Preliminary Report of a True NasoMaxillary Infantile Fibrosarcoma: Single-Modality Management and 2-Year Follow-Up

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Abstract

Infantile fibrosarcoma (IFS) is a malignant tumor, commonly presenting in long bones and seldom encountered after 2 years of age. It is extremely rare in the faciomaxillary region. The condition is often mistaken for teratomas, and histopathology/immunohistochemistry is confirmatory. Treatment involves surgical resection as primary modality. Prognosis is much better compared to the adult variant with even scope for spontaneous regression following "conservative" resection. We present a case of IFS of the nasomaxillary complex in a 3-month-old female child. We managed our case with surgical resection, and the patient has remained disease free for over 2 years.

Keywords: Fibroblastic tumor, hemangioma, infantile fibrosarcoma, teratoma

Introduction

Infantile fibrosarcoma (IFS) is a malignant fibroblastic tumor usually arising in the extremities. It has an incidence of 1% of all childhood malignancies and is even rarer in the faciomaxillary region (0.05%).[1] IFS is morphologically and genetically related to congenital mesoblastic nephroma. It is clinically more likely to be confused as a teratoma or hemangioma. No known etiological factors have been proven beyond doubts, though a few sporadic reports of prenatal radiation, Gardener's syndrome, etc., have been published. Although being histologically similar to the adult variant, it has a favorable prognosis with less likelihood of metastasis. Early diagnosis is the key to successful management.

Case Report

A 3-month-old female child presented with a progressively expanding mass in the upper jaw for 1 month of age. There were apparently no associated feeding or breathing difficulties. The full-term, transvaginally delivered baby had no relevant medical, social, or family history attributable to the current affliction. On examination, a single, spherical bluish mass in the anterior maxillary vestibule was seen extending on to the upper lip. There was considerable elevation of the alar base and obliteration of the right nasal cavity [Figure 1]. This sessile mass measuring about 2 cm × 2 cm was soft in consistency and was neither expansile nor pulsatile. Aspiration was inconclusive. A wide bore 48 channel 1.5 T-high-field magnetic resonance imaging (MRI) revealed a soft-tissue mass in the central aspect of the right maxilla extending into nasal cavity with cranial displacement of the deciduous central incisor [Figure 2a]. The child was subsequently taken up for excision of the nasomaxillary mass under general anesthesia [Figure 2b and c]. Histopathological analysis reported IFS (Grade 1) and that the margins were free of tumor. Microscopy revealed densely packed spindle cells arranged in fascicles and demonstrating herringbone pattern [Figure 3]. Immunohistochemical staining reported positivity for vimentin [Figure 4], smooth muscle actin,

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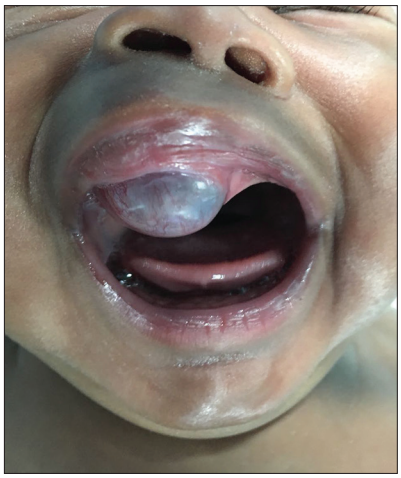

Figure 1: Preoperative image showing nasomaxillary mass causing cheilo-nasal dysmorphism

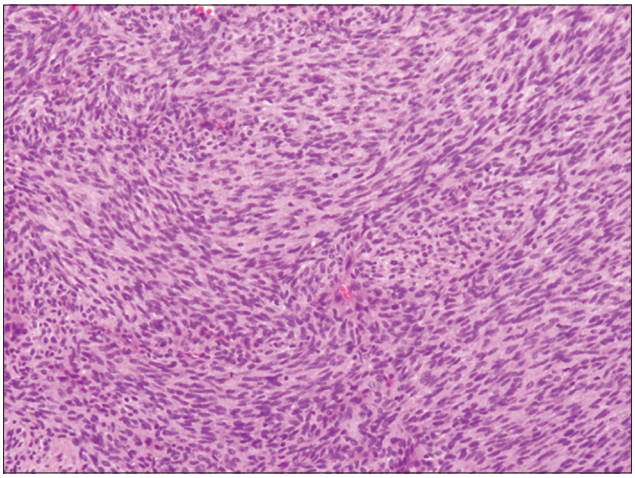

Figure 3: Photomicrograph showing cellular spindle cell lesion arranged in fascicles with "herringbone pattern"

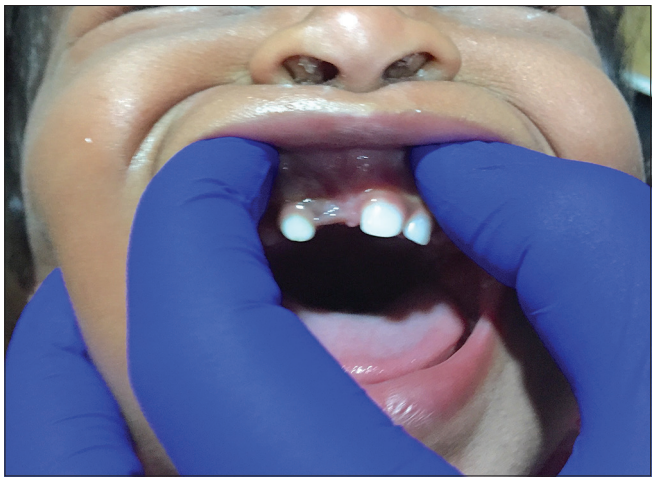

Figure 5: Eighteen months' postoperative follow-up showing complete regain of facial architecture

and CD 34 while being negative for S100, CD99, BCL-2 myogenin, and desmine. Postoperatively, the child was

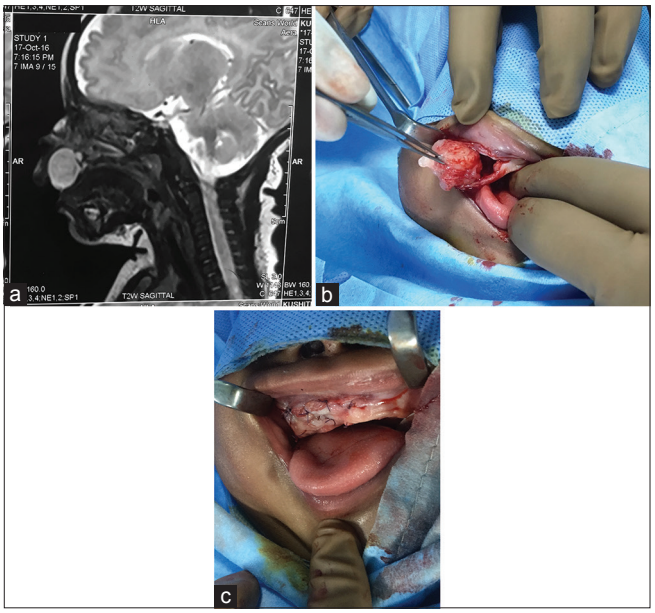

Figure 2: (a) Preoperative magnetic resonance imaging showing soft-tissue mass in the nasomaxillary region. (b) Delivery of the excised mass. (c) Closure of the defect

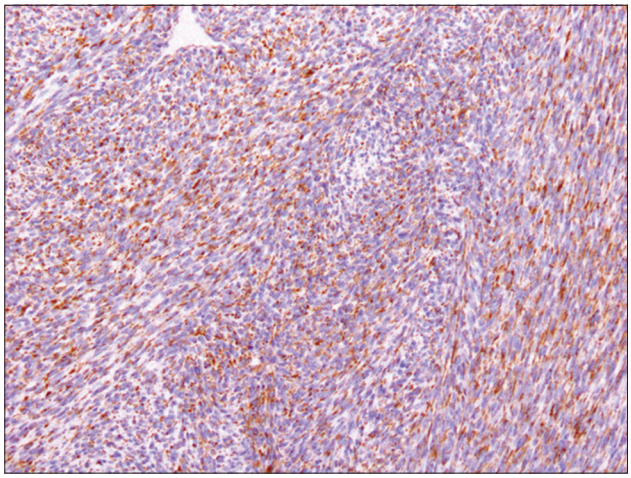

Figure 4: Photomicrograph showing vimentin positivity in neoplastic cells

referred to medical oncologist who deferred chemotherapy considering low-grade and tumor-free margins. The patient was hence planned for close observation and follow-up. The patient is being followed up for the last 2 years and has remained disease free so far [Figure 5].

Discussion

IFS is a soft-tissue malignancy usually involving extremities. Etiology is unknown, and no hereditary diseases, predisposing factors, or causative agents have been demonstrated so far, excluding reports associating it with prenatal irradiation and Gardener's syndrome. It characteristically presents as a rapidly enlarging solitary mass in the soft tissues very similar to our case. It is very rare in the maxillofacial region. Sporadic reports of "infantile" fibrosarcomas in children have been published

involving tongue,[1] orbit, cheek, mastoid process[2] maxilla, and mandible.[3]

Differences between infantile and the adult fibrosarcomas are a topic attracting wide range of discussion, including onset, clinical features, histopathology, treatment strategy, and outcome. IFSs occur during the 1st year of life and often present at birth as against the adult variant which has peak incidence around the fourth decade, a subset of which originates after irradiation or burns. Considering suggestions of the World Health Organization (WHO) regarding the age of presentation to be addressed as the true infantile variant (younger than 2 years),[4] our case is probably the first ever reported to be occurring in the maxilla. The WHO also proposes that, for cases to be reported as the infantile variant beyond this age, cytogenetic confirmation has to be mandated. Cytogenetic studies of IFS have revealed several abnormalities, particularly a t (12;15) (p13;q25) resulting in ETV6-NTRK3 gene fusion (readily diagnosed by molecular methods) and a nonrandom gain of extra chromosomes.

Infantile type has a slight male predilection as against the adult type, which is gender neutral. Infants usually present with a rapidly expanding tumor causing mass effect correlating with the site of occurrence. Feeding and breathing difficulties are common presenting signs for the fibrosarcomas occurring in the maxillofacial region.

Although it is generally agreed that the infantile variant is histologically identical to the classic adult fibrosarcoma, certain specific differences proposed were the characteristic spindle-shaped fibroblastic cells which formed interlacing bundles accompanied by variable amounts of collagen or reticulin fibers in the adult type, whereas the IFSs are characterized by proliferation of immature fibroblasts forming indistinct bundles. Quite often, this phenomenon may exhibit areas of angiosarcoma-like pattern and cavernous blood vessels. This explains why it is commonly confused to be a hemangioma or teratoma. Histopathology in our case as expected revealed fascicles of spindle cells arranged in a characteristic "herringbone" pattern which is pathognomonic of fibrosarcomas.

MRI characteristically reveals soft-tissue mass with heterogenous enhancement pattern and variable osseous erosion, very similar to the presentation in our case. Diagnosis of IFS is usually by imaging, while histopathology and immunohistochemistry are confirmatory. Positivity of vimentin is 100% in IFS.

Complete resection is usually curative, and chemotherapy can be reserved for inoperable tumors or involved margins.[5,6] For all clinical purposes, it has the natural course similar to that of fibromatoses. Fortunately, as against the adult type, it has a more favorable clinical course and prognosis[5] (<10% metastasis) and rarely requires adjuvant therapy beyond excision. This is contrasting to 5-year survival rate of 40-50% in the adult variety depending on the histologic grading which varies depending on the severity of cytologic atypia and the character of stroma. Our patient has been on follow-up for over 2 years now and has remained disease free till date.

Learning points:

1. IFSs though rare must be kept in the differential diagnosis of tumors of infancy
2. They are often mistaken for teratomas or hemangiomas
3. Diagnosis is by imaging and pathology
4. Early surgical resection is usually curative
5. Regular follow-up is of prime importance.

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.

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