Fibrosarcoma of the bone is seen in association with Paget’s disease, fibrous dysplasia, and post-radiotherapy. In 1940 Ewing established the initial concept of primary intraosseous fibrosarcoma, and since that time the discussion regarding its existence has been ongoing. Intraosseous fibrosarcoma is an uncommon tumor accounting for approximately 5% of all malignant intraosseous tumors. Its rate of occurrence in the cranium is approximately 15%, with the mandible being the most common site. In the English literatures only 82 cases of fibrosarcoma of the jaw have been reported, also using the key words fibrosarcoma and HIV, the oral manifestation of HIV at Medline and PubMed did not reveal any other case of fibrosarcoma occurring in HIV patients. We report one additional case of intraosseous fibrosarcoma of the maxilla occurring in an HIV-positive patient. Immunosuppression in HIV-positive patients is known to cause three types of malignancies: Kaposi’s sarcoma, non-Hodgkin’s lymphoma, and invasive cervical cancer. Rare neoplasms like Hodgkin’s disease, anal cancer, leukemia, basal cell carcinoma, and squamous cell carcinoma have also been demonstrated. This paper presents one such a rare incidence of an intraosseous fibrosarcoma occurring in an HIV-positive patient.

Introduction

Fibrosarcoma is a malignant neoplasm of fibroblastic origin affecting both soft and hard tissue that can occur in any location, with bone extremities being the main affected sites. Fibrosarcoma of the bone is seen in association with Paget’s disease, fibrous dysplasia, and post-radiotherapy. Ewing established the initial concept of primary intraosseous fibrosarcoma, and since that time the discussion regarding its existence has been ongoing. Intraosseous fibrosarcoma is an uncommon tumor accounting for approximately 5% of all malignant intraosseous tumors. Its rate of occurrence in the cranium is approximately 15%, with the mandible being the most common site. Occurrence in the maxilla is rare, with an incidence ranging from 0% to 6.1%. In the English literatures only 82 cases of fibrosarcoma of the jaw have been reported, also using the key words fibrosarcoma and HIV, the oral manifestation of HIV at Medline and PubMed did not reveal any other case of fibrosarcoma occurring in HIV patients. We report one additional case of intraosseous fibrosarcoma of the maxilla occurring in an HIV-positive patient. Immunosuppression in HIV-positive patients is known to cause three types of malignancies: Kaposi’s sarcoma, non-Hodgkin’s lymphoma, and cervical cancer, which are classified as AIDS defining cancers. Other malignancies occurring in AIDS patients are referred to as non-AIDS defining cancers. Allardice et al. have reported the occurrence of squamous cell carcinoma, small cell carcinoma, adenocarcinoma of the lungs, hepatocellular carcinoma of the liver, transitional cell carcinoma of the bladder, and Ewing’s sarcoma of the bone.

Case Report

A 35-year-old female patient referred to the outpatient department of Hospital Karnataka, India with the chief complaint of a growth in the maxillary anterior region. Past history revealed extraction of teeth 21, 22, 23, and 24 from a general dental practitioner, as the teeth were mobile. Seven days later the patient has noticed a small growth in the same region which was asymptomatic. As the patient is from a low socio-economic status she did not return to the dentist. One month later due to an increase in the size of the growth she had difficulty in speaking and eating; for this reason she was referred to the clinic.

Extra-oral examination showed swelling in the anterior region of the upper lip, extended from the vermilion border of the lip to the ala of the nose, and laterally extended from midline to the canine fossa. On palpation, the temperature of the overlying skin was increased and tender.

Intra-oral examination showed a growth with areas of ulceration. The growth was seen on the left quadrant, extending from 12 to 25. The lesion measured 3 × 2 cm and was an irregular, ill-defined. The surface was pale pink with patchy areas of redness. Submandibular lymph nodes were palpable, enlarged, tender, and firm (Figure 1).

An orthophantomograph showed ill-defined radiolucency extending from midline to the mesial of 25. Posteriorly there was discontinuity in the floor of the maxillary sinus (Figure 2). Upon routine laboratory investigation the patient was found to be HIV-positive. The initial test performed for HIV was the tri-dot test; subsequent confirmation was by Western blot test. The patient did not have any symptoms and was unaware of her HIV status. Since the patient did not agree for further investigation, the CD4+ count and viral load estimation were not performed.

Incisonal biopsy was taken and histopathologically the section showed highly cellular stroma with proliferating fibroblasts. The cells showed a typical “herringbone pattern” with sheets of cells arranged in intertwining whorls. Tumor cells showed hyperchromatism, cellular pleomorphism, and increased mitotic activity. In a few areas the cells were arranged in interlacing fascicles and a moderate amount of collagen was seen (Figure 3). Additionally, immunohistochemical staining with vimentin was done, which showed positivity for fibroblasts (Figure 4). The diagnosis of low-grade fibrosarcoma was made, mainly based on characteristic histomorphological features. For confirmation of the diagnosis the patient underwent a maxillectomy. The excisional specimen showed...
similar histological features, and the final confirmatory diagnosis of low-grade fibrosarcoma was made. The patient showed no recurrence for six months and later was lost to follow up.

Discussion

Soft tissue sarcomas are rare in the maxillofacial region, which accounts for less than 1% of cancers. In the past, fibrosarcoma was the most common soft tissue sarcoma reported, but with the introduction of the electron microscope and immunohistochemistry, it was evident that many previously diagnosed fibrosarcomas were other spindle cell malignant lesions. Soft tissue sarcomas are rare in the maxillofacial region, which accounts for less than 1% of cancers. In the past, fibrosarcoma was the most common soft tissue sarcoma reported, but with the introduction of the electron microscope and immunohistochemistry, it was evident that many previously diagnosed fibrosarcomas were other spindle cell malignant lesions. Fibrosarcoma accounts for approximately 15% of all soft tissue sarcomas, of which only 1% occur in the head and neck region. Frankenthaler et al. studied 118 cases of head and neck fibrosarcoma and found that the neck was the most common site (25%), followed by the face (20%), scalp (16%), and the maxillary sinus (12%). Only 12% of the fibrosarcomas were located intraorally, with half located in the lower jaw. Intraosseous fibrosarcomas may develop endosteally or periosteally, the latter affecting bone and spreads from adjacent soft tissue.

Clinically, the lesion can cause pain, swelling, paresthesia, loose teeth, and ulceration of the overlying mucosa. Intraosseous fibrosarcoma is symptom-free until it reaches a considerable size, then symptoms begin and are usually non-specific. They are reported in all age groups but commonly seen in the 3rd and 5th decades of life. Radiographically, fibrosarcoma presents a destructive lytic lesion with thinning and disruption of the cortex, resulting in soft tissue invasion. The classic fibrosarcoma is characterized microscopically by uniform spindle shaped cells distributed in interlacing fascicles with a herringbone growth pattern, demonstrating immunoreactivity for vimentin. The ultrastructural finding exclusively shows presence of fibroblastic cells, showing a mild degree of nuclear pleomorphism and rare mitosis, with a collagenous stroma representing low grade fibrosarcoma. High grade lesions show an intense nuclear pleomorphism, greater cellularity, and atypical mitosis. The nuclei can be spindle shaped, oval, or round. Microscopically, the well-differentiated tumors are easily diagnosed as fibroblastic lesions due to their characteristic histologic appearance, showing cells arranged in fascicles with a herringbone appearance. Otherwise the histologic appearance of the undifferentiated tumors...
may simulate many other tumors of soft tissue origin\cite{16}; such as nodular fascitis, cellular benign fibrous histiocytoma, fibromatosis, malignant peripheral nerve sheath tumor, malignant fibrous histiocytoma, and monophasic fibrous synovial sarcoma. Each of these lesions have characteristic histomorphologic features when compared with fibrosarcoma.\cite{19} The present case was of a low-grade fibrosarcoma that showed characteristic histomorphologic features, thus we only did vimentin staining which showed strong positivity for fibroblasts.

Nodular fascitis is reactive myofibroblastic proliferation that grows rapidly and is marked by its cellularity and immature cellular appearance. It differs from fibrosarcoma by its irregular growth pattern and cells that are arranged in short bundles. Cellular benign fibrous histiocytoma may be distinguished from fibrosarcoma by its fascicles, which are not as regular or as sweeping as those seen in fibrosarcoma. Areas of more conventional benign fibrous histiocytoma may be present and are extremely useful in this distinction.\cite{19} In fibromatosis, mitosis are generally absent and the grade of cellular atypia is very low.\cite{16} Malignant fibrous histiocytoma is characterized by a storiform to haphazard growth pattern and multinucleated bizarre giant cells. Siderophages and xanthoma cells are common features. Malignant peripheral nerve sheath tumors may display areas that are virtually indistinguishable from fibrosarcomas. The cells showing neural differentiation often have a wavy or buckled appearance, rather than the finely tapered fibroblasts of fibrosarcoma. Although cells can be arranged into an irregular fascicular growth pattern, the long and sweeping fascicles characteristic of fibrosarcoma are usually not present. Monophasic synovial sarcoma cells are more ovoid and arranged in an irregular fascicular pattern.\cite{19} The histological appearance of high grade fibrosarcoma may be similar to malignant fibrous histiocytoma, liposarcoma, synovial sarcoma, or Kaposi’s sarcoma.\cite{16,19} Immunohistochemical markers help to differentiate between these spindle cell neoplasms and high grade fibrosarcoma. The cases presented in the early seventies and eighties were probably other spindle cell tumors. Many spindle cell neoplasms were placed into this category without further sub-classification because of the lack of advanced techniques. However, over the past decade with expanding knowledge about soft tissue lesions, it is evident that a much smaller percentage of head and neck neoplasms are true fibrosarcomas.\cite{19}

The etiology for most cases reported is not known, but they are known to develop in old scars or in areas of previous radiation.\cite{20} The association of our case with an HIV infection may suggest a possibility of fibrosarcoma being an immunodeficiency-associated cancer.

The most common cancer associated with an HIV infection is Kaposi’s sarcoma. Approximately 30% of patients with AIDS develop this sarcoma, which is commonly seen in young adults with a mean age of 39 years. Initially, the lesion presents itself as small flat, pink patches, which later develop a classic blue-violet popular appearance. Kaposi’s sarcoma occurs in almost all locations and most patients have multiple oral or cutaneous lesions. Histologically, it occasionally shows a bland spindle cell component, initially centered around the proliferating vascular channels, which at times coalesce. Between these spindle cells are numerous slit-like or sieve-like spaces, which are not evident in fibrosarcomas. The cause for Kaposi’s sarcoma in HIV patients is due to profound immunosuppression.\cite{19} Immunodeficiency associated with HIV infection is also reported to cause other cancers, such as anal cancer, colorectal cancers, and Hodgkin’s disease. The strength of the association between these non-AIDS defining neoplasms and HIV infection has varied considerably. Thus, it is important to develop a more complete understanding of the association between HIV infection and cancer. The increased rate of cancer among AIDS patients can be attributed to either immunosuppression due to HIV infection or common risk behaviors shared by the specific cancer and the HIV infection.\cite{11}

There are also reports in HIV/AIDS literature of patients suffering with soft tissue leiomyosarcomas, dermatofibrosarcoma protuberans, metastatic Kaposi’s sarcoma, primary lymphoma, papillary thyroid carcinoma, and simultaneous occurrence of occult papillary thyroid cancer and medullary thyroid cancer, due to chronic immunosuppression among adults.\cite{21} Yurino et al. have reported one case of fibrosarcoma of the crus in their study on the assessment of clinical manifestations of HIV infection in positive sero-adults in Russia.\cite{22} The present case appears to be a rare case report of intraosseous fibrosarcoma of the jaw occurring in an HIV patient, which may be due to immunosuppression.

The treatment of choice for fibrosarcoma is a wide margin surgical resection. The need for adjuvant radiotherapy and or chemotherapy is still unclear, and is normally indicated in high-grade tumors because these tumors may present subclinical or micro-
scopic metastases at the time of diagnosis. Prophylactic neck dissection is also controversial. The treatment protocol carried out for the present case was maxillectomy. Patient follow up was for six months with no recurrence, however later she was lost to follow up.

The overall survival rate of ten years varies from 21.8% to 83%. The clinical stage, the histological grade of malignancy, and local recurrences are most important in determining prognosis of the lesion.

In conclusion, this article presents a rare case of intraosseous fibrosarcoma of maxilla occurring in an HIV-positive patient. The possible etiological role of HIV in fibrosarcoma needs elucidation with further case reports.

Reference