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Unicystic Ameloblastoma: Cyst or Tumour

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ABSTRACT

One of the most common and significant tumour of odontogenic origin affecting the jaws is ameloblastoma. Even though it is considered a benign tumour, in certain cases it can lead to significant morbidity and even lead to mortality. A distinct yet less frequent type of ameloblastoma which resembles a cyst more than a tumour is Unicystic ameloblastoma (UCA), which has characteristic histopathological, clinical and radiographic features. A review of the literature is presented to help better understand the various types of UCA with a view to proper management

Key words: Ameloblastoma, radiographic, histopathological.

INTRODUCTION

Ameloblastoma has been traditionally considered a benign tumour of odontogenic origin even though it is locally aggressive and can reach a large size with destruction of surrounding structures. Falkson in 1879 and Churchill in 1934 have been credited with first describing ameloblastoma in detail.[1]There are three clinical types of ameloblastoma, namely multicystic (solid conventional), Unicystic and peripheral.[2]In 1977 Robinson and Martinez defined Unicyclist ameloblastoma as a distinct entity.[3] A lesion which clinically and radiographically resembles a cystic lesion but with histopathologic examination of the cystic lining revealingameloblastomatous epithelium, is termed a unicystic ameloblastoma. The term unicysticisderived from its gross appearance of a single cystic cavity with histopathology showing a cystic lesion with a cyst lining and a cyst wall .

Pathogenesis

Ameloblstomas are thought arise fromneoplastic transformation of odontogenic epithelium, the source of which might be from enamel organ, odontogenic epithelial rests (Cell rests of malassez, Serre), reduced enamel epithelium, epithelial lining of odontogenic cysts (dentigerous) and from basal cells of oral mucosa.[4]It is debatable whether UCA arises de novo or from pre existing cyst. The pathogensis of unicystic ameloblastoma has been put forward by Leider et al as 1. Arising de novo from odontogenic epithelium, 2. From transformation of the cystic epithelial lining of odontogenic cysts and 3. From cystic degeneration of a solid ameloblastoma.[5]The mechanism of cyst formation in a solid ameloblastoma has been suggested as being due to epithelial dysadhesion or due to the production by the tumour of proteinases (metalloproteinases, serine proteinases); enzymes that are concerned with degeneration of the enamel organ after crown formation.

Clinical and radiographic features

Unicystic ameloblastomas usually present as nonpainful swelling of the jaw. Smaller lesions are usually discovered in routine radiographs as they are symptomless. Large lesions can lead to perforation of cortex with ulceration of surface mucosa. They tend to occur in an younger age group

predominantly in the second decade of life. A slight male predilection is seen with the most common site of occurrence being the posterior mandible and ascending ramus area. They may or may not be associated with an impacted tooth with the mandibular third molar being the most often associated tooth. In a series of 193 cases Philipsen et al found the incidence to be biphasic and dependent on the association of the lesion with impacted teeth. He grouped them as being dentigerous (associated with impacted teeth) and non-dentigerous(not associated with impacted teeth) and found that the dentigerous type occurred much earlier than the non-dentigerous type and even found a reversal in sex predilection between the two groups with the non-dentigerous type having a female sex predilection.[6]

Radiographically while the conventional soild ameloblastoma presents with a radiolucent multilocular soap bubble appearance the unicystic ameloblastoma can presents as either a unilocular or multilocular radilucency with or without scalloped margins. Unilocular pattern isthepredominant presentation in UCAs associated with impacted teeth.[7]Other radiographic patterns seen are pericoronal, periapical and inter- radicular radiolucencies.[7]

Histopathology

The Histopathological criteria for diagnosis of UCA is demonstration of ameloblastomatous changes in focal areas of the lining epithelium of a cystic lesion. The lesionusually consists of a dense, uniformly thickened, fibrous connective tissue capsule, that consists totally or partially of ameloblastic epithelium, surrounding a solitary large fluid-filled lumen. The epithelial lining of the lumen is uniform in thickness and has a slightly hyperchromatic layer of palisaded basal cells, most of which exhibit reversal of nuclear polarity. The remaining layers resemble stellate reticulum.[8]Hybrid lesions akin to hybrid amloblastomas have also been encountered with hitopathological patterns of plexiform. follicular, acanthomatous, desmoplastic and clear cell type.[9,10]Robinson and Martinez[1]was the first to try and classify UCA.Ackermann et al[11] classified UCA according to Histopathological pattern into three types.

In 1998 Philipsen and Reaichart suggested a slightly modified Histopathological typing based on the Ackermann clasiification.[6]

Treatment

The Histopathological typing of UCA is crucial in the treatment plan. In case of subtype 1 and 1.2, enucleation and curettage with application of Carnoy's solution or liquid nitrogen cryotherapy is usually sufficient as recurrence rates with these types have been observed to be low.[12]In case of 1.2.3 and 1.3 subtypes a more radical approach of resection is usually warranted. In every case a thorough post operative examination of the full specimen should be carried out with multiple sections from many areas to determine the exact subtype of UCA. Furthermore treatment decisions should also be based on the age of the patient as emphasis should bein preserving function along with aesthetic considerations in younger patients.[13,14] Follow up of patients for recurrence should be carried out for 15 to 20 years since recurrence frequently presents very late.Recurrence rates vary depending on not only the histopathological type but also on treatment modality as reported by various studies with the highest rates seen in lesions treated only with enucleationand the least with lesions treated with resection.[13]

Ackermann et al 1988 [11]

Group 1	Luminal	Tumour confined to the luminal surface lining of the cyst
Group 2	Intra luminal(Plexiform UCA – where the projections have a plexiform pattern)	Nodular proliferation into the lumen without infiltration of tumor cells into the connective tissue wall
Group 3	Mural	Invasive islands of ameloblastomatous epithelium in the connective tissue wall not involving the entire epithelium

Philipsen and Reichart 1998[6]

Subtype 1	Luminal UA
Subtype 1.2	Luminal and Intraluminal UA
Subtype 1.2.3	Luminal, Intraluminal, and Intramural UA
Subtype 1.3	Luminal and Intramural UA

DISCUSSION

Unicystic type of ameloblastoma accounts for only 5 to 10% of all intraosseous ameloblstomas.[5]Debate still exists about the pathogenesis of unicystic ameloblastomas. It has been proposedthat UCA can arise denovo from odontogenic epitheloium or from transformation of a pre-existing cyst or from cystic degeneration of a conventional solid ameloblastoma.[5]Most of unicystic ameloblastomas occur in the posterior mandible 90% and are associated with impacted teeth, most commonly the third molar.[16]The diagnosis is based on the gross picture of a cycstic lesion with histologic findings of ameloblastomatous changes of the lining epithelium. The radiographic picture can be of a unilocular or multilocular radiolucency. Unicystic ameloblastomas have been considered to be less aggressive than conventional ameloblastomas because of lower recurrence rate of about 10%.[17,18]In many cases of small lesions the diagnosis of UCA is after the fact in that the initial diagnosis and treatment is based on a provisional diagnosis of an odontogenic cyst and only post operative through examination reveals anUCA. In some cases the differentiation between an odontogenic cyst and UAC is difficult. PCNA and Ki-67 expression has been shown to be more pronounced in UCAs than in regular odontogenic cyst linings and can serve to differentiate the two.[19]Various treatment options have been proposed ranging from simple curettage and enucleation upto segmental resection and radiotherapy. Considering the lower recurrence rate of unicystic type, a more conservative approach is usually advocated.

CONCLUSION

Unicystic ameloblastoma is an interesting variant of ameloblastoma that has charachterestic clinical, radiographic and histopathologic features. The treatment plan for each case should be decided based on these findings as the clinical course depends on variables like histopathologic type, association with impacted tooth, ageand duration. There is a marked similarity to a dentigerous cyst in some cases of UCA.The importance of thorough examination of the post operative specimen by multiple sections from different representative areas is again emphasized for correct diagnosis as well as histological typing, which to a great degree determines the treatment plan and prognosis.

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