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Fibrosarcoma of the Oral Cavity: A Case Report

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ABSTRACT

Fibrosarcomas are relatively uncommon malignant tumors arising from fibrous connective tissue and are characterized by increased abnormal proliferation of malignant fibroblast. Fibrosarcoma is capable of local recurrence and metastasis with poor prognosis. Approximately 0.05% of cases are noted in the head and neck region amongst all the cases recorded in the human race. In the present case, we report a case of fibrosarcoma of the mandible in 24 years old female who was presented with small intraoral swelling in the lower anterior vestibule with the involvement of floor of the mouth. Histopathology revealed a proliferation of malignant fibroblast cells arranged in a classical herringbone pattern.

Keywords: Fibrosarcoma, Fibroblasts, Malignancy, Mandible, Spindle cell tumor, Oral cavity

INTRODUCTION

Fibroblastoma is a tumor of mesenchymal cells that are composed of malignant fibroblast cells. Fibrosarcoma is rare in oral and oropharyngeal region, it mainly affects long bones [1]. It can occur at any age, but the peak age of occurrence is the 3rd and 6th decade of life. The etiology of fibrosarcoma remains obscure. Although radiation exposure has thought to be the most important etiological factor, followed by trauma, and underlying conditions of bone-like Paget's disease, fibrous dysplasia, or chronic osteomyelitis [2,3]. Current research shows that the majority of sarcomas are linked with genetic mutations. Recently, a unique fusion transcript has been detected in 10 out of 11 cases. This fusion results from translocation t(12;15) (p13;q25) giving rise to ETV6-NTRK3 (ETS variant gene 6; neurotrophic tyrosine kinase receptor 3) gene fusion. It is absent in other spindle cell tumors of childhood as well as adult fibrosarcoma [3,4]. Fibrosarcoma can occur as soft tissue mass or as an intraosseous tumor. In the initial phase of the tumor, it is mostly asymptomatic and appears like a benign fibrous growth. But in advanced phase, the tumor begins growing very fast and exhibit firm lobulated mass with a smooth surface but surface ulceration, pain, and secondary infections are also often developed. Intraosseous tumor of fibrosarcoma shows severe bone destruction with loosening or exfoliation of adjacent teeth. Two types of fibrosarcoma exist primary and secondary. Primary fibrosarcoma is a fibroblastic malignancy that produces a variable amount of collagen. Secondary fibrosarcoma of bone arises from a pre-existing lesion or after radiotherapy to an area of bone or soft tissue [4,5]. In this paper, we are presenting a case of primary fibrosarcoma of the oral cavity.

Case Report

A 24-year old female patient came with a chief complain of pain in the lower region of the jaw for 3 months. She had visited a private dentist for the pain but obtained no relief of symptoms as the patient was given antibiotics. After 2-3 weeks of the pain experience, she noticed a small growth in the lower anterior vestibule with the involvement of floor of the mouth. The growth was initially very small then slowly enlarged to present size (6 × 7 cm). She has also complained about the difficulty in chewing and speech. Patients dental, medical and family history was non-contributory, also there was no history of any trauma to the teeth. On intraoral examination large swelling, approximately 6 × 7 cm in size was noticed on the floor of the mouth extending from #32 to #42 regions (Figure 1).

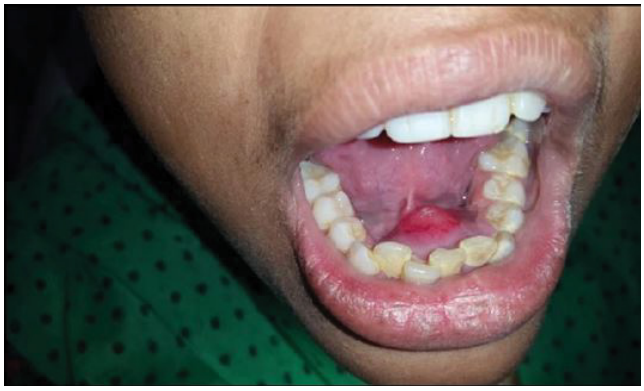

Figure 1 Intraoral picture showing swelling on the floor of the mouth

On palpation, the swelling was firm in consistency and exhibited fixity to underlying tissues. The overlying surface of the swelling was reddish in color and slightly ulcerated. Teeth associated with swelling were vital and non-mobile. Submental lymph nodes were palpable, movable and non-tender. Based on the clinical presentation provisional diagnosis of central giant cell granuloma was made. Radiological examinations revealed osteolytic areas with ill-defined borders on anterior mandible extending from distal surface of #35 to the distal surface of #44 (Figure 2).

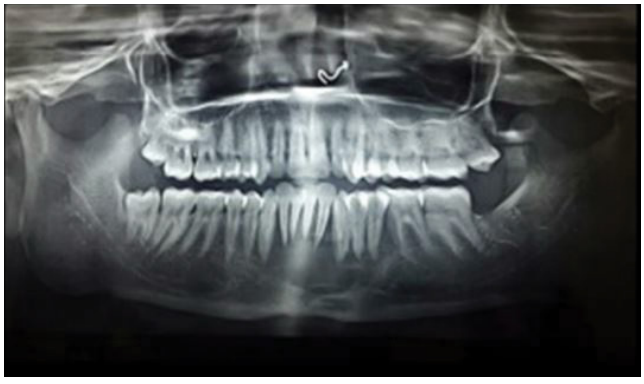

Figure 2 Orthopantomograph (OPG) showing osteolytic areas with ill-defined borders on mandible extending from distal surface of 35 to the distal surface of 44

Internal structure was radiolucent and displacement of teeth with #31, #32, #33, #41, #42 and #43 were seen. Incisional biopsy was performed. On microscopic examination, histopathological section revealed a proliferation of malignant cells with bundles and fascicles of spindle cells arranged in a classical herringbone pattern (Figure 3).

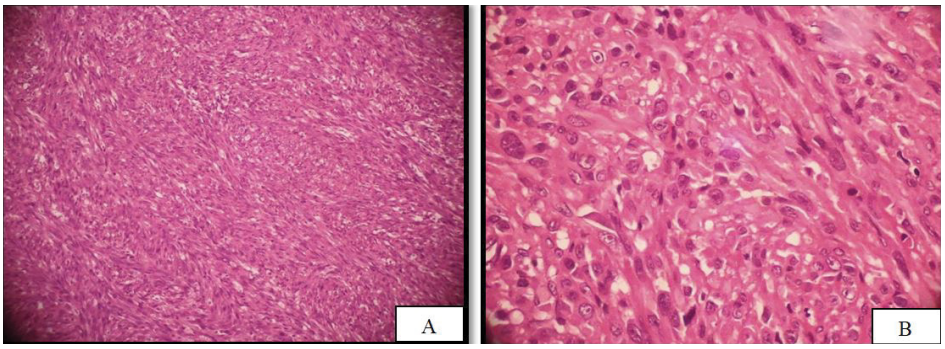

Figure 3 Hematoxylin and eosin-stained section Magnification 10x (a) and 40x (b), showing predominant cellular stroma with fascicles of spindle-shaped cells exhibiting hyperchromatic nuclei, pleomorphic cells, atypical mitotic figures and increased nuclear-cytoplasmic ratio with the presence of herringbone pattern

Cellular and nuclear pleomorphism with hyperchromatic nuclei was noted at places. Increased mitotic activity with numerous bizarre cells was also noted. Focal areas of necrosis and acute inflammatory infiltrate were seen. Few multinucleated tumor-associated giant cells were also seen at periphery. Based on clinical, radiographic and histopathological examination, a final diagnosis of low-grade fibrosarcoma was made. After confirming the diagnosis of fibrosarcoma patient was referred to the oncology center for further evaluation and management. Surgical resection of the fibrosarcoma was done. The patient was kept under regular follow-up (Figure 4).

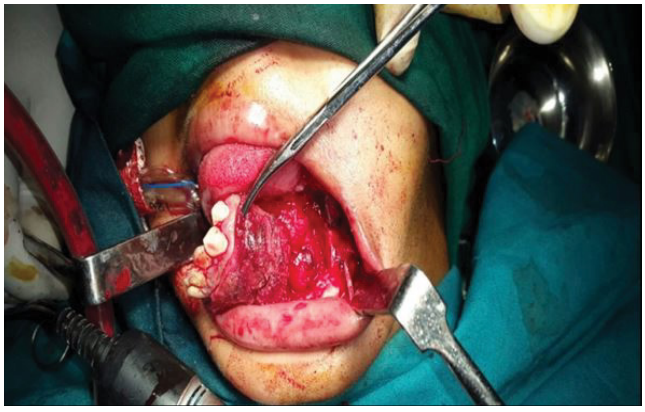

Figure 4 Intraoperative photograph showing surgical resection

DISCUSSION AND CONCLUSION

Fibrosarcoma (FS) is malignant mesenchymal neoplasm affecting long bones of extremity and soft tissues that rarely involves the oral cavity. World Health Organization (WHO) 2002 defined fibrosarcoma as a malignant tumor, composed of fibroblasts with variable collagen and in classical cases, a herringbone architecture [6]. Fibrosarcoma can arise as a primary tumor in jaws and may be classified as a central or peripheral type of fibrosarcoma. Fibrosarcoma of oral cavity clinically exhibits painless, lobulated sessile, non-hemorrhage mucosal mass and sometimes loosening of teeth, pain, and ulceration of overlying mucosa [4]. Although primary fibrosarcoma of jaws is usually asymptomatic, in the present case-patient was symptomatic, had pain swelling and ulceration of overlying mucosa. Clinically in oral cavity fibrosarcoma may be misdiagnosed with ossifying fibroma, squamous cell carcinoma, soft tissue sarcoma, lymphomas, or ulcerated granuloma. Radiographically, it appears as an osteolytic lesion with irregular margins as seen in the present case. However radiographically fibrosarcoma of jaws may not be distinguished from other osteolytic lesions of the bone [7,8]. Microscopically there is a proliferation of spindle-shaped malignant fibroblasts with a variable amount of collagen formation in connective tissue stroma [9]. Histopathologically they are graded into 3 forms, well differentiated, intermediate grade and high grade according to the degree of cellularity, degree of cellular differentiation, mitotic activity, amount of collagen produced by tumor and presence of necrosis [4]. Characteristically, malignant cells of fibrosarcoma are uniform, spindle-shaped with a large, elongated, hyperchromatic nucleus arranged in a typical herringbone pattern. Herringbone pattern shows the diagnostic parallel sheets of cells

arranged in a streaming fashion or interlocking bands that goes in different directions. In many cases, the tumor shows abnormal mitotic figures and cellular atypia. Rarely multinucleated giant cells are also seen. Here in the present case, the malignant cells displayed pleomorphism with hyperchromatic nuclei arranged in a classical herringbone pattern with cellular atypia. Few multinucleated tumor-associated giant cells at the periphery and focal area of necrosis were also noted in this case. Usually, the final diagnosis of fibrosarcoma can be made by careful histopathological examination as we have done in our case. But sometimes it is challenging to differentiate fibrosarcoma from other spindle-shaped sarcomas like fibroblastic osteosarcoma, leiomyosarcomas, lymphoma, liposarcoma, fibrous histiocytoma, and diagnosis often achieved by exclusion. Immunohistochemical analysis of fibrosarcoma is required in cases with diagnostic difficulties. Fibrosarcoma stains positive for intermediate filament vimentin and negative immunostaining for muscular markers (smooth muscle actin, HHF-35, and desmin) will be helpful in establishing the diagnosis of fibrosarcoma [10]. The treatment of fibrosarcoma is radical surgical excision with a wide margin. The role of adjuvant radiotherapy and chemotherapy is still controversial but is generally indicated in high-grade tumors. As in the present case, surgical resection was performed. The prognosis is influenced by the site of origin of the tumor and histopathological grades. Primary fibrosarcoma of bone has a worse prognosis than other bone sarcomas; with 5-year survival rate is around 60% [4]. The prognosis of fibrosarcoma depends upon the clinical stage, histological grades, and local recurrence. The local recurrence for low-grade fibrosarcoma is high as compared with high-grade fibrosarcoma [4]. Therefore it is noteworthy that wide surgical resection is very important to suppress the risk of local recurrence. So that we can conclude, this rare tumor of the oral cavity must be differentiated from a similar form of another lesion in the oral cavity. A dentist must be well aware of the clinical presentation, histological features and immunohistochemical markers of fibrosarcoma to propose an appropriate diagnosis and aid in treatment planning.

DECLARATIONS

Conflict of Interest

The authors declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

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