and Martinez [1] in 1977 and later this was variously termed as mural, monostotic, intracystic, cystogenic, or cystic ameloblastoma [1]. It occurs at a mean age of 22 years and is located commonly in the mandible (mandible to maxilla = 3 to 13:1) with female: male preponderance of approximately 1:1.3 [2]. It is a unilocular, cystic lesion whose clinical features are similar to a non-neoplastic cyst [3]. The unicystic ameloblastoma deserves special consideration on the basis of its clinical & radiologic appearance, its histopathology, and its response to treatment. The importance of the unicystic ameloblastoma is that it possesses a much better prognosis after enucleation or curettage than does the classic intraosseous ameloblastoma [1].

Our case, on the contrary highlights a more aggressive counterpart of unicystic ameloblastoma in which the lesion shows dysplastic features (carcinomatous changes) to a great extent. This case represents first among the category of malignant transformation of unicystic ameloblastoma lining; which however, is reported on and often among classical ameloblastomas. Also, a brief review of malignant changes within ameloblastomas of maxillofacial region is presented herewith.

Case Report

A 35 year old female patient visited the Department of Oral Pathology, with the chief complaint of pain and swelling in the left lower back teeth region since 5 months. On examination, extraorally, a smooth and diffuse swelling was located in left lower cheek region causing facial asymmetry, measured approximately 4x5 cm in dimensions and extended from angle of mouth to mid body region antero-posteriorly. This was associated with dull pain and was tender on palpation. Intra orally, the swelling was present in 33 to 36 region, measured approximately 4x6 cm, was smooth surfaced and had an open socket in relation to 36 (Figure 1). The lesion also presented expansion of left buccal cortical plate and obliteration of the left buccal vestibule. An orthopantomographic evaluation revealed a well-defined unilocular radiolucency with scalloped borders extending from 31 to 36 region anteroposteriorly (Figure 2). Based on the above mentioned findings, a differential diagnosis of odontogenic keratocyst, radicular cyst and unicystic ameloblastoma were made. The aspirate was drawn and micro protein estimation value was found to be 9.8 gm% which ruled out the possibility of odontogenic keratocyst. Following this, enucleation was performed with application of Carnoy’s solution at the margins. The histopathology of the sections stained with H & E revealed the presence of stratified squamous epithelium overlying inflamed fibrocellular connective tissue stroma. The epithelial lining facing the lumen was variable in thickness from 4-5 layers to extensive proliferations of up to 15 layers at places (Figure 3). The basal layer appeared to be well defined columnar to cuboidal with polarization of nuclei along with heaping up of superficial cells and suspension of a few clear cells in the suprabasal and spinous cell layers (Figure 4). The epithelial lining at places significantly revealed dysplastic features with hyperchromatism, increased nucleo cytoplasmic ratio, increased mitotic figures, cellular and nuclear pleomorphism (Figure 5) along with massive epithelial proliferations both towards the lumen (luminal/intraluminal) as well as towards the connective tissue wall (mural) (Figure 6). The underlying connective tissue stroma had collagen fibre bundles with fibroblasts, chronic inflammatory cell infiltration chiefly of lymphocytes, endothelium lined blood vessels with few extravasated RBCs and irregular trabeculae of bone with osteocytes and osteoblastic rimming.

Correlation of histopathological, radiographic and clinical features supported a diagnosis of UnicysticAmeloblastoma (UA 1.2.3) with early carcinomatous change. The postoperative course was uneventful and at the one-year follow-up, there was no evidence of recurrence.
Discussion

Leider et al. [2] has proposed three pathogenic mechanisms for the evolution of unicystic ameloblastoma: (1) the reduced enamel epithelium associated with a developing tooth undergoes ameloblastic transformation with subsequent cystic development; (2) ameloblastomas may arise in a dentigerous or other type of dental cyst in which the neoplastic ameloblastic epithelium is preceded temporarily by a non-neoplastic stratified squamous epithelial lining; (3) a solid ameloblastoma undergoes cystic degeneration of ameloblastic islands with subsequent fusion of multiple microcysts to develop a unicystic lesion.
Unicystic ameloblastoma most commonly manifest facial disfigurement with noticeable swelling which coincides with the clinical presentation of the lesion in our case. The most common site for the occurrence of unicystic ameloblastoma in the mandible is the third molar area but mandibular body region is not an uncommon site for its occurrence as was also reported in our case. Unicystic ameloblastoma represents a cyst that is lined by Ameloblastomatous epithelium [2]. This epithelium may grow exophytically in the cyst lumen forming intraluminal nodules, or it may invade the fibrous cyst wall [1].

Yavagal et al. [4] revealed that although cystic ameloblastomas follow a biologically low-grade course, recent evidence suggests that they may often behave as biologically aggressive tumors. This is supported by the high incidence of cortical perforation, tooth resorption, lesion size, bone destruction, and a high rate of recurrence after simple enucleation. Epithelial-mesenchymal interactions are essential in pathogenesis of these type of lesions which is brought about through modification of the extracellular matrix/stroma. Fibroblasts, the cells that synthesize collagen undergo changes in response to tumorigenesis in adjacent epithelial cells. These changes in fibroblasts bring about alteration of collagen fibres, affecting their quality and organization [5]. This aggressive biological behaviour is also strongly supported by numerous immunohistochemical studies which confirm the presence of proliferation and aggressiveness markers in unicystic ameloblastoma (Table 1).

Review of malignant changes arising from ameloblastomas in existing English literature could reveal only a few cases which have been summarised in Table 2. To the best of our knowledge, the case presented here is first to show malignant squamous conversion of ameloblastomatous epithelium in unicystic ameloblastoma. Unicystic-type ameloblastoma are generally removed as a dentigerous cyst without preoperative biopsy, and all of the tissue must be included for a proper diagnosis [12]. Hence, the treatment performed was surgical enucleation with mechanical curettage following application of Carnoy’s solution on the margins to remove the residual tissue bits, if any. After 1 year of follow-up no recurrence was detected [18].

To conclude, it is important to keep in mind that, in general, unicystic ameloblastomas are solitary lesions with less aggressive behaviour in comparison to classical ameloblastomas. However, the clinicians should be aware of the possibility malignant squamous changes in unicystic ameloblastoma lining manifested as early carcinomatous change. These types should be treated carefully and a follow up phase is mandatory as they may be associated with recurrences.
Reference Markers Expression
Altini M et al. [6] Stromal myofibroblasts (MF) Marked expression
Li TJ [7] PCNA & Ki-67 Marked expression in invading islands
el-Sissy NA[8] p53 Marked expression
E-cadherin Weak positive expression
β catenin Marked expression
Piatelli A et al. [10] Calretinin Marked expression

Table 1: Expression of immunohistochemical markers in unicystic ameloblastoma

<table>
<thead>
<tr>
<th>S. no</th>
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<th>Location</th>
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</tr>
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<tr>
<td>1</td>
<td>VILLA VG [13]</td>
<td>Intraosseous ameloblastoma</td>
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<tr>
<td>2</td>
<td>Baden E et al. [14]</td>
<td>Peripheral maxillary ameloblastoma</td>
<td></td>
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<td>3</td>
<td>Ueta E et al. [15]</td>
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<td>mandibular</td>
</tr>
<tr>
<td>4</td>
<td>Nthumba PM [16]</td>
<td>Intraosseous ameloblastoma</td>
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<td>5</td>
<td>Ahuja A et al. [17]</td>
<td>Intraosseous ameloblastoma</td>
<td>mandibular</td>
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<tr>
<td>6</td>
<td>Present case</td>
<td>Intraosseous unicystic ameloblastoma</td>
<td>mandibular</td>
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Table 2: Review of malignant squamous changes in ameloblastoma

References