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Fibrosarcoma of mandible: A case report

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ABSTRACT

Fibrosarcomas is an uncommon connective tissue growth that rises from the proliferation of malignant fibroblasts. Local recurrence is frequent, but metastasis is rare. About 0.05% of cases are affected in the head and neck. We report a case of mandibular fibrosarcoma in a 30-year-old man who presented with intraoral swelling in the ridge of the left mandibular alveolus. Histopathology showed the proliferation of malignant fibroblast cells arranged in a classic herringbone pattern.

Keywords: Fibrosarcoma; Fibroblast; Malignancy; Lower jaw; Spindle cell tumor; Oral cavity.

Introduction

Fibroblastoma is a mesenchymal cell tumor of malignant fibroblast. Fibrosarcoma is rare in the oropharynx, mainly affecting the long bones [1]. It can happen at any age, but the peak age of occurrence is in the third and sixth decades of life. The etiology of fibrosarcoma is unclear. Although radiation exposure is considered as the most important etiological factor, followed by trauma and underlying conditions such as Paget's disease, fibrodysplasia, or chronic osteomyelitis [2,3]. Current research suggests that the majority of sarcomas are associated with genetic mutations. Recently, a unique fusion transcript was identified in 10 of 11 cases. Cases of this fusion are caused by translocation t (12;15) (p13;q25), which causes ETV6-NTRK3 gene fusion (ETS type 6 gene) neurotrophic receptor tyrosine kinase 3). Fibrosarcoma can be a soft tissue mass or an intraosseous tumor. In the initial phase of the tumor, it is mostly asymptomatic and looks like a benign fibrotic growth. But in the advanced phase, its growth is fast and a hard lobular mass with a smooth surface sometimes with superficial wound, pain and secondary infections are also often created. intraosseous Fibrosarcoma tumor shows severe bone destruction with loosening or exfoliation of adjacent teeth. There are two forms of fibrosarcoma, primary and secondary. Primary fibrosarcoma is a fibroblastic malignancy that produces different types of collagen. Secondary fibrosarcoma origi-

nates from bone and arises from a previous lesion or after radiotherapy in an area of bone or soft tissue [4,5]. In the oral region, the 5-year survival rate for oral soft tissue fibrosarcoma is 69%. 27% for those of medullary origin [6]. In this article, we present a case of primary fibrosarcoma of the oral cavity.

Case Presentation

A 31-year-old male patient presented with the main complaint of pain in the lower jaw for 2 months. About 3 years ago, the left first molar of the lower jaw was removed due to caries up to the furcation area. He had visited the dentist for pain, but even though the patient was given antibiotics, the symptoms did not get relieved. After 2/5-3/5 weeks of pain, he noticed a growth in the area of the alveolar ridge of the extracted 6th left mandibular tooth, which was very small at first, then slowly grew to its current size (3 x 2 x 1.5). No such case has been reported in the patient's dental, medical and family history. He also had a history of falling from a height about 4 years ago. In the intraoral examination, a red saddle-shaped exophytic lesion with a smooth surface of approximate dimensions (3cm x 2cm x 1.5cm) extended from the lingual of the 6th lower left tooth to the depth of the vestibule. Telangiectasia was observed on the surface of the lesion and diascopy was positive. The lesion's consistency is rubbery and extended in the vestibule's depth to the alignment of teeth 5 and 7. There is a prolapse of the lower 7th tooth.

Based on the clinical manifestations, a provisional diagnosis of central giant cell granuloma was given. Radiological examinations showed osteolytic areas with an indistinct border behind the left mandible, which extended from distal tooth 5 to distal tooth 7. The internal structure was radiolucent and an incisional biopsy was performed. On microscopic examination, the histopathological section showed the proliferation of malignant cells with bundles and fascicles of spindle cells arranged in a classic herringbone pattern. Cellular and nuclear pleomorphism with hyperchromatic nuclei was observed in some places. Increased mitotic activity, was also noticed with many strangely shaped cells. A small number of multi-nucleated tumor giant cells were also observed in the periphery. Based on the clinical appearance, radiography and Histopathological examination, the final diagnosis of low-grade fibrosarcoma was given. After confirmation of the diagnosis, the patient was referred to the oncology center for further evaluation and management.

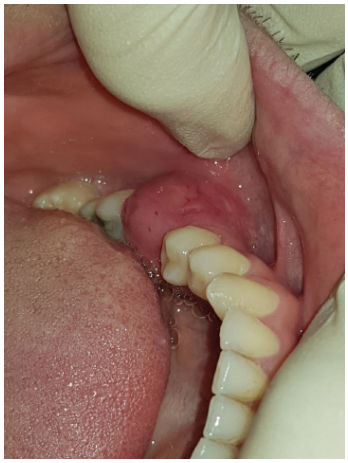
Discussion

Fibrosarcoma (FS) is an uncommon malignant mesenchymal neoplasm that accounts for approximately 5% of all intraosseous malignant tumors. It affects long bones and soft tissues. About 10% occur in the neck. More than 75% of the cases are reported in the lower jaw [7]. The World Health Organization (WHO) defined fibrosarcoma in 2002 as a malignant tumor composed of fibroblasts with variable collagen and, in the classic form, a herringbone structure [8]. Fibrosarcoma can develop as a primary tumor in the jaw and is classified as a central or peripheral type of fibrosarcoma. Fibrosarcoma of the oral cavity is clinically a painless, lobulated, and non-bleeding mucous mass, and sometimes loose teeth, pain, and ulceration of the covering mucosa are seen along with it [4]. Although primary fibrosarcoma of the jaw is usually asymptomatic, in the present case, the patient was symptomatic, with painful swelling and mucosal ulceration.

Clinically, fibrosarcoma of the oral cavity may be differentially diagnosed with ossific fibroma, squamous cell carcinoma, soft tissue sarcoma, lymphoma, or ulcerative granuloma. Radiographically, as seen in the present case, it is in the form of an osteolytic lesion with irregular margins. However, radiographically, fibrosarcoma of the jaw may not be distinguished from other osteolytic bone lesions [9,10]. Microscopically, the Proliferation of malignant fibroblasts is spindle-shaped with a variable amount of collagenization in the connective tissue stroma [11]. From the histopathological point of view, they are graded in 3 forms: fully differentiated, medium grade, and high grade, based on mitotic activity, the amount of collagen produced by the tumor, and the presence of necrosis [4].

Characteristically, malignant fibrosarcoma cells are uniform, spindle-shaped with a large, elongated, hyperchromatic nucleus arranged in a typical herringbone pattern. The herringbone pattern shows the characteristic parallel sheets of cells that form interlocking bands. The threads that are placed in different directions are arranged. In many cases, the tumor shows itself in the form of abnormal mitosis and cell atypia. Rarely, multinucleated giant cells are also seen. in the present case, the malignant cells showed pleomorphism with hyperchromatic nuclei arranged in a classic herringbone pattern with cellular atypia. A small number of giant cells associated with multinucleated tumor in the periphery and focal area of necrosis were also noted in this case. Usually, the final diagnosis of fibrosarcoma is made by detailed histo-

pathological examination as in the present case. But sometimes it is challenging to distinguish fibrosarcoma from other spindle-shaped sarcomas such as fibroblastic osteosarcoma, leiomyosarcoma, lymphoma, liposarcoma, fibrosing histiocytoma, and the diagnosis is often rejected. Immunohistochemical analysis of fibrosarcoma is required in cases where there are diagnostic difficulties. Fibrosarcoma is positive for intermediate filament vimentin, and a negative result in immunostaining for muscle markers (smooth muscle actin, HHF-35, and desmin) will be useful in the diagnosis of fibrosarcoma [12]. Secondary fibrosarcoma of bone may be associated with fibro dysplasia, Paget's disease, it may also appear as a malignant giant cell tumor of the bone or caused by previous radiation [7] and may be misdiagnosed with an abscess or an odontogenic cyst of the root of the adjacent teeth [11]. The treatment of fibrosarcoma is radical surgical resection with wide margins. The role of adjuvant radiation therapy and chemotherapy is still uncertain, but it is generally used in high-grade tumors. As in the present case, surgical excision was performed. Prognosis is influenced by tumor location and histopathological grade. Primary fibrosarcoma of bone has a worse prognosis than other bone sarcomas. The prognosis of fibrosarcoma depends on clinical stage, histological grade and local recurrence. Local recurrence of fibrosarcoma with low differentiation grade is high compared to fibrosarcoma with high differentiation grade [4]. Therefore, wide surgical resection is crucial to suppress the risk of local recurrence. Therefore, we can conclude that this rare oral cavity tumor should be differentiated from other similar lesions in the oral cavity. The dentist should know well about the clinical presentation, histologic features, and immunohistochemical markers of fibrosarcoma to suggest an appropriate diagnosis and assist in treatment planning.



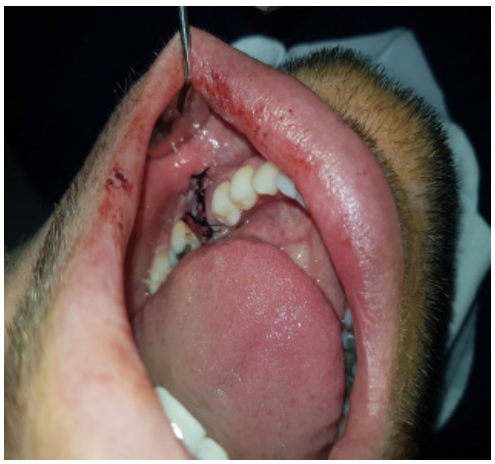
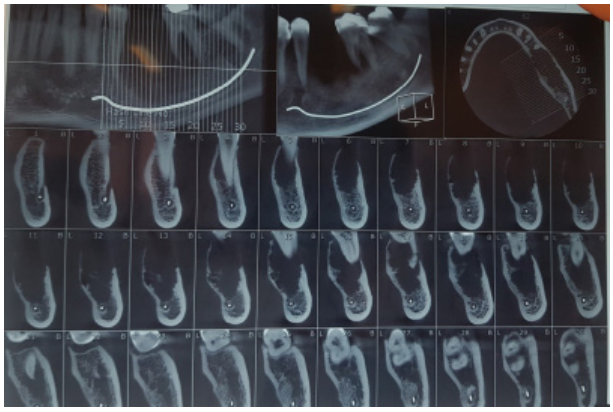


Figure 1 & 2. Intraoral manifestation of fibrosarcoma before and after biopsy.



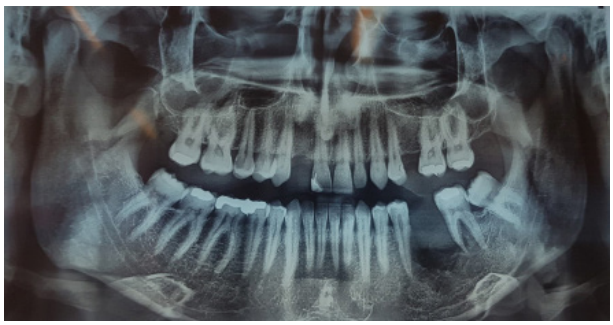


Figure 3 & 4. Radiographically, in the present case, it is in the form of an osteolytic lesion with irregular margins.

Acknowledgments

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Conflict of Interest

There is no conflict of interest to declare.

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