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Fibrosarcoma of the maxilla with maxillary sinus invasion: a case report and review of the literature

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Abstract

Background Fibrosarcoma is a rare, invasive soft tissue sarcoma that is more common in males aged 30–60 years. It accounts for 1% of all malignancies in the human body. Treatment is radical surgical removal, radiotherapy, and chemotherapy.

Case presentation A 58-year-old Khorasani Persian male patient with a complaint of expansion, pain, and paresthesia in the left maxillary quadrant was diagnosed with infiltrative high-grade sarcoma. Tumoral cells were strongly diffuse positive for vimentin and negative for S100 and desmin, indicating high-grade adult fibrosarcoma.

Conclusion This report presents a rare case of a fibrosarcoma causing paresthesia, pain, and expansion in the maxilla with maxillary sinus invasion, which requires early diagnosis and immediate referral.

Keywords Fibrosarcoma, Maxilla, Oral cavity, Immunohistochemistry, Case report

Introduction

According to the World Health Organization, fibrosarcoma (FS) is classified as a malignant spindle cell tumor characterized by fibroblastic/myofibroblastic differentiation and fascicular architecture, along with variable collagen matrix production [1]. Adult fibrosarcoma is an exceptionally rare and invasive type of soft tissue sarcoma. It is often difficult to distinguish fibrosarcoma from other soft tissue sarcomas and is only diagnosed after all other types of mesenchymal and non-mesenchymal forms are ruled out by a thorough study of their morphological characteristics and further assessment [2].

Adult fibrosarcoma is more common in individuals aged 30–60 years, and its occurrence is greater in males [3, 4].

Fibrosarcoma accounts for 1% of all malignancies in the human body, and it is more common in soft tissue. Its intraosseous occurrence has been reported to be approximately 5%. Only 0.05% of cases occur in the head and neck, and 10% of these cases occur in the maxillofacial region and oral cavity. Fibrosarcoma is also very rare in the maxilla, with a prevalence of 0–6.1% [5–8].

Fibrosarcomas often manifest clinically as slowly developing tumors that may enlarge significantly prior to causing discomfort. Histologically, the composition of well-differentiated fibrosarcomas mostly comprises fascicles of spindle-shaped cells, which are known to have a characteristic arrangement resembling a "herringbone pattern." Cellular morphology typically exhibits minimal variation in both size and form; however, there is typical variability in the number of mitotic figures that may be observed. The cells in poorly differentiated tumors are less often arranged and may have an oval or spherical

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shape. The presence of mild pleomorphism, accompanied by an increased frequency of mitotic activity, may be observed. The histological characteristics of high-grade fibrosarcoma may be similar to those of other tumor types, including malignant fibrous histiocytoma, liposarcoma, or synovial sarcoma [9].

The treatment of choice is radical surgical removal in combination with radiotherapy and chemotherapy [10].

This report presents a rare case of a fibrosarcoma in the maxillary left quadrant causing pain and paresthesia.

Case presentation

A 58-year-old Khorasani Persian male patient with a history of smoking for more than 20 years was referred to the Tehran School of Dentistry with a chief complaint of expansion, pain, and paresthesia in the left maxillary quadrant for 2 months. His medical history and family history were unremarkable.

Radiographic evaluation was performed with the aid of cone beam computed tomography, revealing a 40 mm × 33 mm radiolucent soft-tissue shadow destructive mass with ill-defined borders on the left side of the maxilla evading the maxillary sinus floor (Fig. 1).

An incisional biopsy of the lesion was performed through an intraoral approach. On microscopic examination, large epithelioid to spindle and occasionally histiocytic cell proliferation with significant atypia in extensive sheets and nests with irregular crisscrossing fascicles and storiform patterns were observed. Numerous atypical mitotic figures and focal areas of necrosis were also present. The hyperplastic parakeratotic stratified squamous epithelium of the mucosa was partially invaded by tumoral cells, and ulceration was also observed. Sheets of hemorrhage and superficial bacterial colonies were evident. The microscopic findings led to the diagnosis of infiltrative high-grade sarcoma (Fig. 2). Further immunohistochemistry analysis of tumor cells revealed strong diffuse positivity for vimentin, and the Ki-67 proliferation index was estimated to be 40–50%, with a mitotic rate of 7/10 in a high-power field. Smooth muscle actin (SMA)

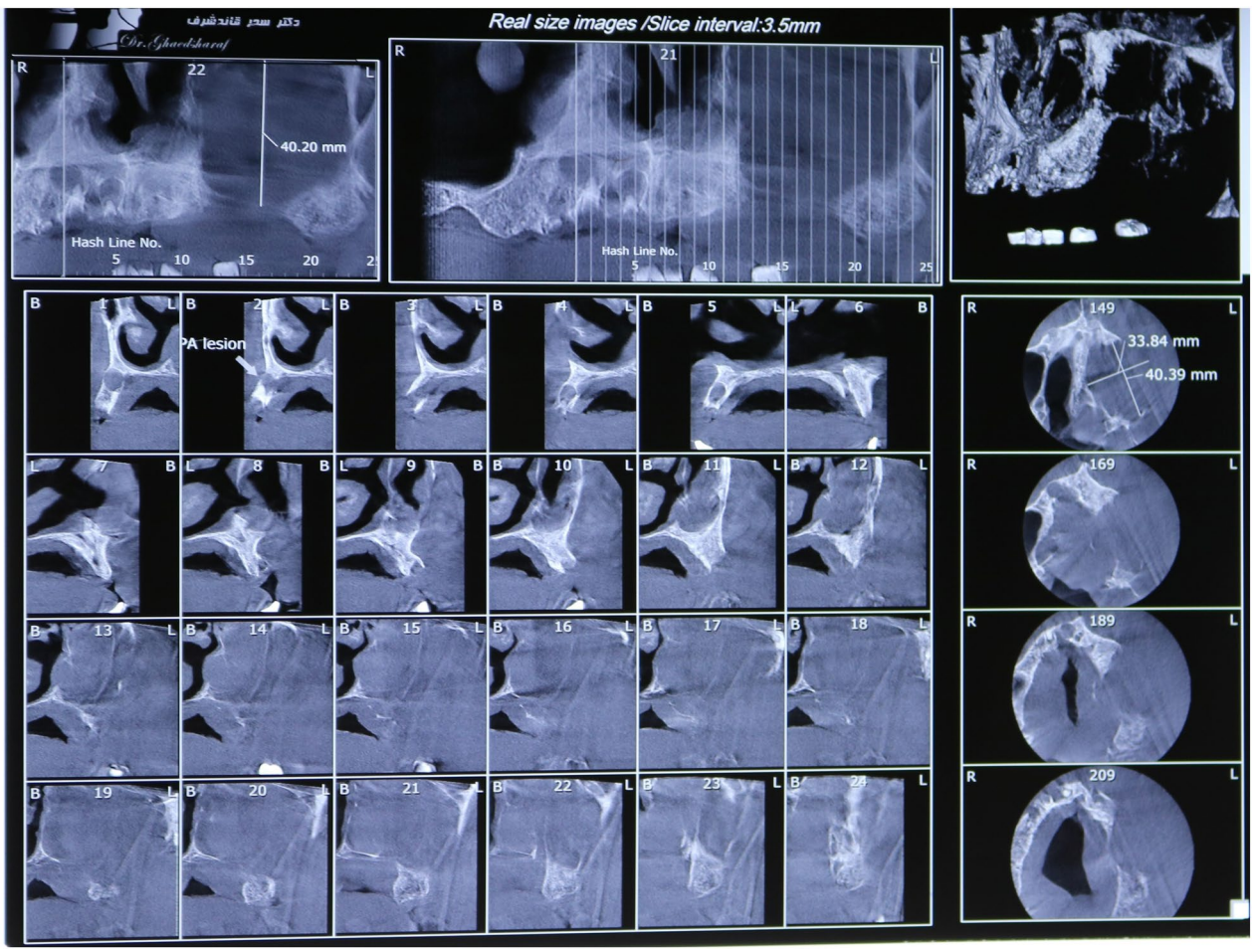

Fig. 1 Radiographic evaluation and cone-beam computed tomography showing a diffuse, ill-defined radiolucent mass in the left maxillary area

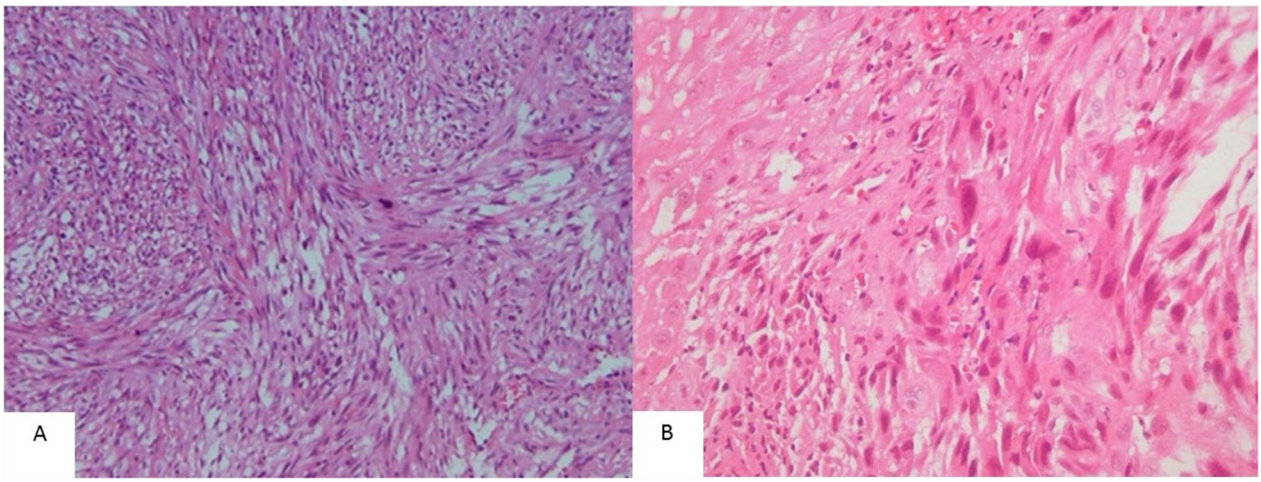

Fig. 2 Hematoxylin–eosin-stained sections. Interlacing fascicles of spindle cells with occasional nuclear polymorphisms (**A**; 200×). Higher magnification shows spindle cells with nuclear atypia and hyperchromatism (**B**; 400×)

was positive only in the stromal blood vessels, and Pan-CK was positive in the superficial mucosa. Tumoral cells were negative for S100, desmin, EMA, CD99, Bcl-2, and H-caldesmon (Fig. 3) Thus, a diagnosis of adult fibrosarcoma of the maxilla (high-grade) was given, and the patient was referred for surgical excision.

The patient was referred to an oncology center for further evaluation and management. Because of the extent of the lesion, surgery was not carried out, and chemotherapy and radiotherapy were started. The chemotherapy regimen consisted of doxorubicin 20 mg/m^2 intravenously, ifosfamide 2000 mg/m^2 intravenously, and Mesna 225 mg/m^2 intravenously for 3 days and was repeated every 21 days. The dosage for radiotherapy was 50 Gy. The lesion had not yet been resected, and the patient was not followed up.

Discussion

Fibrosarcoma is a malignant tumor of fibroblasts with variable collagen synthesis and "herring bone" architecture. Today, the diagnosis is made through ruling out other mesenchymal and non-mesenchymal tumors. There are two types of fibrosarcoma: infantile or congenital and adult-type fibrosarcoma. The adult type is highly malignant, whereas congenital fibrosarcoma rarely metastasizes [2, 9, 10].

Fibrosarcoma commonly arises from tendons and fascia located within deep soft tissue. The tumor can manifest as either a primary or secondary neoplasm. Primary fibrosarcoma of the bone has the potential to originate from either the medullary canal or the periosteum. Secondary fibrosarcoma might be associated with a range of conditions, including but not limited to preexisting bone lesions, bone injury, radiation-induced fibrous dysplasia, Paget's disease, and osteomyelitis [7, 11]. In our case, no definite etiological factor was identified.

Fibrosarcoma is a rare neoplasm, approximately 0.05% of which occurs in the head and neck. Approximately 10% of these involve the maxillofacial region, and the maxilla accounts for 0–6.1% of all primary fibrosarcomas of the bone [12, 13]. Males are more likely to develop adult fibrosarcoma, which is more common in those between the ages of 30 and 60 years [2, 10].

Patients are frequently asymptomatic, but occasionally, the main symptoms include jaw expansion, either accompanied by pain or without it, paresthesia, and loosening of teeth. Furthermore, in large masses, ulceration may occur [13]. Pain and paresthesia commonly manifest as late signs, indicating the involvement of nerves [8, 10, 14]. In the mentioned case, swelling, pain, and paresthesia were present.

Radiographic imaging reveals radiolucent lesions with ill-defined osteolysis and moth-eaten patterns of bone destruction [15, 16]. Our patient presented with ill-defined borders, thinning, and disruption of the cortex, maxillary sinus, and inferior cortical bone.

Diverse fascicles of spindle cells with varying collagen production comprise the histology of fibrosarcoma. Fibrosarcoma grading is determined by a number of criteria, including the amount of collagen matrix produced by tumor cells, cellularity, pleomorphism, cellular differentiation, level of necrosis, and mitotic activity [17].

Tumors classified as grade I (well-differentiated) have a homogeneous nuclear appearance and a noticeable amount of collagenous intercellular material. Giant cells cannot be observed, and mitotic figures are either scarce or nonexistent. Tumors classified as grade II (intermediate) have less intercellular material and more cellularity.

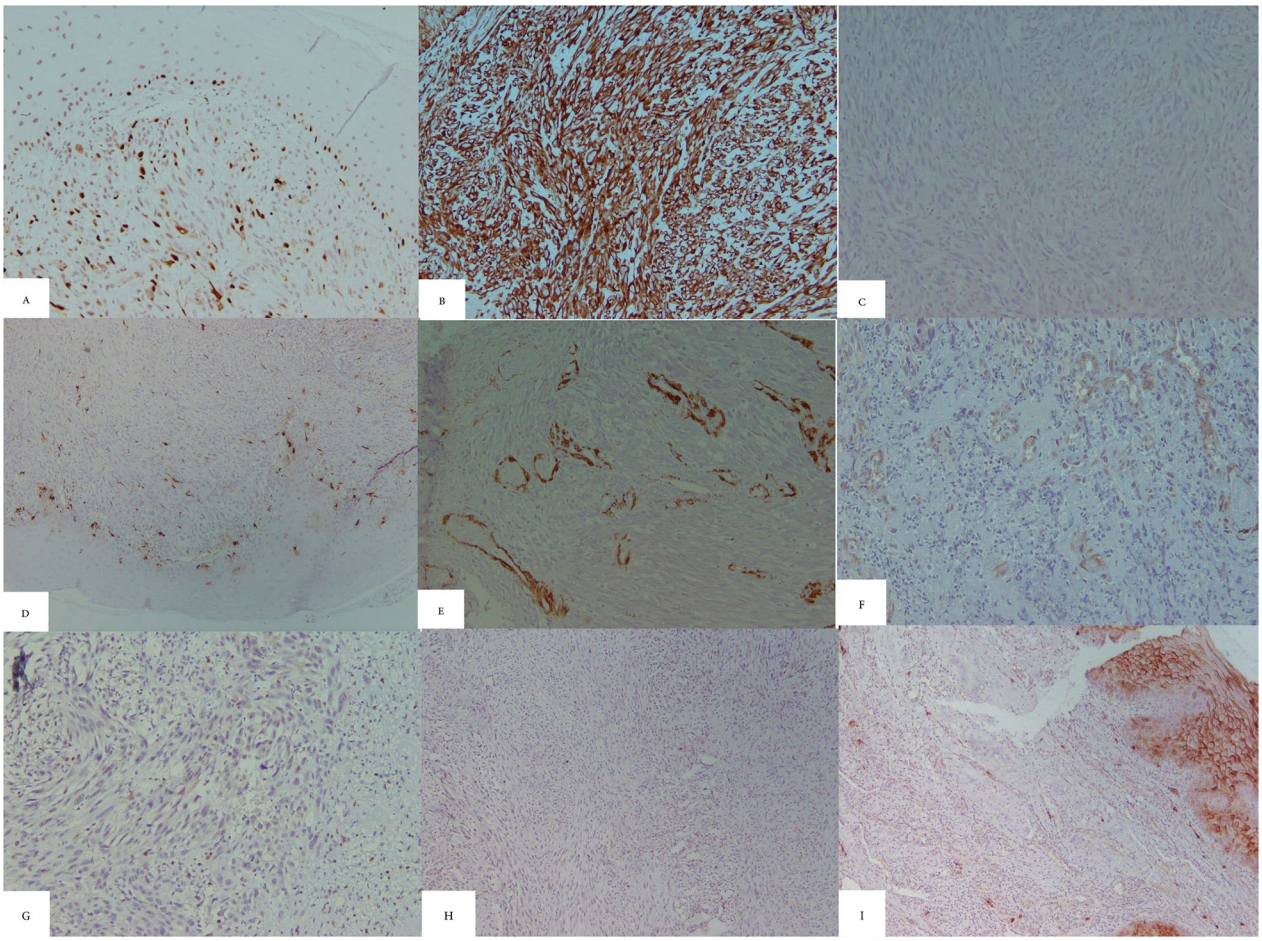

Fig. 3 Immunohistochemical stained sections. Expression of Ki-67 shows high proliferative activity in the tumor cells. Positive immunoreactivity in the basal cells of the epithelium served as an internal control (**A**; 100×), diffuse strong immunoreactivity for vimentin (**B**; 200×), and negative expression for desmin (**C**;100×). Immunohistochemical staining for S100 revealed few scattered positive cells (**D**; 100×), negative immunoexpression of SMA (**E**; 200×), H-caldesmon (**F**; 200×), Bcl-2 (**G**; 200×), CD99 (**H**; 100×), and EMA (**I**; 100×)

Even so, the nuclei exhibit the characteristic herring bone pattern and are somewhat homogeneous in size, shape, and staining characteristics while being more densely packed. Mitotic figures are occasionally observed. Anaplastic tumors with several large cells and mitotic figures are known as grade III (high) tumors. The prognosis of a tumor is directly correlated with its histological grade, which is highly important [18, 19].

Fibrosarcoma is primarily diagnosed by ruling out other spindle cell mesenchymal tumors. It is characterized by the absence of immunocytochemistry (IHC) stains for epithelial, myogenous, and neural markers, as well as for CD34, CD99, Bcl-2, and nuclear beta-catenin [20–22]. The histological morphology of a fibrosarcoma closely resembles that of fibromatosis, fibroblastic odontogenic sarcoma, malignant fibrous histocytoma (MFH), malignant peripheral nerve sheath tumor (MPNST), liposarcoma, and synovial sarcoma. Fibromatosis is characterized by the absence of mitosis and a very low level of cellular atypia. Odontogenic sarcomas exhibit odontogenic tissues that are not observed in fibrosarcoma. An immunohistochemical (IHC) study was conducted to differentiate between MFH, liposarcoma, synovial sarcoma, MPNST, and fibrosarcoma. This study focused on the markers vimentin, pan-cytokeratin, desmin, actin, EMA, H-caldesmon, CD99, Bcl-2 and S100 protein. Immunohistochemistry revealed the presence of vimentin-positive cells, while the other immunomarkers were negative. Fibrosarcoma is mostly diagnosed by ruling out other possibilities, and it is characterized by the absence of actin, cytokeratin, S100, and desmin expression [23, 24].

Vimentin is the most potent positive stain for fibrosarcoma. There are no markers for neurological tissue, such as S100 and neuron-specific enolase; macrophages (CD68); melanoma (HMB-40); muscles, such as desmin

and actin; or epithelial tissue, such as cytokeratin and epithelial membrane antigen [13]. In the present case, the tumor cells were strongly diffuse positive for vimentin, the Ki-67 proliferation index was estimated at 40–50%, and these cells were negative for other markers.

There have been few reports of fibrosarcoma in the maxilla. Khanna *et al*. in 2014 reported a case of fibrosarcoma in the maxilla with extension into the maxillary sinus. The lesion caused discomfort in speech and mastication and revealed diffuse swelling [25]. Similar to our patient, Yuwanati *et al*. reported a patient with swelling, paresthesia, and pain in 2011 [26]. In 2023, Hrishi *et al*. reported a case of fibrosarcoma in a patient with an enlarged gingiva in the lower front region [6].

The published cases identified as fibrosarcoma of the jaws are listed in Table 1. The prevalence of fibrosarcoma was more evident in males between the ages of 30 and 60 years. The most common symptom was pain and swelling, and resective surgery with or without chemotherapy was the most frequently used treatment.

The diagnosis of the lesion is based on histopathological analysis. The patient's chances of survival increase with an early diagnosis of any sarcoma. Fibrosarcomas are aggressive lesions, with high-grade tumors accounting for 80% of lesions. The preferred course of treatment for sarcomas is surgical excision. Clear margins and wide excision are necessary for patient survival. Radiation and surgery together are known to decrease recurrence [6].

Conclusion

Fibrosarcoma is a rare neoplasm, with approximately 0.05% of cases occurring in the head and neck. It can manifest as a primary or secondary neoplasm and can be associated with various conditions. The tumor is characterized by ill-defined osteolysis and moth-eaten bone destruction. The prognosis is directly correlated with the histological grade. The most potent positive stain for fibrosarcoma is vimentin. Early diagnosis increases the patient's chances of survival. The preferred treatment is surgical excision, with clear margins and a wide excision required for patient survival.

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Author contributions
NM and ME drafted the manuscript. NM, ME, and SD performed the experiments.

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Availability of data and materials
Data sharing is not applicable to this article as no datasets were generated or analyzed during the current study.

Table 1 Patients identified in English-language literature as having fibrosarcoma of the jaws; only those with demographic features are listed below

Author	Year	Sex/age	Location	Signs and symptoms	Treatment	Recurrence	Follow-up
Wadhwan *et al*. [27]	2010	Male/38	Posterior mandible	Pain and slow-growing swelling	CT	NA	NA
Wadhwan *et al*. [27]	2010	Male/55	Posterior maxilla	Pain and rapid-growing swelling	CT	Patient succumbed to the lesion	1 year
Yuwanati *et al*. [26]	2011	Female/44	Posterior mandible	Pain and swelling and paresthesia	NA	NA	NA
Dhanavelu *et al*. [5]	2012	Female/73	Posterior maxilla	Pain and swelling	NA	NA	NA
Swain *et al*. [7]	2013	Male/8	Anterior maxilla	Rapidly growing swelling	Surgery	Patient succumbed to the lesion	6 months
Nanda *et al*. [28]	2013	Female/17	Posterior mandible	Pain and swelling	NA	NA	NA
Khanna *et al*. [25]	2014	Male/63	Posterior maxilla	Rapidly growing swelling	Surgery	NA	NA
Reddy *et al*. [29]	2015	Male/45	Anterior maxilla	Pain and swelling	Surgery + CT	Yes	6 months
Shrivastava *et al*. [13]	2016	Female/22	Anterior maxilla	swelling	Surgery	No	Few months
Sood *et al*. [8]	2016	Male/60	Premolar region of maxilla	Pain and swelling	Surgery	Yes	8 months
Akhtar *et al*. [15]	2016	Female/25	Posterior mandible	Swelling	Surgery + CT	No	2 years
Singh *et al*. [16]	2019	Male/4	Posterior mandible	Rapidly growing swelling	Surgery + CT	No	18 months
Anisuzamn *et al*. [30]	2020	Male/15	Posterior mandible	Pain and swelling	Surgery	No	2 months
Parvathi *et al*. [14]	2022	Female/30	Posterior mandible	Pain and swelling	NA	NA	NA
Hrishi *et al*. [6]	2023	Male/44	Anterior mandible in gingiva	Swelling Bleeding from gingiva pain	Surgery + RT	NA	NA

CT chemotherapy, *N/A* not assessed

Declarations

Ethics approval and consent to participate
Not applicable.

Consent for publication
Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Competing interests
The authors declare that there are no competing of interest regarding the publication of this article.

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