Fibrosarcoma of the oral cavity

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ABSTRACT

Fibrosarcoma has been defined as a malignant tumor of the fibroblasts that shows no other evidence of cellular differentiation and is capable of recurrence and metastasis. Fibrosarcomas are rare but may occur anywhere in the body, most commonly in the retroperitoneum, thigh, knee and distal extremities. Fibrosarcoma is uncommon in the head and neck region and constitutes about 1% of all the malignancies affecting the human race. Of all the fibrosarcomas occurring in humans, only 0.05% occurs in the head and neck region. Of this, almost 23% is seen in the oral cavity. Fibosarcomas generally have a poor prognosis and the overall survival rate is 20–35% over a period of 5 years.

Key words: Fibroblasts, fibrosarcoma, oral cavity

Fibrosarcoma is a malignant mesenchymal neoplasm of the fibroblasts that rarely affects the oral cavity proper. The etiology of fibrosarcoma remains obscure. Although radiation exposure has been 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. Fibrosarcomas may occur intraosseously as well as in the soft tissues. Clinically, the lesion may cause pain, swelling, paresthesia, loosening of the teeth and ulceration of the overlying mucosa.

Histologically, the degree of differentiation is variable, from being comparable to a benign fibroma to a highly anaplastic tumor, thus presenting a diagnostic dilemma to the histopathologists. Fibrosarcoma can be graded as low and high grade of malignancy. Low-grade fibrosarcoma shows spindle cells arranged in fascicles with low to moderate cellularity and a herringbone appearance. There is a mild degree of nuclear pleomorphism and rare mitosis, with a collagenous stroma. High-grade lesions show an intense nuclear pleomorphism, greater cellularity and atypical mitosis. The nuclei can be spindle shaped, oval or round. The histological appearance of high-grade fibrosarcoma may be similar to other tumors such as malignant fibrous histiocytoma, liposarcoma or synovial sarcoma. The positive immunostaining for vimentin, together with negativity for muscular immunomarkers, helps in diagnosing the fibrosarcoma. The treatment of choice is radical surgery; radiation therapy and chemotherapy can be used in inoperable cases. We are hereby presenting two cases of primary fibrosarcoma of the oral cavity reported to the outpatient department of Sharad Pawar Dental College and Hospital, Sawangi (M), Wardha, India.

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CASE REPORTS

Case 1

A 55-year-old male patient came with the complaint of slow-growing swelling over the left side of the face [Figure 1]. The patient did not give any history of systemic illness or trauma to the head and neck region. The patient was a farmer by profession. There was no significant contributing family history. The patient gave a history of tobacco chewing since 38 years. The patient was a resident of the Vidarbha region of central India. He gave a history of pain in the posterior mandibular region since 4 months. After 1 week of the onset of pain, the patient noticed a small swelling in the left buccal mucosa. The patient gave a history of a slowly enlarging swelling, which attained to the present size of 10 cm × 15 cm. There was associated history of difficulty in speech and mastication, loss of appetite with weight loss and fever. Extraorally, the swelling extended anterioposteriorly from the left corner of the mouth to the tragus of the ear and superioinferiorly from the ala-tragus line to 4 cm below the lower border of the mandible. The swelling was irregular in shape and the borders were ill defined. Skin over the swelling was stretched, but was normal in color. The swelling was firm and fixed to the underlying structures. The right submandibular lymph nodes were palpable and the left side nodes could not be palpated due to the size of the swelling. Intraorally, the swelling appeared to be arising from the buccal mucosa and was crossing the midline toward the right side. The buccal

and the lingual cortical plates were expanded, but intact. There was displacement of all teeth on the left side of the mandibular arch. The tongue was displaced toward the right side [Figure 2]. The buccal and the lingual vestibule were obliterated.

The orthopantomograph (OPG) revealed displacement of teeth and superficial erosion of buccal cortical plate. There was displacement of all the teeth and root resorption associated with the posterior teeth on the left side of the mandibular arch [Figure 3]. A clinical diagnosis of osteosarcoma was made.

The intraoral mass was subjected to incisional biopsy. The hematoxylin and eosin (H and E)-stained section showed nonencapsulated tumor tissue made up of rich collagenous stroma. The tumor tissue showed the presence of spindle-shaped cells with a large nucleus and scanty cytoplasm. Few mitotic figures were seen. In few areas, there was presence of Herring Bone pattern characteristic of fibrosarcoma [Figure 4]. A histopathological diagnosis of fibrosarcoma was made and the patient was referred for surgical excision. Because of the extent of the lesion, the surgery was not carried out and chemotherapy was started. After a follow-up of 1 year, the patient succumbed to the lesion.

Case 2

A 38-year-old male patient came with the complaint of pain and rapidly enlarging swelling on the right side of the face. The patient did not give any history of systemic illness or trauma to the head and neck region. The patient was a daily wage labor by profession. There was no significant contributing family history. The patient gave a history of beedi smoking since 24 years. The patient was a resident of the Vidarbha region of central India. The patient noticed the swelling on the right side of the face since 1 month. The swelling started 1 month back when the patient noticed the loosening of teeth in the maxillary posterior region. Extraorally, a single large swelling measuring 7 cm × 8 cm, extending superioinferiorly from the infraorbital margin to the angle of the mouth and anterioposteriorly from the angle of the mouth to the zygomatic arch were seen. The overlying skin was taut and fixed to the underlying swelling. Intraorally, a single growth was seen extending from the corner of mouth toward the maxillary tuberosity region and was extending laterally toward the midline of the palate [Figure 5]. Intraorally, the swelling was ulcerated as the patient gave a history of repeated cheek biting. The teeth on the affected side were displaced toward the palatal side.

The orthopantomograph revealed the displacement of teeth, and bone involvement was seen. Severe bone loss was seen in the mandibular posterior area. A clinical diagnosis of malignancy involving the alveolus was made.

The intraoral mass was subjected to incisional biopsy. The H and E-stained section showed closely packed cells with few collagen fibers. The stroma was entirely composed of bizarre-looking tumor cells that were rounded in shape [Figure 6]. Numerous mitotic figures were seen with few areas of necrosis [Figure 7]. A differential diagnosis of lymphoma, round cell tumor, rhabdomyosarcoma and nasopharyngeal carcinoma was made. The lesion was positive for p53 protein expression [Figure 8]. Immunohistochemically, the cells showed immunoreactivity for vimentin, whereas they showed negativity for S-100 and cytokeratin cocktail. Based on clinical behavior and histological assessment, the final diagnosis of poorly differentiated fibrosarcoma was made. The patient was subjected to whole body scan and no distant metastasis was detected. The lesion was inoperable and chemotherapy was started. The patient has been put on periodic recall.

DISCUSSION

Fibrosarcoma is a malignant tumor that arises from the fibroblasts (cells that produce connective tissue). This is a type of sarcoma that is predominantly found in the area around the bones or in soft tissue. In earlier studies of soft tissue neoplasm, this tumor has been greatly overdiagnosed, and this diagnosis has been frequently applied to virtually any richly cellular, collagen-forming spindle cell tumor, including malignant fibrous histiocytoma, malignant Schwannoma and a host of other sarcomatous and pseudosarcomatous lesions.[1] Malignancies of the fibroblasts are decidedly rare in the oral and oropharyngeal region, but fibrosarcoma is, nevertheless, the most common mesenchymal cancer of the region, representing more than half of all sarcomas. Of all the fibrosarcomas occurring in human, only 0.05% occurs in the head and neck region. Of this, 23% of head and neck fibrosarcomas occur within the oral cavity.[2,3]

The exact cause of fibrosarcoma is not entirely understood; however, studies have indicated that genetic alterations may play a role. A chromosomal rearrangement has been found in some fibrosarcomas. Radiotherapy to the local site has been proposed as a predisposing factor for increased risk of Fibrosarcoma. Other factors that can give rise to fibrosarcoma of the oral and paraoral region include tissues damaged by scarring and heat. Disease processes like Paget's disease and osteomyelitis have also been implicated in few cases in which the fibrosarcomas developed in the bone. Fibrosarcoma elsewhere in the body develops in people between the ages of 25 and 79 years. The peak incidence of occurrence of this tumor is 55–69 years. However, fibrosarcomas in the head and neck region develop in the 3rd and 5th decade of life, but there is a wide age range and many patients are below 20 years of age. Infantile or congenital fibrosarcoma is the most common soft tissue sarcoma found in children under 1 year of age.[2-6] There is no gender predilection. Generally, the tumors develop with equal frequency in males and females. However, in

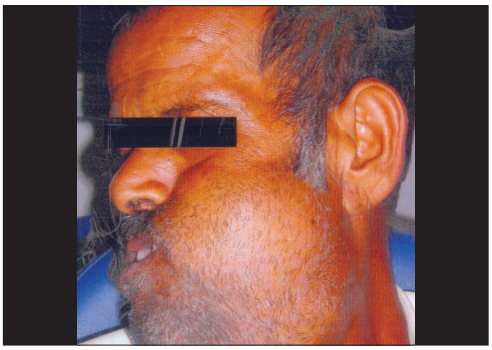

Figure 1: Photograph showing lateral view of lesion in case 1

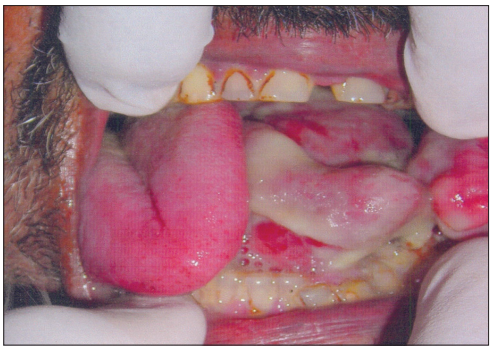

Figure 2: Photograph showing intraoral view of the lesion with evident of displacement of tongue and teeth

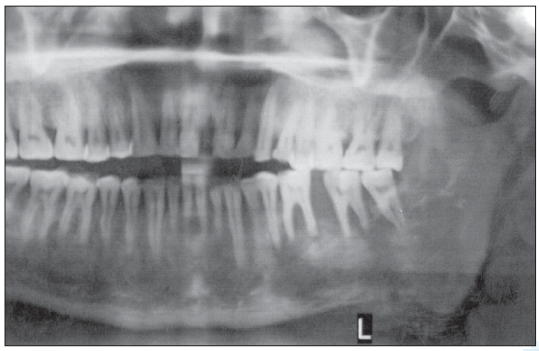

Figure 3: Showing radiograph of displacement of teeth, superficial erosion of the buccal cortical plate and root resorption associated with the posterior teeth on the left side of the mandibular arch

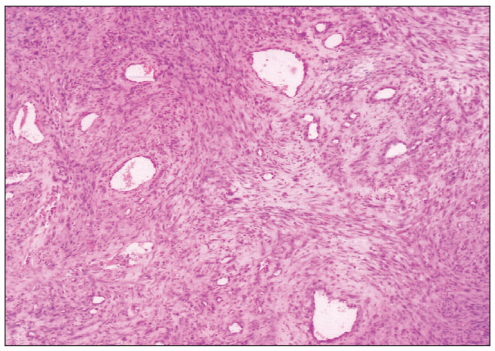

Figure 4: Hematoxylin and eosin stained section showing Herring bone pattern

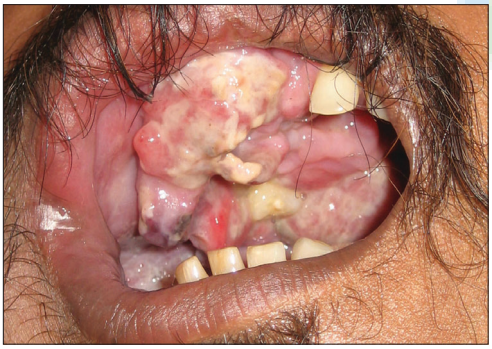

Figure 5: Intraoral view of the lesion in Case 2

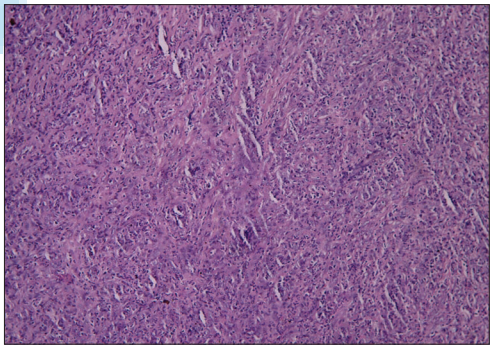

Figure 6: Hematoxylin and eosin-stained section showing fascicular arrangement of cells and predominant cellular stroma

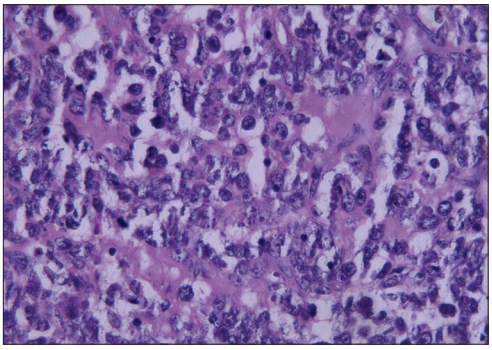

Figure 7: 40x, showing pleomorphic cells, hyperchromaticity and few mitotic figures

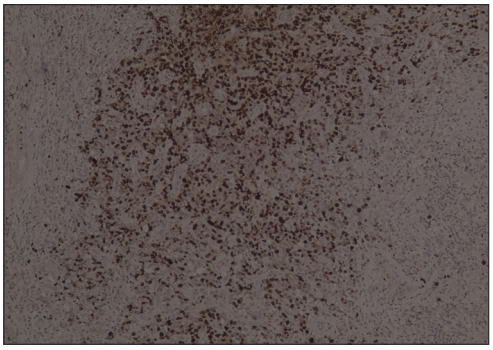

Figure 8: Showing positive for Ki-67 labeling index

some studies, male predilection has been reported. Any submucosal site may be involved although buccal mucosa, tongue and alveolus accounts for more than half of the cases. The symptoms of fibrosarcoma vary depending on size, location and spread of the tumor. Symptoms may include pain, swelling or ulceration. Fibrosarcoma of the oral cavity most often manifests as a clinically innocuous, lobulated, sessile, painless and nonhemorrhagic submucosal mass of normal coloration. On the other hand, aggressive fibrosarcomas tend to be a rapidly enlarging, hemorrhagic mass similar in clinical appearance to an ulcerated pyogenic granuloma, peripheral giant cell granuloma or peripheral ossifying fibroma. Even lesions that do not demonstrate surface ulceration or rapid growth may show destruction of underlying muscle and bone.[6-8]

The American Joint Committee on Cancer developed the most widely used staging system for fibrosarcomas all over the body. The foremost categories of this system include grade (G), size of the tumor (T), lymph node involvement (N) and presence of metastases (M). Low grade and high grade are designated G1 and G3, respectively. The size of the tumor can be <5 cm (2 inches), designated as T1, or >5 cm, designated as T2. If the lymph nodes are involved, it is designated N1, while no lymph involvement is designated N0. Finally, there may be a presence of distant metastases (M1) or no metastases (M0). The following is a list of stages and their indications:Stage IA: G1, T1, N0, M0
Stage IB: G1, T2, N0, M0
Stage IIA: G2, T1, N0, M0
Stage IIB: G2, T2, N0, M0
Stage IIIA: G3, T1, N0, M0
Stage IIIB: G3, T2, N0, M0
Stage IVA: Any G, any T, N1, M0
Stage IVB: Any G, any T, N1, M1

Tumors with lower stage numbers, such as IA and IB, contain cells that look very similar to normal cells, while tumors with higher stage designations are composed of cells that appear very different from normal cells. In higher staged tumors, the cells appear undifferentiated.[8,9]

The differential diagnosis for fibrosarcomas include all spindle cell tumors, and only careful examination of multiple sections and special stains as well as immunohistochemical analysis will permit a correct diagnosis.[1,8]

The treatment of choice for fibrosarcoma is radical surgery. Radiation therapy and chemotherapy can be used in inoperable cases or as a palliative treatment. Prognosis of the tumor is dependent on histological grade, tumor size and adequate surgical treatment with disease free margins. The 5-year survival rate for this disease is poor, ranging from 20 to 35%.[9-12]

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