Squamous cell carcinoma arising in a dentigerous cyst lining: A case report

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Abstract: A 33 year old man presented with a cystic lesion of the maxilla which, on clinical and radiological grounds, was thought to be a dentigerous cyst associated with an un-erupted right maxillary third molar. The lesion was treated by enucleation and primary closure. Histopathological examination revealed a Squamous Cell Carcinoma (SCC) arising in continuity with a benign epithelial cystic lining. The patient declined further surgery but continues to attend follow-up clinics. No recurrence has been noted one year after surgery. SCC arising from an odontogenic cyst is a distinct but relatively uncommon lesion. Because of its uncharacteristic presentation this lesion is rarely diagnosed preoperatively. It is on histopathological examination that the diagnosis is often made. A valid diagnosis of a true carcinoma arising in a cyst requires demonstration of an area of microscopic transition from the benign epithelial cystic lining to the invasive SCC. This report describes a histopathologically proven case of SCC arising in a maxillary dentigerous cyst.

Keywords: Squamous cell carcinoma, odontogenic cysts, epithelial cyst lining, malignant transformation and histological evidence.

Introduction

Squamous Cell Carcinoma (SCC) arising from the epithelial lining of an odontogenic cyst is a rare but distinct pathologic entity. The exact incidence of carcinoma rising in odontogenic cysts is unknown but is reported to vary between 0.01% and 0.02% with less than 200 cases having been reported in the world literature.10 The pathogenesis of SCC arising from an odontogenic cyst is unknown. Longstanding chronic inflammations in the adjacent connective tissue and continuous intraosseous pressure have been suggested as possible causative factors.6

The clinical and radiographic findings of SCC arising in an odontogenic cyst are nonspecific and preoperative differentiation from a simple odontogenic cyst is difficult.8 It is on histopathological examination of the whole cyst lining that definitive diagnosis is often made. In making the diagnosis however, the possibility of the tumour arising from the adjacent tissues rather than from the epithelial cyst lining must be ruled out.4 Furthermore, a valid diagnosis of a true carcinoma arising in a cyst requires demonstration of an area of microscopic transition from the benign epithelial cystic lining to the invasive SCC. This report presents a histopathologically proven case of SCC arising in dentigerous cysts in a 33 year old man.

Case report

A 33 year old man was referred to the University of Nairobi Dental Hospital on 13th October 2005 by his general dental practitioner for evaluation of a painful right maxillary swelling of two year duration and a 6 month history of pus discharge. He had previously consulted several other dental practitioners for this problem and had been treated with antibiotics without remission of symptoms. In his past medical history the patient had a previous hospitalisation in 1980 to the Kenyatta National Hospital for treatment of what he described as a cyst of the mandible. All his teeth, except the molar, were removed as part of that treatment.

On examination, the patient was in good general health. He had a full complement of the upper teeth except the (right maxillary third molar) while the mandibular arch was partially dentate. There was a fluctuant swelling in the right maxilla extending from the upper first molar to the tuberosity region. The swelling

Fig. 1: A panoramic radiograph showing a well defined radiolucent lesion in the right maxilla enclosing the (white arrow). The inset depicts a close-up view of the lesion.
was slightly tender with pus exudation from the gingival crevices on gentle pressure. The overlying oral mucosa was otherwise normal in appearance and no lymph nodes were palpable. There was no associated history of epistaxis, nasal blockage or paraesthesia on the affected side. A panoramic radiograph showed a radiolucent lesion in the posterolateral wall of the right maxillary antrum that was well defined and encompassing the right maxillary third molar (Fig. 1). There was no evidence of erosion into the maxillary antrum or the orbital cavity. A clinical diagnosis of a dentigerous cyst was made and enucleation of the lesion accomplished under general anaesthesia in October 2005. The specimen was submitted for histopathological examination which revealed a well differentiated SCC (Fig. 2 and 3). The patient declined any further surgery but follow up was done for a period of 16 months which showed complete healing of the surgical site. (Fig. 4)

Discussion

SCC arising in an odontogenic cyst is a distinct but relatively uncommon lesion with the incidence estimated to vary between 0.01% and 0.02%. Although the exact number of documented cases is difficult to determine, Gardner in 1975, reviewed all cases from 1889 to 1967 and determined that there were 25 acceptable instances of malignant transformation within the epithelial lining of odontogenic cysts. In 1991, Muller and Waldron found 81 documented cases in the world literature. In a more recent review, Yasuoka et al. found 56 reported cases. Carcinoma may arise from different types of odontogenic cysts; including dentigerous cysts, radicular cysts, lateral periodontal cysts, odontogenic keratocyst and residual cysts. Carcinomas arising in dentigerous cysts are reported only occasionally. In the review of literature Yosuoka et al found that of the 56 cases of SCC arising from odontogenic cysts only 16 cases arose from dentigerous cysts and of which only 2 occurred in the maxilla.

SCC arising in an odontogenic cyst appears to be twice as common in men as in women and occurs at least twice as often in the mandible as the maxilla with a predilection for the posterior region of the mandible. Reported clinical symptoms include swelling, dull pain, draining sinuses, and cervical lymphadenopathy. The aetiology of carcinoma arising in an odontogenic cyst is unknown. Chronic inflammation, keratinization in the cyst wall, and trauma from tooth extraction have been considered as possible predisposing factors for malignant transformation. Gardner states that the possibility of malignant transformation of odontogenic cysts is small in the absence of inflammation. However, there is no way of ruling out secondary infection where there is communication with the oral cavity. Some authors have considered the presence of keratinization in the cyst wall to be a risk factor for malignant transformation. Browne et al. have indicated that odontogenic cysts with keratinization may have a greater tendency to develop carcinoma than non keratinizing cysts.

In making the diagnosis of carcinoma arising from an odontogenic cyst it is important to eliminate several other possibilities such as a tumour arising from the adjacent tissue or metastasizing from distant tissues with subsequent invasion of the cyst; and cystic degeneration of a primary or metastatic carcinoma. The definitive diagnosis of malignant change is made by the recognition of epithelial dysplasia in a cyst lining. Hamp and Harrigan, in 1973 stated that the only valid index of malignancy arising within an odontogenic cyst is the presence of a transition zone between the normal and malignant epithelium. While this has been accepted in principle it has been pointed out that transition is only possible in the early stages of malignant transformation and identifying the precise areas of origin can be difficult. In our case we were able to demonstrate progressive epithelial changes in the cystic wall from benign epithelial lining (Fig. 2), to a chronically inflamed fibrous connective tissue cystic wall supporting a lining of variably hyperplastic stratified squamous epithelium with severe epithelial dysplasia (Fig. 3), malignant epithelial islands exhibited nests of keratin and abnormal mitoses (Fig. 3b).

Malignancy in the cyst wall is often unexpected at initial operation and treatment is usually by enucleation. Resection or sometimes radiation, are then performed as additional treatment following the
histopathological diagnosis. The decision to carry out further treatment is often based on the perceived relative risk associated with SCC and the occasional evidence of bone erosion. This case was significant in that simple enucleation of the cyst appears to have achieved complete cure contrary to what had been planned for the patient. The case challenges the premise upon which decision for radical operations are often made and calls for a review of the clinicopathologic criteria which could discriminate between cases that are manageable conservatively from those requiring a more radical approach. This case also illustrates the importance of adequate and complete histological examination of all cystic lesions, especially those excised in older patients.

Acknowledgements

We express our gratitude to the Dean, School of Dental Sciences University of Nairobi and the Chairman of the department of Oral and Maxillofacial Surgery for permission to publish this work. Our sincere thanks also go to Prof. Chindia for editorial review as well as Alice Limo and Josiah Gichana for technical assistance.

Bibliography