INTRODUCTION

The unicystic ameloblastoma (UA) represents an ameloblastoma variant, presenting as a cyst. There is no gender predilection. Five to 15% of all ameloblastomas are of the unicystic type. More than 90% of cases involve the mandible, usually the posterior region. It is more common in the second and third decades of life and is rare in children under 12 years of age, and better response to conservative treatment. It shares many clinical and radiographic features with odontogenic cysts/tumours and/or periapical disease of endodontic origin. Reported here is an unusual case of unicystic ameloblastoma involving the crown of an unerupted mandibular first premolar in a 9-year boy in an uncommon location, which was misdiagnosed as periapical lesion of inflammatory origin clinically, and as a dentigerous cyst radiographically. This highlights the importance to routinely submit the removed surgical specimen for histopathological examination.

CASE REPORT

A 9-year boy presented with pain in the right side of the lower jaw for 6 months. There was history of dental caries involving the primary second molar (Figure 1a). Past dental and medical history was unremarkable.

He was taking no medication and had no history of known drug allergy. His physical examination revealed no abnormality other than those related to the chief complaint.

On extra oral examination, facial asymmetry was apparent with a diffused swelling of 1x1 inch involving the right side of mandible, with the intraoral examination on palpation, showing a hard, non-tender mass on the right side of the mandible, covered by intact and immobile mucosa. Radiologic appearance of the lesion on the panoramic X-ray was unilocular rather well-defined radiolucent lesion localised distal to the lower second right molar, likely a dentigerous cyst (Figure 1b). Differential diagnosis included odontogenic tumours like a odontogenic keratocyst (OKC/KCOT), ameloblastic fibroma, a calcifying odontogenic cyst (COC) and an adenomatoid odontogenic tumour (AOT).

Enucleation of the lesion was performed to completely
extirpate the cystic lesion (Figure 1c) with extraction of the primary second molar. The enucleated tissue was a monocytic lesion on gross examination. On histopathological examination, the cystic lesion was mainly lined by a thin layer of non-keratinising stratified squamous epithelium with a basal layer of columnar cells, showing hyperchromatic nuclei/reverse polarity/cytoplasmic vacuolisation. The suprabasal epithelial cells were loosely cohesive and resembled a stellate reticulum like cells; (d) suggestive of unicystic ameloblastoma.

DISCUSSION

There are three forms of ameloblastomas: multicystic, peripheral, and unicystic tumours. A unicystic ameloblastoma was first described by Robinson and Martinez in 1977. Although it is a variant of ameloblastomas, it has a relatively benign biologic behaviour and better response to conservative treatment, which makes it a distinguishable entity. It accounts for 15% of all intraosseous ameloblastomas. Unicystic ameloblastomas are characterized as a slow growing and relatively local agressive cystic lesion. Radiographically, the lesions commonly show expansive unilocular radiolucencies with a well-demarcated border. Approximately 50 - 80% of cases are associated with an impacted or unerupted tooth. Therefore, the clinical and radiographic presentations of unicystic ameloblastoma are sometimes indistinguishable from those of dentigerous cysts.4,5

Various contradictory theories on the development of UA have been proposed by many authors. Some authors suggested that UA arises de novo. Leider et al. proposed three pathogenic mechanisms of evolution of UA.

Reduced enamel epithelium associated with a developing tooth undergoes ameloblastic transformation with subsequent cystic development. Ameloblastomas arise in dentigerous cyst or other types of odontogenic cysts, in which the neoplastic ameloblastic epithelium is preceded temporarily by non-neoplastic stratified squamous epithelial lining. Solid ameloblastoma undergoes cystic degeneration of ameloblastic islands with subsequent fusion of multiple micro-cysts and develops into a unicystic lesion.5,7

In 1988, Ackerman and Altini proposed schematic representation of histologic subtypes.7 According to this, group I consisted of a cystic lesion lined by simple odontogenic epithelium. Group II consisted of a cystic lesion showing intraluminal plexiform proliferation of the epithelial lining. Group III consisted of a cystic lesion with epithelial invasion of the supporting connective tissue in either a follicular or plexiform pattern. In 2004, histologic subgrouping was given by Philipsen and Reichart, as subgroup 1-luminal UA; subgroup 1.2-luminal and intraluminal; subgroup 1.2.3-luminal, intraluminal and intramural; and subgroup 1.3-luminal and intramural.8

Figure 1 (a,b,c,d): (a) Dental caries involving primary second molar; (b) OPG reveals unilocular rather well-defined radiolucent lesion below the cemento-enamel junction(CEJ) of the developing premolar; (c) enucleated tissue with the premolar tooth, on histological evaluation H & E stained section reveals epithelial cells that were loosely cohesive and resembled a stellate reticulum like cells; (d) suggestive of unicystic ameloblastoma.
Ameloblastoma is rare before the age of 10 years. According to statistical analysis of 1,036 cases of ameloblastoma collected from the literature found, only 2% of cases occur before 10 years of age. Fortunately, unicystic ameloblastomas have less-aggressive, and a better prognosis even after conservative surgical treatment. Studies have revealed that treatment of UA in children is complicated by following factors: The continued facial growth, different bone physiology (greater percentage of cancellous bone facilitating rapid spread, increased bone turnover, and reactive periosteum), presence of unerupted teeth.\textsuperscript{3,6,9}

The majority of odontogenic tumours occur intraosseously within the maxillofacial skeleton, while extra osseous odontogenic tumours occur nearly always in the tooth-bearing mucosa. The clinical features of most benign odontogenic tumours are non-specific; benign odontogenic tumours show slow expansive growth with no or slight pain, the tumours may be generated at any stage in the life of an individual. Knowledge of basic clinical features such as age, gender, and location can be extremely valuable in developing the differential diagnoses of odontogenic tumours. Most UAs are enucleated with the pre-operative clinical diagnosis of dentigerous cyst and it is only on pathologic examination that their true nature is determined. Certain radiological features, such as large lesion size, bone scalloping, relationship to an impacted tooth or the mandibular canal, tooth resorption, as well as ill-defined lesion borders, require further radiological work-up; and biopsy is indicated for definitive diagnosis. The luminal variant does not infiltrate the surrounding bone; and as a result, no further treatment is required for these lesions. Long-term follow-up is recommended.\textsuperscript{4,5,10}

REFERENCES