Case Report

Infantile fibromatosis of the mandible- Is radical treatment justified?

ABSTRACT

Infantile fibromatosis is characterized by proliferation of fibrous soft tissue with a potential of invading the adjacent structures but lacks the ability to metastasize, thus making it a fairly benign lesion with borderline characteristics. The pathology resembles sarcomatous growth, therefore making it difficult for the clinician to correctly diagnose. There are two variants of extra-abdominal desmoid juvenile and adult variant depending upon the age group it predominately involves. Fibromatosis is comparatively a rare tumour with unpredictable growth and varying local recurrence rates. The mass usually grows slowly, rapid growth and recurrences being mostly associated with the juvenile forms. The disease may present as single or multifocal lesion with widespread distribution, thus requiring whole body scans to identify any insidious growth elsewhere. Here, we report a case of recurrent juvenile/infantile fibromatosis in a 2-year-old child, conservatively managed without any growth disturbance and signs of recurrence.

Keywords: Aggressive fibromatosis, extra-abdominal desmoid, infantile fibromatosis

INTRODUCTION

Infantile fibromatosis (IF), also called desmoid tumour, juvenile fibromatosis, is a rare, benign but locally aggressive neoplasm of the soft tissues. The incidence of IF is estimated to be about 3-4 cases per 1 million people per year with a female/male ratio of 3:1.[1] These tumours develop in the deep soft tissue as fibroblast-derived clonal expansions. Considered to have intermediate biologic behaviour, without ability to metastasize, they are characterised by infiltrative growth patterns and a tendency toward local recurrence.[2] IF often poses a diagnostic challenge because of their rarity, atypical and easily misinterpreted, histologic features. Fibromatoses of childhood often show signs of increased cellularity, rapid growth, and infiltrative margins and these features may be attributed to a malignant process. Differentiation of IF from malignant soft-tissue tumour is important for an appropriate management protocol. The treatment of desmoids is controversially discussed. Most authors suggest radical resection.[3-6] Genetic disorders such as the Gardner syndrome may also lead to desmoids and patients with desmoid should be evaluated for it. Here, we report a rare case of recurrent growth mimicking malignant lesion in a 2-year-old child requiring meticulous management and careful follow-up.

CASE REPORT

A 2-year-old male child reported with a painless firm swelling involving the left dentate region of mandible for about a year. The swelling developed rapidly over the last

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3 months and recurred within a short span after excisional biopsy. The lesion was asymptomatic without any pain and paraesthesia of the adjacent structures. On physical examination, the lesion led to mild facial asymmetry involving the left lower third without any distinct margins and normal overlying skin. The lower border of mandible was regular and continuous without any detectable bruit on palpation. Mouth opening was adequate with no obvious deviation. Intraorally, a firm growth was seen involving the body of mandible from distal of the first deciduous molar on the left side into the retromolar region with evident buccal and lingual expansion. The lesion was sessile and well encapsulated. Mucosal ulceration was present due to occlusal trauma from overlying tooth and thinning. There was obliteration of buccal vestibule corresponding to the extraoral swelling with gross displacement of tongue to the opposite side to accommodate the growing lesion. The lesion had well-defined margins and no definite extension to the floor of mouth. The lesion had no active fluid discharge and was found to be noncompressible [Figure 1]. Owing to the history of aggressive growth rate of the lesion, the diagnosis of malignant lesion more likely of infantile fibrosarcoma or high-grade sarcomatous lesion of soft-tissue origin could not be ruled out clinically.

Initial histopathological examination of the excised tissue revealed benign soft-tissue tumor composed of spindle-shaped fibroblasts with no evidence of atypia diagnosed as benign fibroma with profuse vascularity.

A computed tomography scan was performed after the lesion recurred following excisional biopsy which revealed a mass lesion of size 2.8 × 2.1 × 2.1 showing homogenous density with peripheral contrast enhancement and central necrosis. There were mild osteolytic changes involving the body of mandible with possible cortical invasion of the lesion upto the angle of mandible. Loss of fat plane with left mylohyoid and effacement of left geniohyoid was seen, therefore depicting the probable infiltrative nature of lesion but with a focal presentation giving an impression of possible malignant conversion [Figure 2].

Considering the rapid growth rate and scalloped bony margins, an incisional biopsy and positron emission tomography (PET) scan was justified. Incisional biopsy revealed spindle cell tumour arranged in interlacing fascicles with collagenous stroma. No significant mitotic activity or cellular atypia was seen. Immunohistochemistry revealed beta catenin positivity, scattered desmin positivity, and CD31 positivity highly suggestive of an extra-abdominal IF of mandible.

PET scan rafter injection of 18 FDG revealed FDG avid soft-tissue density measuring approximately 2.9 (AP) ×3.5 (TR) cm involving the left lower alveolar region and left retromolar trigone. The lesion is extending inferiorly eroding the margins of alveolar margins of mandibular ramus to the floor of mouth and extension in submandibular region. Mild FDG avid subcentimetric left level IB, bilateral II, left level III nodes noted. There was no active uptake elsewhere thus ruling out any possibility of intestinal involvement or metastatic nature [Figure 3].

A definitive diagnosis of IF was established and subsequently treated by complete surgical excision of the tumour mass. The lesion was explored and removed after ensuring adequate local hemostatic measures. Curettage and peripheral ostectomy of the involved site was performed thoroughly after ensuring preservation of the underlying second molar

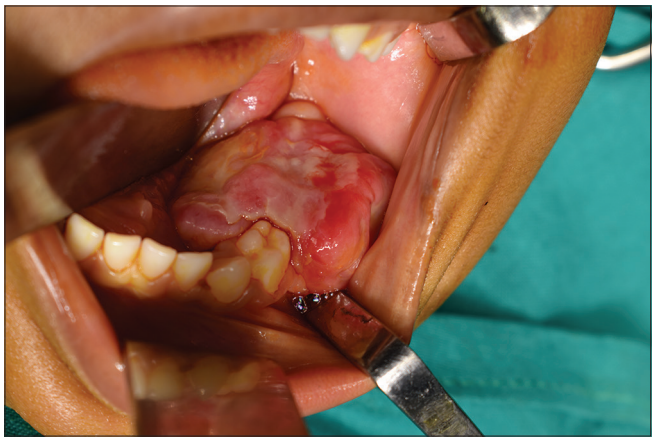

Figure 1: Preoperative image showing sessile proliferative growth arising from left body of mandible. The overlying epithelium is ulcerated due to trauma from opposing teeth

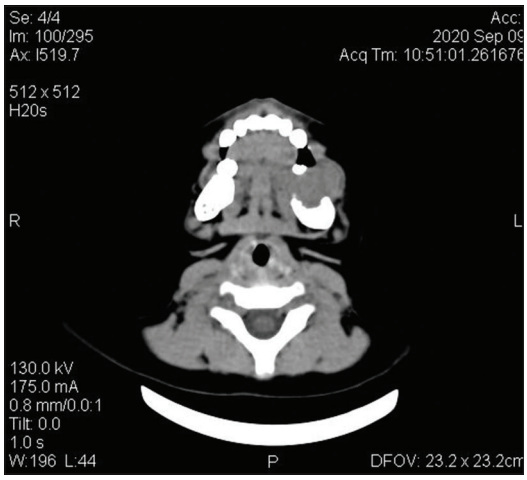

Figure 2: NCCT face axial view in soft-tissue window showing a sessile exophytic soft-tissue growth involving left body of mandible loss of fat plane with left mylohyoid and effacement of left geniohyoid

tooth bud and adjacent neural structures. The defect was closed primarily after adequate undermining without requiring any bony reconstruction [Figure 4]. Histology of the lesion also again confirmed the diagnosis of IF requiring regular follow-ups to monitor and report any chances of recurrence [Figure 5].

Patient was followed up over a period of 1 year without any signs of recurrence [Figure 6].

DISCUSSION

IF, also known as desmoid tumour, is a rare benign tumour that develops from the tissue of the musculoaponeurotic system and is characterized by local aggressive behaviour. The lesion is a rare entity and occurs in 10%-20% in head and neck region. Surgery has been considered the first-line treatment.[7] The NCCN recommends that surgery be the initial treatment modality for lesions that are surgically resectable.[8] Adjuvant radio or chemotherapy is then recommended based on the status of the surgical resection margin. In the largest single series, Meazza analysed the outcomes of 94 paediatric desmoids.[9] They report significantly lower local recurrence rates for resections with microscopic clearance (22%) compared to resection with both macroscopic and microscopic clearance and resection with macroscopic residual disease (both 47%). Buitendijk performed a meta-analysis of 12 pediatric case series performed between 1986 and 2004.[7] Recurrences were seen in 49% of patients, with 89% recurring within 3 years (97% at 6 years). The current consensus is that while there is benefit in gross total resection, microscopically negative margins do not improve local control, so the pursuit of negative margins does not justify increased postoperative functional and esthetic deficits.[1,10] In a series of eight pediatric patients treated for head and neck IF with surgery alone, only one patient had a recurrence, although seven of them had microscopically positive margins (median follow-up 3.5 years).[11] In our case, patient reported with recurrent lesion as excision of the lesion was done in some other centre without margin status. Considering the risk benefit ratio in

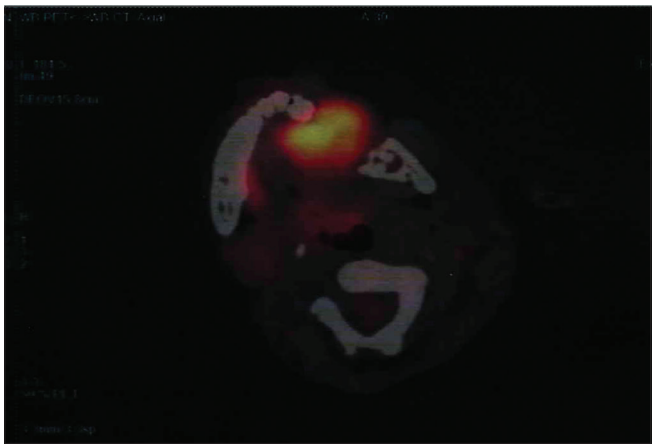

Figure 3: PET scan with 18 FDG uptake involving left body of mandible showing increased proliferative mass with bony erosion (SUV max = 8.0)

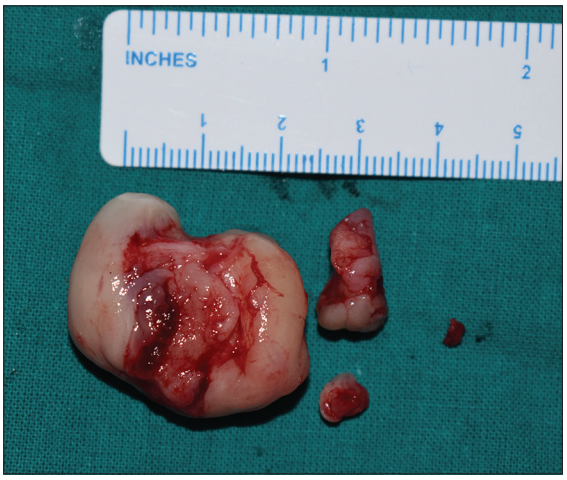

Figure 4: Tumour specimen measuring over 3 cm

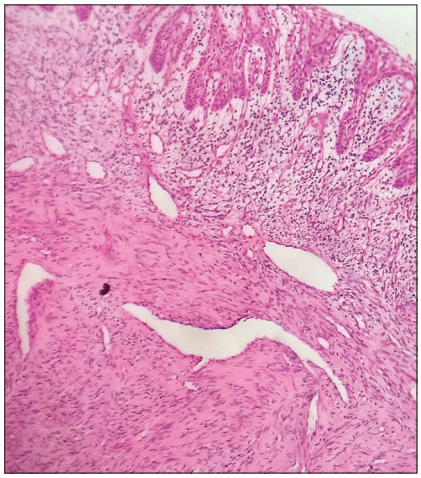

Figure 5: Photomicrograph at 10x showing overlying keratinized squamous epithelium with underlying fibrocellular connective tissue stroma

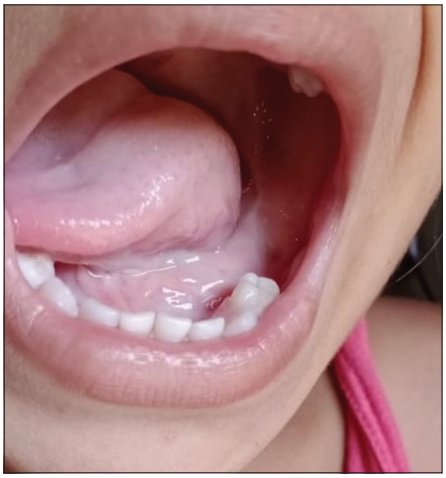

Figure 6: Follow-up at 1 year showing satisfactory wound healing and no signs of recurrence

this age group regarding residual deformity, complete surgical excision was done. No evidence of any recurrence noted after 1 year follow-up. We screened the patient for Gardener syndrome but could not find any association. A critical point is whether or not radical resection with clear histological margins is necessary, it would be logical to expect less relapse following more radical resection. But considering the residual deformity after radical resection in such a young age group, a conservative surgical approach may be advocated.

CONCLUSION

IF is a rare benign lesion, sometimes associated with Gardner's syndrome with a high recurrence rate. Due to young age group it predominately affects, it becomes seemingly important not to endanger patient to considerable postsurgical functional deformity and thereby hindering the psychosocial development. Owing to recurrence rate, it is especially important to follow-up patient over a long duration. If the tumour recurs, resection might be considered as a viable treatment option on a later stage of life.

Informed consent

An informed assent form was obtained from the patient's relatives.

Ethical approval

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the legal guardian has given his consent for images and other clinical information to be reported in the journal. The guardian understands that names and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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