CASE REPORT

Primary fibrosarcoma of maxilla in an 8-year-old child: A rare entity

Niharika Swain, Shwetha V Kumar[1], Richa Dhariwal[1], Samapika Routray[2]
Departments of Oral Pathology, Mahatma Gandhi Mission Dental College and Hospital, Kamothe, Navi, Mumbai, Maharashtra, [1]Oral Pathology, Dr. R. Ahmed Dental College and Hospital, Kolkata, West Bengal, [2]Institute of Dental Sciences, Bhubaneswar, Odisha, India.

Address for correspondence:
Dr. Niharika Swain,
304, Siddhi Vinayak Residency, Sector-9,
Plot no-40, Kamothe, Navi Mumbai - 410 209,
Maharashtra, India.
E-mail-niharikadec30@gmil.com

ABSTRACT

Fibrosarcoma (FS) is a malignant mesenchymal neoplasm of the fibroblasts that is uncommon in the head and neck and constitutes less than 1% of malignancies and approximately 6% of the soft tissue sarcomas. FSs rarely occur before the third decade except infantile type. This condition primarily affects long bones, and its occurrence in the cranium is rare (15%), with the mandible being the most commonly involved cranial site. Here a case of primary FS in anterior maxilla of an 8-year-old male child is reported. This article is presented to document the rarity of FSs in the jaws of children with review of literature.

Key words: Bone, child, fibrosarcoma, maxilla

INTRODUCTION

Fibrosarcoma (FS) is a malignant neoplasm of the fibroblastic origin. Fibrosarcomas are extremely rare lesions in the oral cavity and maxillofacial area accounting for 10% of the head and neck region.[1] Its occurrence in jaws, especially in the maxilla, is rare with an incidence ranging from 0% to 6.1% of all primary FS of the bone.[2,3]

Here we report a case of primary FS of maxilla in a child, which is probably the first ever such presentation according to the previous English literature.

CASE REPORT

An 8-year-old male child reported with the complaint of fast growing swelling over middle of face. The patient did not give any history of systemic illness or trauma to the head and neck region. There was no significant contributing family history. He gave a history of painless but rapidly growing swelling in the anterior maxilla since 20 days, which attained to the present size of 7×10 cm. There was no associated history of difficulty in speech and mastication with incomplete closure of mouth. The patient had also given the history of previous incisional biopsy, which was conducted by a general physician with histopathological report of benign fibrous lesion. Intraorally, a globular sessile mass with an area of ulceration due to previous biopsy procedure was present in the anterior hard palate extending from 55 to 65 [Figure 1]. The buccal and palatal cortical plates were completely destroyed. Missing regional teeth was also noticed, which had the history of spontaneous exfoliation. The labial vestibule was completely obliterated. The orthopantomograph (OPG) revealed a purely radiolucent destructive lesion in the anterior maxilla [Figure 2]. A chest radiograph was advised to rule out the metastasis, which was found to be clear without any significant and relevant finding. With a clinical provisional diagnosis of bone malignancy, the intraoral mass was subjected to incisional biopsy. The Hematoxylin and Eosin (H and E) stained section showed nonencapsulated tumor tissue made up of highly cellular stroma arranged in interlacing fascicles [Figure 3]. The tumor tissue showed the presence of spindle-shaped cells with large nuclei and scanty cytoplasm. Mitotic figures (6-7/10 HPF) were observed [Figure 4]. In few areas, Herring bone pattern characteristic of FS was also noticed. Immunohistochemically, vimentin positive cells were abundant [Figure 5], whereas other immunomarkers like desmin, smooth muscle actin (SMA), S-100 were negative. A histopathological diagnosis of primary intraosseous FS (grade-II) was made. Patient had undergone surgical excision. The excisional biopsy was also consistent with the diagnosis of incisional biopsy. Finally the patient succumbed to the tumor within time period of 6 months of diagnosis.

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DISCUSSION

FS is a malignant mesenchymal neoplasm of the fibroblasts that rarely affects the oral cavity, which may either arise in the soft tissue or be of primary intraosseous origin. (Between

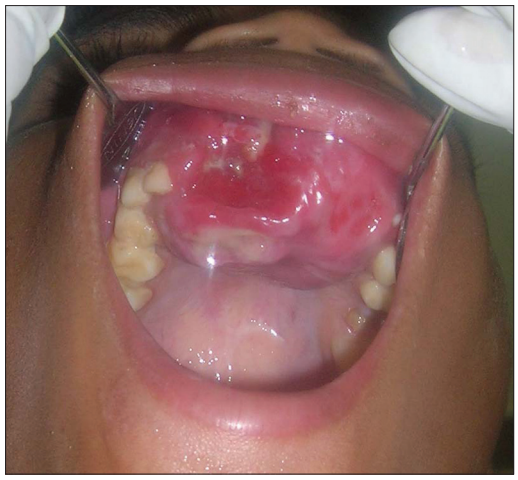

Figure 1: A large swelling on anterior maxilla extending from 55 to 65

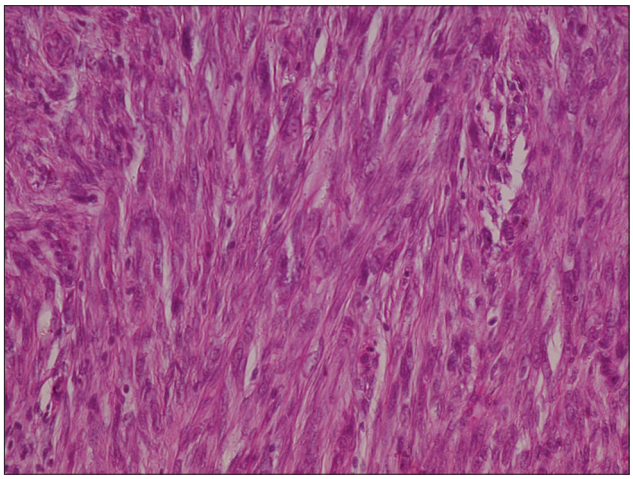

Figure 3: Highly cellular stroma arranged in interlacing fascicles showing herringbone pattern (H&E stain, ×400)

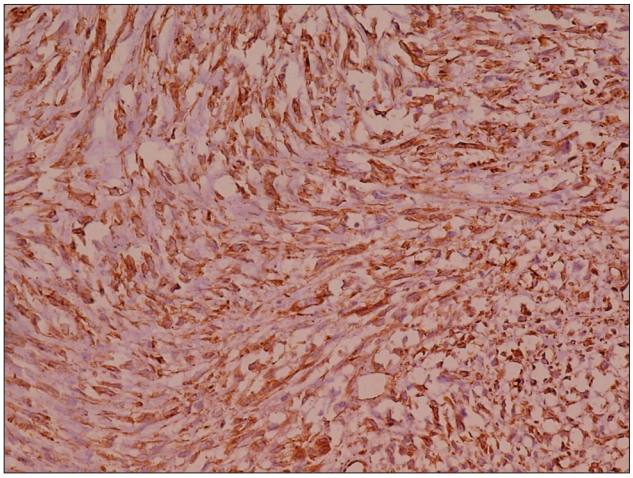

Figure 5: Tumor cells showing intense immunoreactivity for vimentin (IHC stain, ×200)

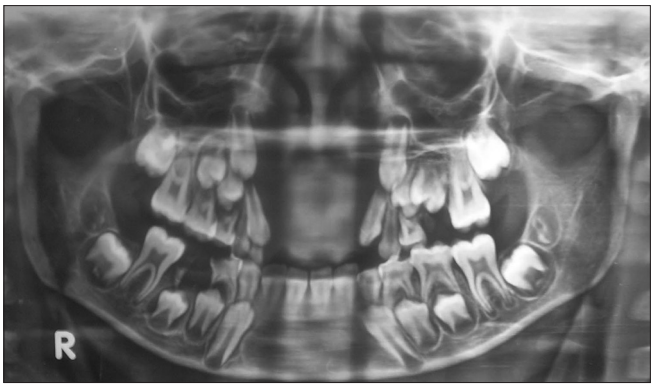

Figure 2: Orthopantomograph revealed a purely osteolytic lesion in the anterior maxilla

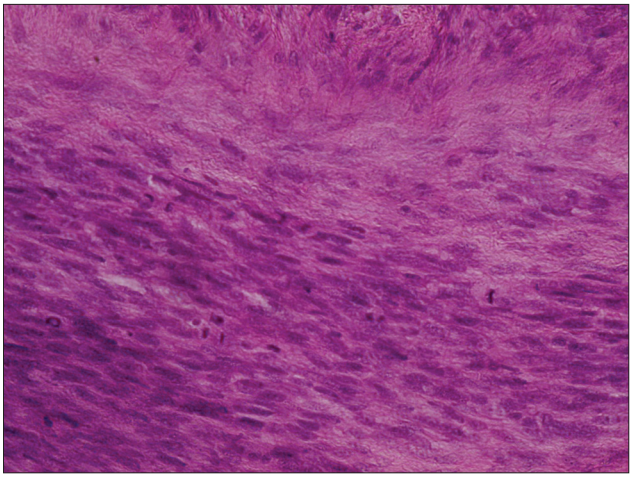

Figure 4: Neoplastic spindle-shaped cells showing mitotic figures (H&E stain,×400)

1950 and 1975, this was believed to be the most common soft tissue malignancy. However, the identification and separation of other similar spindle cell tumors, all of which were previously regarded as FSs, has unmasked the rarity of true FS.) FS may arise as a primary tumor in any part of the jaws and may be classified as of either peripheral (periosteal) or central (endosteal) type.[4-6] It is more frequent in the fifth and sixth decades of age but cases in children and adolescents were also described in the literature.[7-9] Because of different clinical behavior and distinctive molecular alterations, infantile fibrosarcoma (IFS) must be considered a separate entity from the identical lesion in adults. Previous literature did not show any unanimous opinion regarding the cut-off age for use of term infantile FS. Some authors have mentioned a cut-off age of 2 years,[10,11] whereas others have suggested a cut-off age up to 5 years.[12-15] Extensive review of previous literatures showed only few cases of primary intraosseous FS affecting jaws of children (age group ranging 2-13 years) as depicted in Table 1. We have considered 2 years to be the cut-off age for IFS as suggested by World Health Organization (WHO).[19] As all the reported cases of primary FS in children affected the mandible, the present case is probably the first ever reported case of primary FS of maxilla in present literature.

The most frequent clinical manifestations of FS of jaw are pain and swelling. Other associated symptoms like loosening of teeth, trismus, pathological fracture, and parasthesia may be

Table 1: Summary of clinical findings for reported cases of intraosseous fibrosarcoma of the jaw in children

Reference	Age/gender	Site	Diagnosis	Treatment	Follow up	Outcome
Van Blarcom *et al*.[16]	13 years/F	Body, angle and ramus of mandible	Grade 2	NS	Lost to follow up	NS
Dehner LP *et al*.[7]	7 years/M	Body of mandible	Well diff.	Hemimandibulectomy and neck dissection	5.5 years	No recurrences Living well
Slootweget al.[17]	11 years/F	Ramus of mandible	Well diff.	Radiation	2 years	Local recurrences Died of disease
Bang *et al*.[14]	2.5 years/M	Body of mandible	Fibrosarcoma	Local and radical Excision	15 years	Recurred 5 times Living well
Lo Muzio *et al*.[2]	4 years/M	Body of mandible	Fibrosarcoma	Radicular surgery	4 years	No recurrences
Divya *et al*.[18]	10 years/F	Parasymphysis to angle of mandible	Well diff.	Partial mandibuletomy	8 months	No recurrences
Present case	8 years/M	Anterior maxilla	Moderately diff.	Surgical exicision	6 months	Died of disease

NS*: Not stated

present.[17,20-22] Radiologically they share similar features with other malignant tumors. FS of bone appears as an ill defined lytic lesion with a destructive pattern and minimal internal structure[2,23,24] but few authors also reported about the well circumscription or cyst like appearance of the lesion.[16,18]

Histopathologically FSs of bone are invasive tumors with no distinct margins. The cells are rather uniform and spindle shaped and arranged in fascicles, often forming a herringbone pattern. Histologically, the degree of differentiation of this neoplasm is variable, comparable to either a benign fibroma or an anaplastic tumor. Therefore care should be taken to distinguish it from other spindle cell neoplasm. Combined histological and immunohistochemical analysis aid in the definitive diagnosis of such spindle cell lesions. Histological grading of FS of bone is based on the degree of cellularity, degree of cellular differentiation, mitotic activity, the amount of collagen produced by the tumor cells, and the extent of necrosis. Van Blarcom *et al*., categorized this lesion into four grades (grade 1 being best differentiated to grade 4 being least well differentiated) on the basis of cellular differentiation by Broders' method.[16] Using the following criteria of amount of fibers, nuclear atypia, and mitotic figures, Taconis *et al*., graded FSs of jaws into three types. Grade I – well differentiated, less than 2 mitoses in 20 HPF, Grade II – moderately differentiated less than 10 mitoses in 20 HPF, and Grade III – high mitotic rate, 10 or more mitoses in 20 HPF.[25] Depending on the number of mitotic figures, tumor differentiation, and the presence of tumor-necrosis, French Fédération Nationale des Centres de Lutte Contre le Cancer (FNCLCC) grading system is currently, the most widely accepted.[9,26,27] Most common complication of FS is frequent local recurrences, which can lead to infiltration, local destruction, airway compression, esophageal compression, and extension into the mediastinum Although incidence of local and distant metastasis is low, both lymphatic and hematogenous metastasis can worsen the prognosis by involving lungs, mediastinum, abdominal cavity, and bone.[24,28-31]

Treatment of choice for FS of bone is wide resection with clear margin.[17,32,33] Prophylactic lymph node dissection is not required.[34,35] In FSs that cannot be completely excised because of their location or extreme size, postoperative radiotherapy of 6000-7,000 cGy is appropriate.[1,20,36] In grade III FSs, postoperative adjunctive chemotherapy is recommended, ostensibly to treat potential subclinical or microscopic metastasis.[11] When chemotherapy is employed, agents used successfully for sarcomas are preferred, including adriamycin, actinomycin D, oncovin, cyclophosphamide, prednisone, and daunorubicin.[1]

Apart from the treatment modality, the prognosis of FS is significantly influenced by site of origin and histopathological grading of the neoplasm. Unlike soft tissue FS, FS of bone has a poorer prognosis with 5-year survival rate of 4.2-31.7%.[37,38] A difference in clinical course between FS of jaw and its long bone counterpart has been reported by many authors. A consistent favorable prognosis of FS of jaw has been observed during the 5, 10, and 20 year observation period as compared with long bone FSs.[25,39,40] Like most sarcomas, there is a strong correlation between the prognosis of FS and its histologic grade. Low grade differentiation influences the survival rates in a negative way.[1,26,39,41]

CONCLUSION

Although FS of jaws are rare, meticulous patient histories, detailed clinical examination, and appropriate radiographic examination, careful histopathological and immunohistochemical analysis are very important in the evaluation of such cases. Oral physicians must always consider the possibility and be able to recognize the features of FS to propose appropriate investigations and help in treatment planning. Though FS of jaw has a better prognosis in comparison to the long bone counterpart, Scarcity of follow up data of FS of jaws in children could not provide any significant conclusion regarding the prognosis of this neoplasm.

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