Intraoral Soft Tissue Fibrosarcoma in a Four-Year-Old Child: An Unusual Case Report

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Fibrosarcoma is a rare soft tissue tumor of connective tissue origin that includes about 0.05% of all the malignancies in the head and neck region of which almost 23% is seen in the oral cavity. This paper describes a rare case of 4-year-old boy who presented with swelling on the right side of face diagnosed as soft tissue fibrosarcoma of the intraoral region. The histopathological and immunohistochemistry confirmed the diagnosis by the presence of spindle-shaped cells arranged in fascicles with mitotic figures and cellular proliferation reproducing fibroblasts. The patient was successfully treated with combination of chemotherapy and surgery with a good clinical outcome. This case report is presented to highlight the rarity of fibrosarcoma in orofacial region of children which requires special attention of pediatric dentist and should be considered as differential diagnose of soft tissue mass in orofacial region of children. Clinical and histopathological features must be correlated with immunohistochemistry in the final diagnosis in fibrosarcoma.

Keywords: *Fibrosarcoma, child, mandible, Immunohistochemistry*

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INTRODUCTION

Fibrosarcoma is a rare malignant neoplasm of fibrous connective tissue origin that may arise either within the soft tissue or may have a primary intraosseous origin.[1] Of all the fibrosarcoma occurring within humans, only 0.05% occur in the head and neck region. Out of these, 23% of head and neck fibrosarcomas occur in the oral cavity with the mandible being the most common site. [2,3] Clinically, fibrosarcoma most often presents as slow growing masses that may reach considerable size before producing pain, swelling, loosening of the teeth paresthesia and occasional ulceration of the overlying mucosa.[1,4,5] Histologically, well differentiated fibrosarcomas consist of fascicles of spindle shaped cells that classically form a "herringbone pattern." [6] The diagnosis of the tumor is based on various imaging modalities like magnetic imaging resonance (MRI), computed tomography (CT) but the confirmatory diagnosis is obtained by histopathological evaluation and immunohistochemistry (IHC).[7] Surgery alone or in combination with chemotherapy and radiotherapy remains the mainstay of the treatment.[8] In clinical practice, pediatric dentists encounter a spectrum of different tumors and tumor like lesions in the orofacial region of children and more likely to first examine. Therefore a detailed description of clinical, histopathologic and immunohistochemical features of an intraoral soft tissue fibrosarcoma in a four-year-old child has been described.

Case report

A 4-year-old male child was referred to the department of Pedodontics with the chief complaint of rapidly growing painless swelling on the right side of the cheek region over last six months. The parents reported that the child was having difficulty during mastication with no associated history of difficulty in speaking and mouth opening. On anamnesis the parents did not give any preceding history of systemic illness and trauma to the head and neck region.

On extraoral examination face was asymmetrical with swelling present in lower right region of face where the overlying skin appeared to be stretched (Fig 1a). Intraorally, the swelling was present in right buccal vestibular region presenting as firm, immobile, reddish mass measuring 5.5 cm × 4 cm covered with fibropurulent discharge extending from retromolar region to the angle of the mouth. There was obliteration of right buccal vestibule with associated ulceration in relation to the fibrous mass (Fig 1b). There was absence of tooth mobility in the lower right quadrant of the jaw and the regional lymph nodes were non palpable. Panoramic radiograph (OPG) revealed absence of any radiolucency in relation to lower right region of the mandible (Fig 1c).

The patient was referred to the department from the associated medical college for incisional biopsy of the swelling to rule out any malignancy. The patient had already undergone ultrasonography (USG) of the right cheek region and fine needle aspiration cytology. USG of the right cheek region revealed a well defined heterogeneous lesion measuring 37mm × 28mm along with the internal calcification and no dominant vascularity and the cytological report of lesion showed small cells having scanty cytoplasm and vesicular nuclei arranged in clusters with vague acinar pattern giving the impression of minor salivary gland tumor. An incisional biopsy under local anesthesia was performed for histopathological evaluation which revealed presence of spindle shaped cells surrounded by collagen fibers resembling that of fibrosarcoma after which the neoadjuvant chemotherapy was administered followed by the complete surgical excision of the lesion under general anesthesia. The histopathology of the excised lesion showed spindle cells arranged in fascicles with varying shapes and size and scanty cytoplasm with indistinct cell borders which were separated by interwoven collagen fibers arranged in parallel fashion (Fig 2a). A high number of mitotic figures (8-9/10 HPF) were observed within the lesional tissue (Fig 2b). A strategic panel of immunohistochemical markers was applied to aid in the diagnosis, which included S100, Nestin, TLE 1, EMA, CK7, CK19, P63 and calponin. Mild patchy positivity was noted only with S-100 (Fig 3) whereas all other tumor markers were negative. Based on USG, cytology, biopsy and Immunohistochemistry (IHC) report of lesion confirmed diagnosis of fibrosarcoma was made. The child is under routine follow up and shows no signs of recurrence and metastasis after 18 months (Fig 4a-b).

Fig 1a) Extraoral photograph showing swelling of lower right side of face; b) Intraoral photograph showing solitary mass of tissue obliterating right buccal vestibule and c) Panoramic radiograph (OPG) revealed absence of any radiolucency in the lower right region of the mandible.

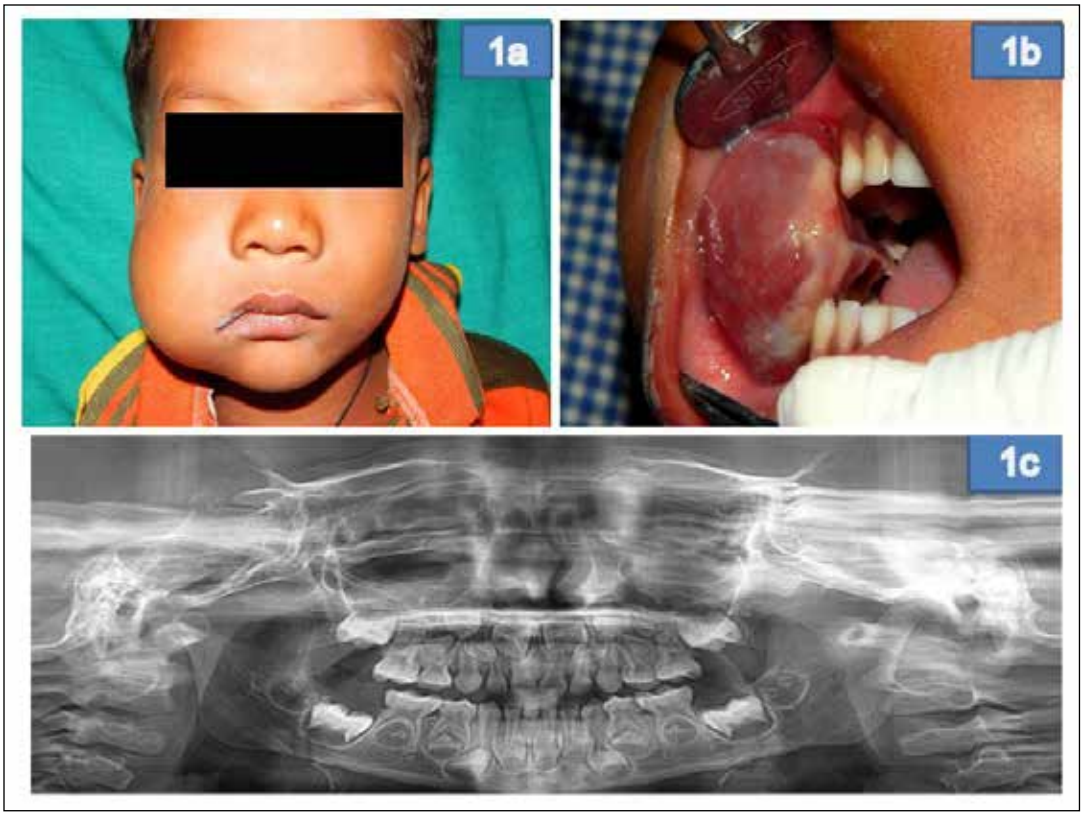


Fig 2a) Histopathological photograph showing spindle shaped tumor cells arranged in fascicles (100X magnification); b) High power field showing mitotic figure (8-10 mitotic figures per HPF) (400X magnification)

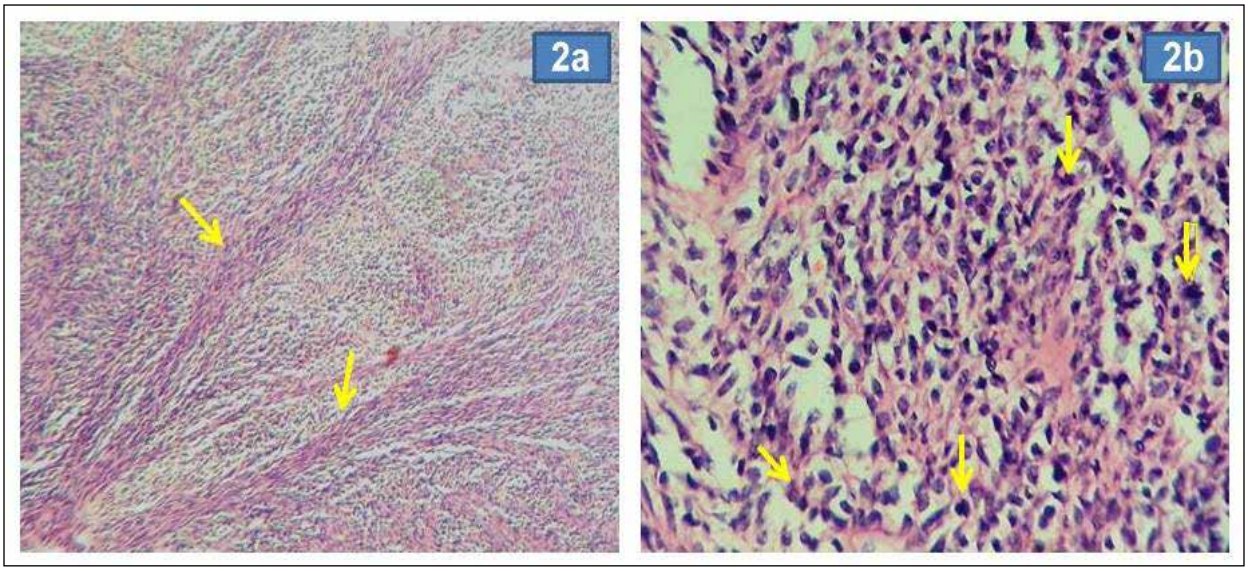


Figure 3 Immunohistochemistry shows patchy positiveness for S100 antibody

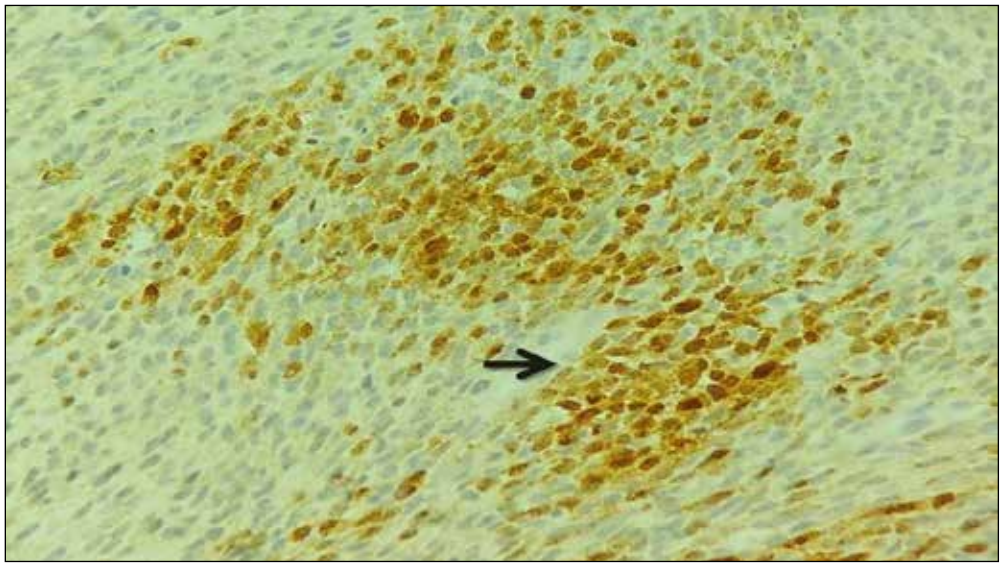
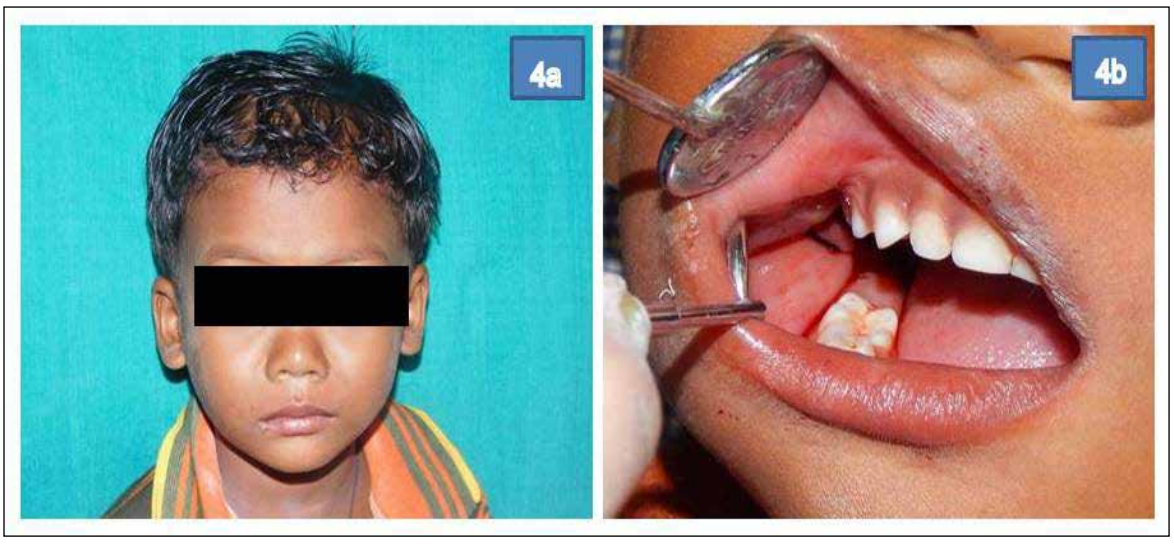


Fig 4 Post-operative view after 18 months follow up a) Extraoral view b) Intraoral view



DISCUSSION

Fibrosarcoma is an uncommon malignant neoplasm of fibroblast with high recurrence rate but rare incidence of metastasis.[2] It is classified in the heterogeneous group of non rhabdomyosarcoma malignant mesenchymal tumor.[9] Similar to other sarcomas, it has no characteristic symptoms and is difficult to diagnose clinically. Depending on the era and criteria in vogue, the incidence of fibrosarcoma has varied widely with time. In 1936 fibrosarcoma was considered to comprise 65% of all the soft tissue sarcomas.[10] However, the number was revised to 12% in 1974 and even lesser in 1989.[11,12] Several factors like recognition of malignant fibrous histiocytoma and fibromatoses as separate entities were responsible for such sharp decline in its incidence. The occurrence of the neoplasm in head and neck region, particularly in jaws is quite low in comparison to deep soft tissues of lower and upper extremities, where the frequency is much higher. In jaws, it is more commonly seen in the mandible and may arise as a primary tumor from any part and can be classified as peripheral or central type.[13] The mean age of occurrence of fibrosarcoma is between 2nd and 6th decade, with equal gender predilection, although, cases have been described among children in the literature.[5] Infantile fibrosarcoma (IF) is usually reported in the first five years, with a preponderance under three years of age and 40 percent of cases are recorded below three months of age and rarely affects older age group between 10 to 15 years.[14] There is a controversy regarding the cut-off age for the use of term infantile fibrosarcoma regarding which the WHO has suggested a cut off age of 2 years for the diagnosis of IF.[5,15]Though, infantile and adult fibrosarcoma are histologically identical, their clinical behavior and molecular pathogenesis are different. In the present case the patient was four years old i.e. the first decade of life, when the diagnosis of fibrosarcoma was given.

The most commonly occurring clinical manifestations of fibrosarcoma of the jaw are pain and swelling which may vary depending on the size, location and spread of the tumor. Other related symptoms like loosening of teeth, trismus, pathological fracture and paresthesia may also be present. [1,4,5] Fibrosarcoma in the oral cavity generally manifests as a clinically innocuous, painless, lobulated, sessile and non hemorrhagic submucosal mass of normal coloration.[8] An aggressive or high grade of fibrosarcoma mainly presents as rapidly enlarging hemorrhagic mass which has a similar clinical appearance to an ulcerated peripheral giant cell granuloma, pyogenic granuloma or peripheral ossifying fibroma. Moreover, the lesion might show destruction of the underlying muscle and bone.[16,17] Although these symptoms were not seen in the present case suggesting a variability in the clinical presentation of the lesion. Radiological imaging of fibrosarcoma reveals an ill defined radiolucent lesion with a moth-eaten or permeative pattern of bone destruction.[8] Ironically, the panoramic view in our case did not demonstrate the presence of any osteolytic lesion or breach within the cortical bone. Histopathologically, fibrosarcoma shows minor variation, but primarily presents in fasciculated growth pattern containing spindle shaped tumor cells having scanty cytoplasm and indistinct cell membrane mainly separated by parallel strands of collagen fibres. In some cases the cells are oriented in curving or interlacing fascicles, forming a classic herringbone pattern.[2,18] It is classified histologically into high grade and low grade fibrosarcoma. In the present case, although the mitotic activity was high, other parameters like arrangement of tumor cells and absence of necrosis were considered and the lesion was categorized as a low grade fibrosarcoma.[8] Low grade fibrosarcoma can present a high level of cellularity and variable amount of collagen. Due to the varying degrees of differentiation, it presents with a diagnostic dilemma. Therefore, utmost care should be undertaken to distinguish it from other spindle cell neoplasms. The diagnosis of fibrosarcoma should chiefly be carried out on the basis of exclusion principle. To prevent any bias in the definitive diagnosis for the present case, histopathological evaluation was followed by IHC. A panel of IHC markers was employed to exclude similar lesions such as malignant peripheral nerve sheath tumor (MPNST), monophasic fibrous synovial sarcoma and myoepithelial carcinoma. The positive and negative immunostaining for S100 and nestin respectively, helped us to exclude the possibility of MPNST. The immunostaining was negative for EMA, TLE1, CK19 and CK7 which ruled out monophasic fibrous synovial sarcoma. Also, a negative p63 immuno expression eliminated myoepithlial carcinoma from the differential diagnoses.[16] Thus, a final diagnosis of fibrosarcoma was made, on the principle of exclusion.

Surgical resection with a wide margin currently remains as the mainstay of the treatment, although the surgical approach has clearly evolved over the years, from primary treatment modality to being part of a multidisciplinary strategy and from mutilating operations to more conservative organ-sparing procedures.[1,2,16,19] Adjuvant therapy is given after the definitive treatment which is usually surgery to lower the risk of recurrence, whereas the neoadjuvant therapy is the administration of therapeutic agents before a definitive treatment. The need for adjuvant radiotherapy and/or chemotherapy is still unclear and is usually indicated in high-grade tumors as these tumors may present sub-clinical or microscopic metastases at the time of diagnosis. The literature review suggests a high observed response rate to chemotherapy, particularly to the anthracycline and alkylating agent–free regimen of vincristine and dactinomycin.[20] One of the major advantage of neoadjuvant therapy in contrast to adjuvant therapy is that it is given before the definitive treatment in order to reduce the size of a tumor. Therefore, the present case was treated with neoadjuvant chemotherapy followed by the surgical resection of the lesion.

CONCLUSION

The rare and variable presentation of fibrosarcoma suggests a crucial role of the pediatric dentist in its diagnosis. Thus, it should be considered as a differential diagnosis among soft tissue masses related to head and neck region, in infants and children. The difficulties in achieving a correct diagnosis for such a rare tumor emphasizes on a multidisciplinary approach by pathologists, radiotherapists and pediatric dentists as well.

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