A Rare Presentation of Congenital Benign Fibrous Histiocytoma Mimicking Mandibular Tumor in an Infant

Dear Editor,

Benign fibrous histiocytoma (BFH), also known as dermatofibroma, is a common skin lesion in the adult population. It most commonly presents as a slow-growing cutaneous nodule in the sun-exposed skin and rarely as a non-cutaneous lesion in the head and neck.[1] Age groups of 20 to 60 years are most typically involved.[2] Congenital cases are rare, with only a few prior cases reported.[3,4] We report one such rare case of congenital BFH that gave the impression of a mandibular origin due to the involvement of the periosteum of the underlying mandible.

A 7-month-old girl presented to the outpatient department with a solitary spherical swelling in the left lower jaw. The swelling was noticed at birth and was progressive in size. On examination, the diameter of the swelling was 2 cm; it was non-tender with well-defined margins. Consistency was firm with fixity to the skin and the underlying mandible [Figure 1]. Fine-needle aspiration cytology (FNAC) was suggestive of a spindle cell lesion.

Magnetic resonance imaging (MRI) of the neck showed a mild heterogeneous well-defined lesion in the subcutaneous plane of the left submandibular space causing skin tethering and abutting the mandible.

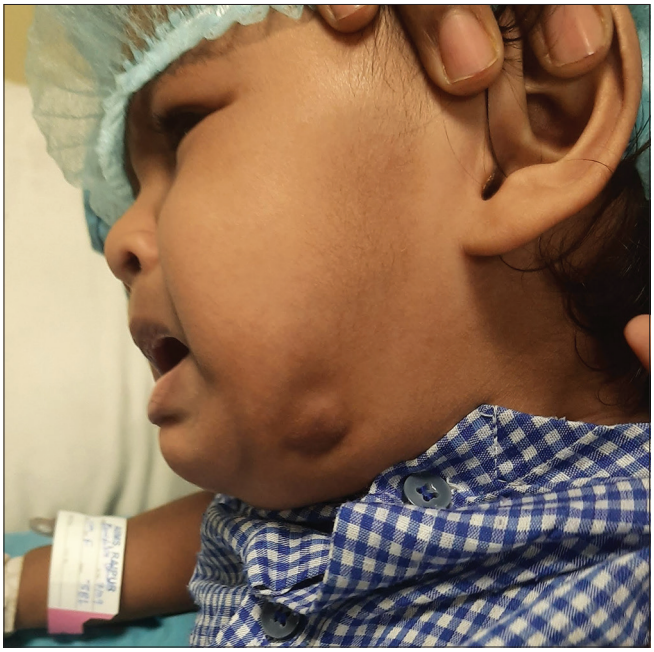

Figure 1: Pre-operative image of the lesion over the left mandible showing a 2 cm × 2 cm swelling with skin tethering. Discharge, opening, or puncta were not seen. The swelling was non-tender and fixed to the underlying bone on palpation

The patient was taken up for surgery, and the mass was excised under loupe magnification, keeping an adequate margin. The specimen was sent for histopathology [Figure 2].

Histopathology showed tissue lined by the stratified squamous epithelium. The dermis showed an ill-circumscribed lesion composed of spindle cells arranged in a fascicular and storiform pattern, collagenous stroma, and admixed with varying numbers of histiocytes, foam cells, and inflammatory cells suggestive of cutaneous BFH with periosteal margins free of the tumor [Figures 3 and 4]. The patient was followed up to 6 months postoperatively with no clinical signs of recurrence or neurological deficit [Figure 5].

The term fibrohistiocytic tumors is a descriptive designation for a group of heterogeneous lesions that share morphological features of histiocytes and fibroblasts on light microscopy.[2] Various histologic types have been described, including common BFH, epithelioid, aneurysmal, cellular, atypical, angiomatoid, and plexiform. The cutaneous BFH is further classified based on the histological types, including lipidized, hemosideric, keloidal, lichenoid, granular cell,

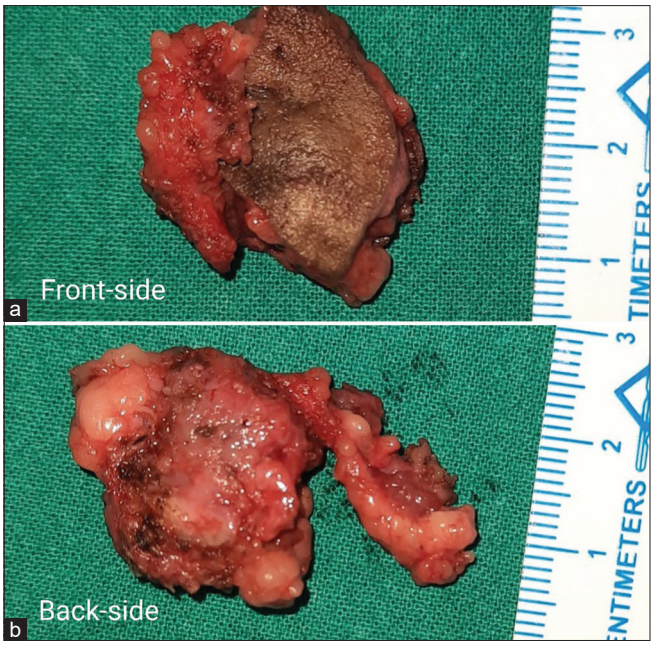

Figure 2: Excised specimen of the lesion viewed from – (a) The front side showing excised elliptical skin island with visible dimpling. The lesion was 2.5 cm × 2 cm, firm in consistency. (b) Back side showing a whitish patch of the periosteum that was fixed to the lesion and hence excised along with the lesion. Surrounding normal fat tissue can be seen which was taken out along with the lesion

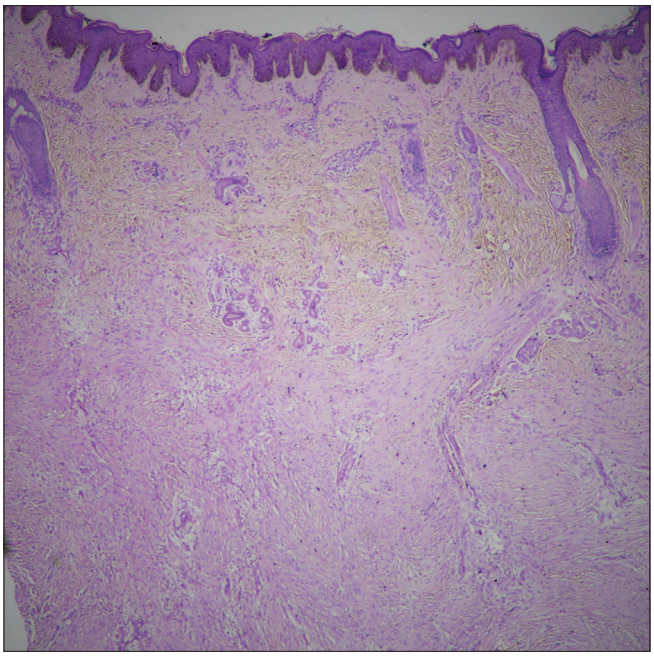

Figure 3: Low-power micrograph (H&E stain, 2×) showing dermal-based lesion with extension into the deeper dermis

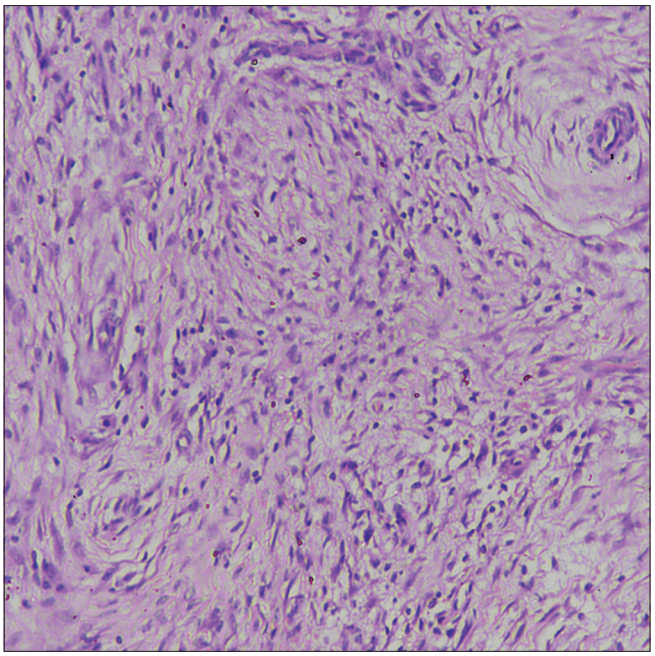

Figure 4: Micrograph (H&E stain, 40×) showing typical histopathological features, i.e., spindle cells in a storiform pattern with admixed foamy histiocytes, lymphocytes, and plasma cells

palisading, balloon cell, signet ring cell, and a few others.[2,5]

They generally occur in young individuals with a female preponderance, as was with the present case.[6] However, they are said to be rare in the first two decades of life, and congenital cases like the present case are very rare.[5]

Clinically, they are solitary nodular swellings with the overlying skin commonly reddish-brown to darkly pigmented and scaly.[2] The size is usually less than 3 cm. However, a giant dermatofibroma can be more than or equal to 5 cm in size.[7] Dermoscopic evaluation of these lesions will most commonly exhibit a central white patch with a peripheral pigmented network.[6]

Histologically, there is hyperplasia of the epidermis with hyperplastic basal keratinocytes. The proliferation of immature keratinocytes is seen. Often, there is a tumor-free zone between the epidermis and dermis known as the Grenz zone.[2,6] Focal extension into the subcutis may be present in a few patients. The differential diagnoses would include dermatofibrosarcoma protuberans (DSP), Kaposi's sarcoma, and BCC.

The tumor is excised with regular follow-up. The recurrence rate is <2% following incomplete excision.[2] Metastasis is rare but has been reported. BFH usually carries a good prognosis.

Congenital cutaneous BFH should be considered as a differential diagnosis in children presenting with congenital swellings. The following learning points were gathered from the present case:

- Tethering of the mass to the periosteum gave an impression of the mandibular origin of the mass.
- FNAC could be inconclusive.
- The lesion may involve important structures like facial nerve fascicles in this case which may require proper dissection and repair.

Declaration of patient consent

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

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Conflicts of interest

There are no conflicts of interest.

Nitin Borkar, Moumita De[1], Jiten Kumar Mishra[1], Shreya Malhotra[2], Shamendra Anand Sahu[1], Amit Kumar Chowhan[3]

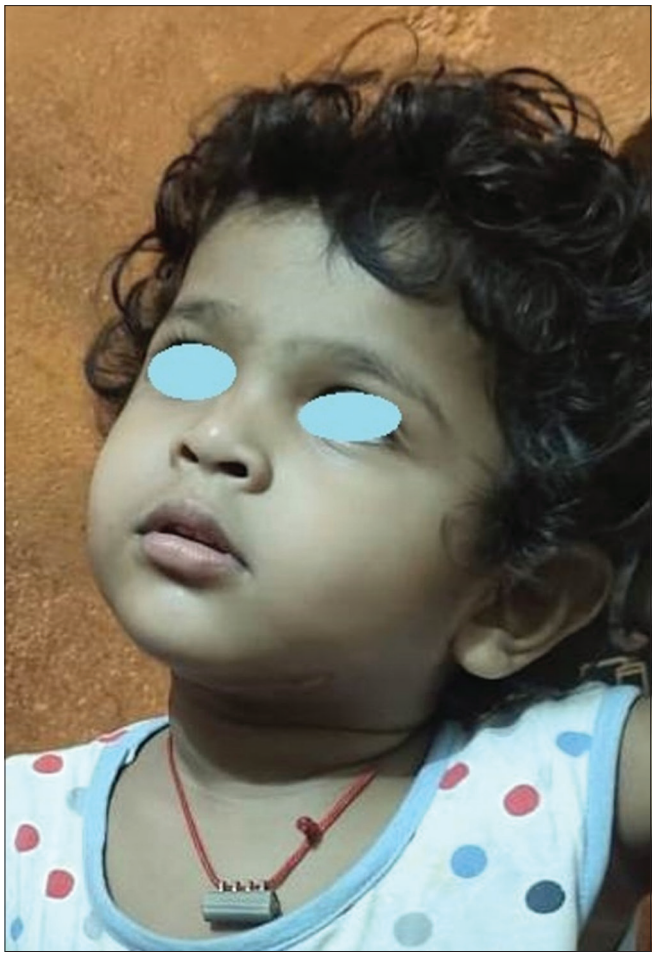

Figure 5: Follow-up at 6 months showing a linear scar with slight hyperpigmentation. The scar is supple without any tenderness. No neurological deficit was seen

Departments of Paediatric Surgery, [1]Burns and Plastic Surgery, [2]General Surgery, and [3]Pathology, All India Institute of Medical Sciences, Raipur, Chhattisgarh, India

Address for correspondence:
Dr. Moumita De,
Department of Burns and Plastic Surgery, All India Institute of Medical Sciences, Raipur, Chhattisgarh, India.
E-mail: drmoumitade@gmail.com

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