EXTRA-ABDOMINAL FIBROMATOSIS INVADING THE MANDIBLE: CASE REPORT

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SUMMARY

Extra-abdominal fibromatosis (desmoid tumour) is a rare aggressive neoplasm with a tendency to infiltrate local structures but rarely metastasises or undergoes spontaneous malignant transformation. The treatment of choice is surgery, however, recurrences have been reported even after wide-field resection. This article presents a case of extra-abdominal fibromatosis that had extensively invaded the mandible.

INTRODUCTION

Extra-abdominal fibromatosis, also known as desmoid tumour, is a rare aggressive tumour arising from fascial sheaths and musculoaponeurotic tissues which may affect any part of the body. Its aetiology is unknown but genetic, hormonal and traumatic factors have been implicated (1-4). Superficial fibromatoses grow slowly whereas musculoaponeurotic fibromatoses are locally aggressive and grow rapidly (5). Metastasis and spontaneous malignant transformation to fibrosarcoma are rare and only a few cases have been reported (6).

The parts of the body that are commonly affected include the shoulder girdle, inguinal region and lower extremities (5). Reports of this lesion in the maxillofacial region are rare. We present a case of extra-abdominal fibromatosis that had extensively invaded the left body of the mandible.

CASE REPORT

A 38 year old woman presented in the University of Nairobi Dental Hospital oral diagnosis clinic with a left mandibular swelling which had been enlarging over one year. The patient had first noticed loosening of the left mandibular molars which was followed by the appearance of a painless swelling. The lesion had progressively increased in size in spite of unspecified treatment. The patient’s general medical history had been essentially unremarkable.

On intraoral examination a large fungating lesion measuring approximately 8 x 6 cm was noted in the molar area of the left mandible and extending into the ramus. The pinkish mucosa over the tumour was intact and under it were many small blood vessels. The lesion was generally firm in consistency, but with some soft areas and was attached to deeper structures. The associated teeth (36,37) were mobile. An orthopantomogram revealed an osteolytic lesion involving the ramus and angle of the mandible with 38 displaced posteriorly (Figure 1). Under local anaesthesia, an incisional biopsy was performed and submitted for histopathology, the analysis of which indicated a subepithelial myxoma. Subsequently block resection of the tumour was accomplished under general anaesthesia with the anterior margin at the 35; and posteriorly the ramus was removed sparing the condyle. A stainless steel plate was used to reconstruct the defect.

Figure 1
Orthopantomogram showing extensive invasion of the left mandible by the tumour

Histopathological examination of the resected tumour revealed a lesion consisting of spindle cells with few mitotic figures arranged in fascicles, with a vascular component but the vessels appeared non-neoplastic. Microscopic examination of the lymph node (submandibular) showed reactive and hyperplastic changes but no metastatic disease (Figures 2 and 3). A diagnosis of a fibromyxomatous tumour of low malignant potential was made. Immunohistochemical analysis performed to enhance tumour specificity revealed the tumour to be an extra-abdominal fibromatosis (desmoid tumour). Hormonal receptor studies demonstrated the presence of oestrogen and progesterone receptors in the tumour.
DISCUSSION

Extra-abdominal fibromatosis involving the mandible may present a diagnostic challenge. As a slow growing, non painful, radiolucent swelling of the jaw, the clinical and radiographic presentation may mimic ameloblastoma, myxoma, or fibro-osseous lesions. Histopathologically, the tumour has features of a myxoma. Immuno-histochemical analysis may be necessary to give a definitive diagnosis.

The presence of oestrogen and progesterone receptors in the present case raises the possibility of involvement of these two hormones in the causation of the tumour. This case falls under the category of musculosaponeurotic fibromatoses and its aggressive nature is demonstrated by bone invasion and extensive destruction. Wide surgical resection is the treatment of choice. Radiation is recommended for lesions for which wide-field resection is not feasible. Other forms of therapy that have been tried include hormonal therapy with testosterone. A case has been reported of a woman followed-up for 22 years in whom testolactone showed tumour regression during the course of observation [7].

Where the tumour is inadequately excised, local recurrence may be as high as 50-75% (5). In the present case the tumour recurred inspite of wide field resection. One study has suggested that a subgroup of desmoid tumours at risk of recurrence may be hypervascular lesions associated with trisomy 8 (2). Hypervascularity was a prominent feature in the present case. Our case remains under close follow-up for any subsequent recurrences which may necessitate surgical intervention.

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REFERENCES