Spindle Cell Neoplasm of Bridge of Nose

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ABSTRACT

Introduction

Spindle cells are a feature of many benign and malignant mesenchymal tumors. Differentiating benign versus malignant tumors is based on the presence and type of extracellular matrix, the nuclear and cytoplasmic features of the spindle cells, and the presence of mitoses and necrosis. In bone tumors, the imaging is often predictive. Fibrous histiocytoma is a benign tumor of mesenchymal origin.[1] The tumor frequently appears in sun exposed areas on skin and orbital tissues.[2] The subcutaneous appearance of this tumor in deep soft tissues is rare. It is usually not life-threatening, though it can be aggressive causing serious complications. However, bridge of nose is a very uncommon area for the origin of spindle cell tumour.

Case Report

A 40-year-old woman presented with painful swelling at the left side of bridge of nose and lacrimation from left eye for 4 months. It was oval and hard 2cm x 2cm swelling at the left side of bridge of nose. Computed Tomography showed a well-defined heterogenous soft tissue lesion with erosion of left nasal bone. Magnetic Resonance Imaging showed an isointense oval mass over the left nasal bridge. Aspiration cytology of lesion yielded spindle cells with bland nuclei. The lesion was excised surgically under general anaesthesia. Microscopically the lesion showed oval to spindle cells with bland nuclei arranged in storiform pattern along with inflammatory cell infiltrate. A diagnosis of Benign fibrous histiocytoma is made. Further Immunohistochemistry study confirmed this lesion as spindle cell neoplasm favouring benign fibrous histiocytoma.

Discussion

Spindle cell tumour of bridge of nose is extremely rare. To the best of our knowledge, no such case has been reported till now in English medical literature. In our case, it was a well-localised, painful, mid-face, subdermal swelling without any ocular, oral lesions and nasal bone was only involved. Fine Needle Aspiration Cytology (FNAC), Magnetic resonance imaging (MRI) and Computed Tomography (CT) images are diagnostic tools for Spindle cell tumour. FNAC has a diagnostic accuracy of 84% and is characterised by the presence of spindle cells with bland nuclei. The treatment of spindle cell tumour is surgical and the approach depends on the extent and location of the tumour. In our case, we did Lynch-Howarth approach. Since the clinical presentation and pre-op FNAC did not suggest malignancy, we removed only the tumour and didn't ensure oncological safe margins.

Keywords

Spindle Cell Tumour; Bridge of Nose

Benign spindle cell tumor is a lesion composed of fibroblast-looking cells, usually haphazardly or focally arranged in short fascicles, and closely admixed with thin or thick collagen fibers. Fibrous histiocytoma is a benign soft tissue tumour that may present as a fibrous mass anywhere in the human body.[3] This tumor frequently appears in sun exposed areas on skin. Its incidence has been reported as only 0.2 to 0.5%.[4] It has no gender preponderance. Surgical removal is curative.

This paper describes a case of Spindle cell tumour at the bridge of nose which presented as a painful, oval swelling over the left side of nasal bridge. The lesion was approached through Lynch Howarth incision and was completely resected. No such case of Spindle cell tumour at bridge of nose has been reported till now. The case is reported for its rarity of location and its ability to masquerade other benign lesions like haemangiomas and dermoid cysts, as in our case.

Case Report

A 40-year-old housewife with no co-morbidity presented with a painful swelling over left side of bridge of nose and lacrimation from left eye for last 4 months which

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was insidious in onset, gradually progressive to reach the present size. There were no aggravating or relieving factors. There was no paresthesia over the swelling. On clinical examination we found a localized, oval, 2cm x 2 cm swelling over left nasal bridge extending from 0.1cm medial to medial canthus of left eye to midline of bridge of nose, superiorly from root of nose to left lateral cartilage of nose inferiorly (Figure 1). The overlying skin appeared stretched and tense with no punctum or ulceration. It was tender, hard in consistency, non-mobile, not fixed to the skin with smooth surface and ill-defined margins. The skin was pinchable over the lesion. Intra-oral and anterior rhinoscopy examination were normal and did not reveal any swelling. Diagnostic nasal endoscopy was within normal limit. There was no proptosis and vision was normal. The cervical lymph nodes were not palpable. However, patient had no history of substance abuse. No significant past, allergic or drug history was present. Overall clinical presentation was suggestive of a benign subdermal lesion.

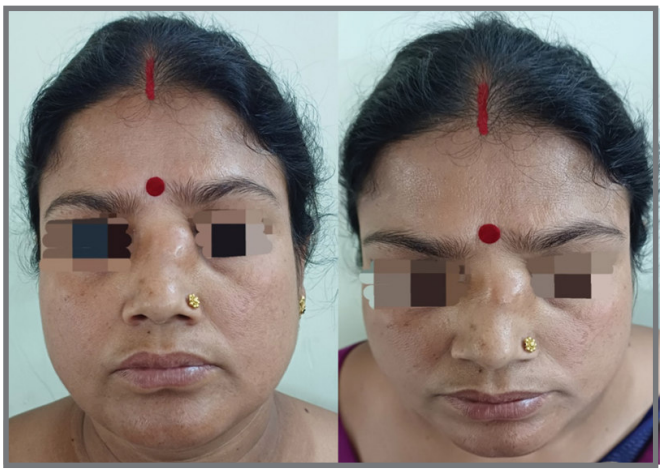


Fig. 1. Lesion presenting as subdermal nodular swelling

Computed Tomography (CT) Scan was advised to study the nature, size and extent of the lesion in relation to nasal bone. It revealed a well-defined heterogenous soft tissue lesion over the left side of bridge of nose with erosion of left nasal bone. There was no evidence of bony erosion or extension of mass into nasal cavity or orbital cavity.

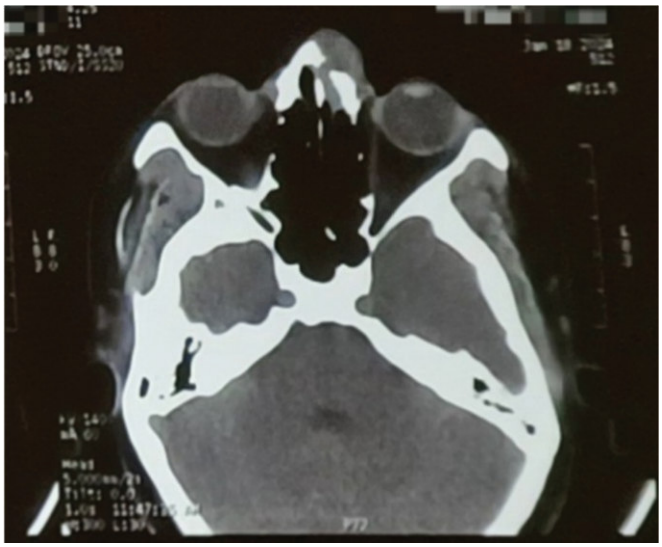


Fig. 2. CT-Scan showing a heterogenous oval mass over left nasal bone

Magnetic Resonance Imaging showed a mass over the left side of nasal bridge. The mass is isointense on T1W, hyperintense on T2W and shows diffusion restriction.

Fine Needle Aspiration Cytology (FNAC) of lesion showed spindle cells with bland nuclei.

A provisional diagnosis of benign spindle cell tumour at the left side of nasal bridge was made.

Surgical excision of the lesion was planned under general anaesthesia. After infiltrating adequately with sterile water and adrenaline, Lynch-Howarth incision was made. Skin flap was elevated, tumour was identified and separated off its margins. Oncological safe margins were not ensured since we did not suspect malignancy. The lesion was not connected to nasal mucosa or skin in the nasofacial groove. Haemostasis was achieved and wound closed in two layers. Post-operative recovery was uneventful.

Histopathological examination of the excised lesion showed predominantly solid and whitish irregular mass. Microscopically the lesion showed oval to spindle cells with bland nuclei arranged in storiform pattern along with inflammatory cell infiltrate. A diagnosis of benign fibrous histiocytoma is made.

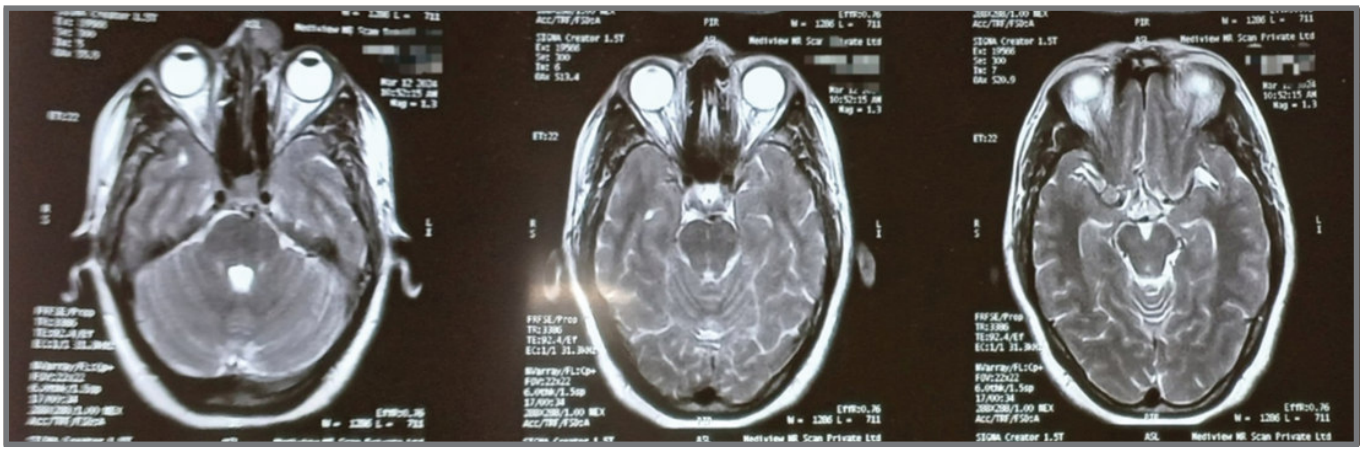
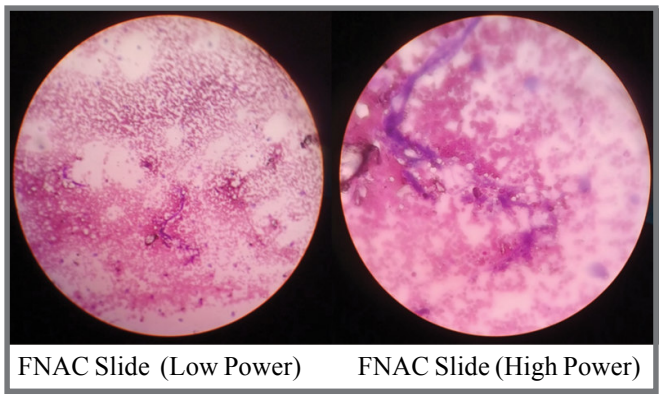


Fig. 3. MRI showing an isointense oval mass over left nasal bridge



FNAC Slide (Low Power) FNAC Slide (High Power)

Fig. 4. H & E-stained section of FNAC slides at 10X magnification

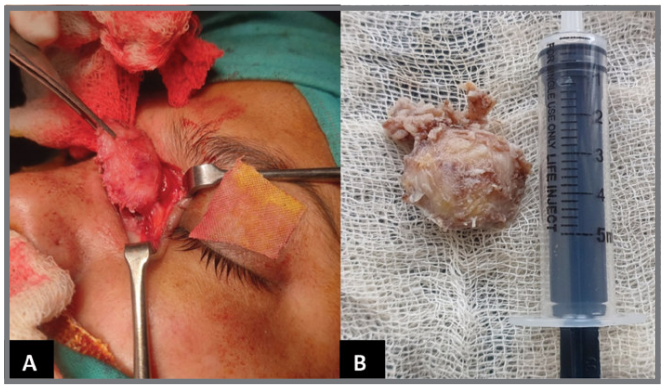
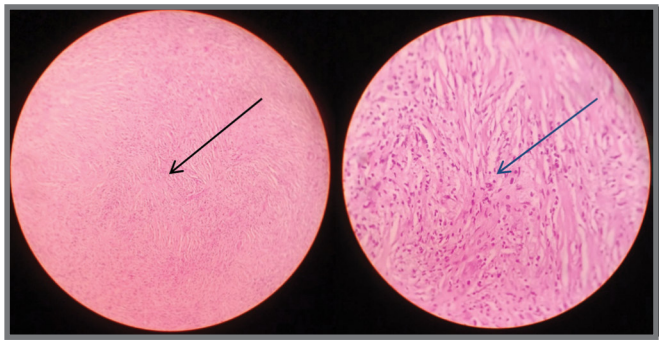


Fig. 5. Lynch-Howarth incision with skin flap elevation, tumour not attached with nasal mucosa or skin(A), Excised tumour (B)



HPE Slide (Low Power) HPE Slide (High Power)

Fig. 6. H & E-stained section of excised specimen at 40X magnification showing Storiform pattern (Black arrow) & Spindle cells with bland nuclei, no nuclear atypia seen (Blue arrow)

Fig. 7. Paraffin wax block of biopsy specimen

Further Immunohistochemistry study showed the tumour is immunopositive for SMA, CD68 and CD10 and patchy weak positive for S100. The tumour is immunonegative for CK, CD34, Desmin, H-CALDESMON, SOX10, STAT6, ERG, and ALK-1. It confirmed that this lesion is spindle cell neoplasm favouring benign fibrous histiocytoma.

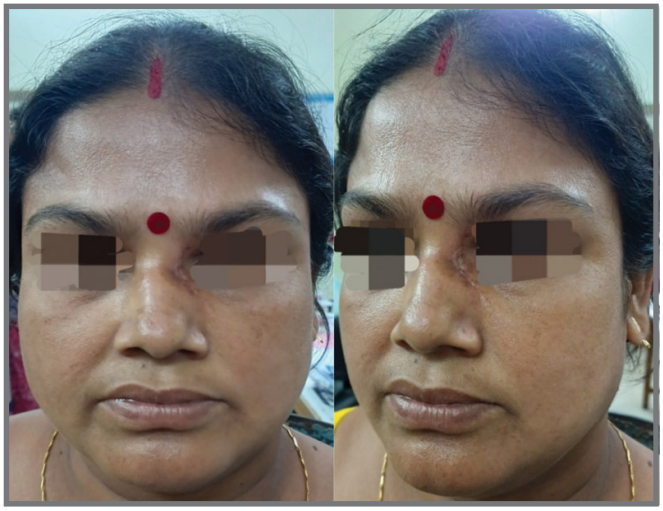


Fig. 8. Post-operative picture of the same patient

Discussion

Benign spindle cell lesions encompass a wide and heterogenous spectrum of fibroblastic and myofibroblastic tumor-like or tumor entities.[5] *Spindle cells* are a feature of many *benign* and malignant mesenchymal *tumors*.

Spindle cell neoplasms arising in the skin comprise a heterogeneous group of tumors with divergent lineages.[6,8] Cutaneous spindle cell neoplasms are relatively common and present surgical pathologists with diagnostic challenges.

Cutaneous neoplasms entirely or predominantly composed of spindle cells constitute a heterogeneous group of mesenchymal and nonmesenchymal tumors.[7] Cutaneous spindle cell neoplasms are often diagnostically challenging because of considerable morphologic overlap among the various tumor types that compose this group.

Cutaneous spindle cell lesions encompass a heterogeneous group of tumors that range from reactive to benign, borderline, and malignant tumors.[8,12] The approach used for the diagnosis of these tumors should be based on knowledge of the relative frequencies of different tumor types, appropriate consideration of clinical context, and correct interpretation of histologic features. Benign lesions are more common than malignant tumors.[9] Fibrous histiocytoma (FH; also known as dermatofibroma) is one of the most common cutaneous mesenchymal neoplasms. Cutaneous soft tissue sarcomas represent less than 1% of malignant tumors.[10]

Fibrous histiocytoma is a common benign dermal mesenchymal tumor characterized histologically by the proliferation of mononuclear, spindle to round or histiocytic cells.[11] Fibrous histiocytoma may develop at any age, but it usually presents during the third and fourth decades. Typically, it occurs on the extremities and trunk and forms a small, dome-shaped elevation that is sometimes pigmented and rarely multiple. Cutaneous FH may rarely metastasize to lung, lymph node, or soft tissue.[12] Although most FHs are dermally based, deep benign FH arises in subcutaneous or deep soft tissue and has a higher recurrence rate than conventional FH.

Cutaneous FH presents as an ill-defined dermal lesion associated with hyperplasia of overlying epidermis. Histologically, the lesion is composed of short spindle and ovoid cells with slightly irregularly shaped nuclei arranged in a storiform fascicular pattern.[13] There is a variable admixture of inflammatory cells, foamy macrophages, Touton giant cells, and siderophages. Entrapped thickened, hyaline collagen bundles are seen at the peripheries of the lesion. Furthermore, hyperplasia of overlying epidermis is present. Immunohistochemically, the tumor cells are often positive for factor XIIIa, SMA, and CD68, and desmin and CD34 are often expressed.[14]

Fibrous histiocytoma(FH) has multiple variants, and many cases exhibit overlapping features of more than one subtype. Furthermore, the recognition of these subtypes is important to achieve accurate diagnosis. The cellular FH variant is characterized by highly cellular spindle cells arranged in a fascicular pattern with increased mitotic activity.[15] Aneurysmal FH shows blood-filled cystic spaces and marked hemosiderin deposition. Epithelioid FH presents a polypoid growth composed of epithelioid cells with vesicular nuclei and abundant eosinophilic cytoplasm.

In addition, the presence of *ALK* gene rearrangement and ALK overexpression in epithelioid FH suggests that epithelioid FH is a biologically distinct tumor type, unrelated to conventional FH and histologic variants.[16] This tumor is another example to express ALK overexpression besides the well-known inflammatory myofibroblastic tumor. Atypical FH displays marked nuclear pleomorphism and hyperchromasia and atypical mitosis.

The differential diagnosis of FH includes Dermatofibrosarcoma and leiomyosarcoma. Dermatofibrosarcoma protuberans shows honeycomb infiltration of subcutis and CD34 expression.[17] Leiomyosarcoma has blunt-ended, parallel-sided nuclei, more eosinophilic cytoplasm, and desmin and h-caldesmon expression. Aneurysmal FH is distinguished from angiomatoid FH by its deep location, fibrous and lymphoid cuff at the periphery, and desmin expression. Atypical FH is distinguished from AFX and superficial pleomorphic sarcomas by its appearance in extremities, epidermal hyperplasia, and the presence of conventional FH features.[18]

The World Health Organisation (WHO) denoted the term "fibrohistiocytic" to a lesion composed of cells resembling round histiocytic and spindle fibroblastic morphology.[19] Vimentin and CD68 positivity reflect its fibroblastic and histiocytic heritage.[20] Prognosis is good and without any recurrence.

Spindle cell tumour of bridge of nose is extremely rare. To the best of our knowledge, no such case has been reported till now in English medical literature.

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The treatment of spindle cell tumour is surgical and the approach depends on the extent and location of the tumour. In our case, we did Lynch-Howarth approach. Since the clinical presentation and pre-op FNAC did not suggest malignancy, we removed only the tumour and didn't ensure oncological safe margins.

Conclusion

Incidence of Spindle cell tumour over nasal bridge is very low making it difficult to diagnose. It is rare for a Spindle cell tumour to arise denovo in connective tissue planes. It is possible to have a Spindle cell tumour not connected to nasal mucosa or skin over the nasal bridge. This is a distinct possibility in our case. Hence, Spindle cell tumour of bridge of nose may be included in the differential diagnosis of a localised subdermal mid-face swelling.

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