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Fibrosarcoma unilateral site in maxilla: a rarest case report

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Abstract

Fibrosarcoma is a malignant mesenchymal neoplasm of fibroblasts that rarely affects the oral cavity and can cause local recurrences or 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.

Keywords: Fibroblasts, fibrosarcoma, immunomarker, case report

Introduction

Soft tissue sarcomas are rare in the oral and maxillofacial region and account for less than 1% of the cancers[1]. At one time, fibrosarcoma was the most common soft tissue sarcoma. With the introduction of electron microscopy and immunohistochemistry, it became evident that many previously diagnosed fibrosarcomas were other spindle cell malignant lesions.

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[2-8]. 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[11-14]. The treatment of choice is radical surgery; radiation therapy and chemotherapy can be used in inoperable cases. We are hereby presenting case of primary fibrosarcoma of the oral cavity reported to the outpatient department of Dentistry in Sardarpatel Medical College in Bikaner, Rajasthan, India.

Case report

A 38 year-old male presented with dental pain in the Right Face in May 2012. Physical examination revealed a 1cmX2 cm tender swelling on the palatal side of area 14 to 17 in the right maxilla. There was no other any abnormality detected. A Intraoral periapical radiograph showed a radio-lucency with bony destruction in the right side of the maxilla in premolar and molar area. Patients given history of extraction of teeth in upper right side before 15 days .The complete blood count, liver and renal function test and chest X-ray were within normal limits. On extra-oral examination facial profile was normal. The submandibular, submental and cervical lymph nodes were not palpable. On intra orally the patient exhibited an ulcerative,granulomatous growth of size 1 x 2 cm extending from the midline of the palate to the buccal gingiva adjoining 14 to 17 region (Fig.1). It showed erythematous mucosa and was firm, fixed and tender on palpation.

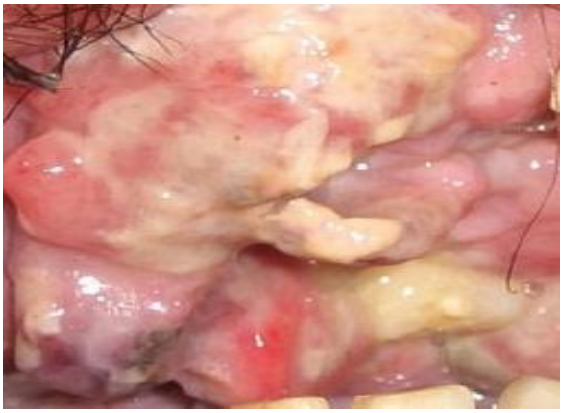

Fig. 1: Ulcerative, granulomatous growth of size 1 x 2 cm in right side of maxilla.

Radiographically investigated by orthopantomograph (Fig.2) and Water s View, which revealed soft tissue shadow of tumor mass with diffuse irregular bony margin, infiltrating and destructive border on right side of posterior maxilla extending from 14 region to 17 area.

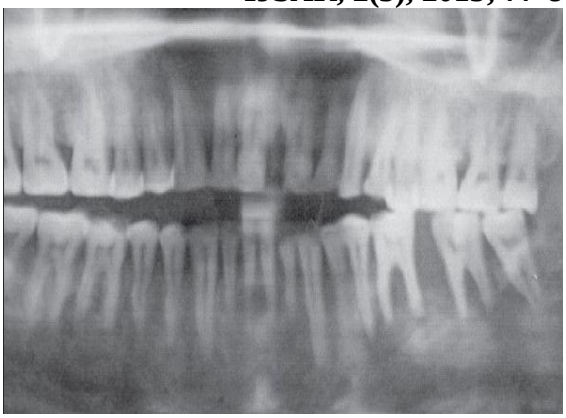

Fig. 2: Radiographically investigated by orthopantomograph.

After routine investigations, Entoto removal by Surgical resection with healthy margin. Biopsy specimens send to our pathology department in Sardar Patel medical college, Bikaner. Hematoxylin and eosin stained sections demarcate the presence of fibro-cellular connective tissue stroma composed of sheets of proliferating oval and spindle shaped fibroblasts. Cells were arranged in herring bone pattern with parallel sheets of cell arranged in intertwining whorls. Cells showed hyperchromatism, moderate cellular and nuclear pleomorphism, increased mitotic figures. Also showed presence of few collagen fibers, diffuse chronic inflammatory cell infiltrate and few areas of hemorrhage (Fig.3). Histopathological diagnosis of intermediate grade fibrosarcoma was made.

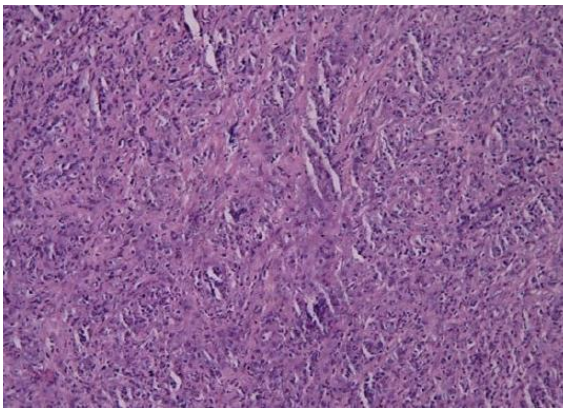

Fig. 3: showed presence of few collagen fibers, diffuse chronic inflammatory cell infiltrate and few areas of hemorrhage.

Discussion

Fibrosarcoma occurs as a primary malignant bone tumor, incidence of less than 5%[1]. Fibrosarcoma arise from the medullary connective tissue of the skeleton or can originate in the soft tissues adjacent the bone, the later lesion called as Periosteal fibrosarcoma has a good prognosis[2] . Fibrosarcoma arising from the normal bone is called as Primary fibrosarcoma, whereas those originating from the pre-existing bony lesions are called as Secondary fibrosarcoma.

World Health Organization (WHO) in 2002 defined fibrosarcoma as a 'malignant tumor, composed of fibroblasts with variable collagen and, in classical cases, a herringbone architecture[3].

Clinically, fibrosarcoma mostly presents as an innocuous, multilocular, sessile, painless and nonhemorrhagic submucosal mass of coloration, however, sometimes may be a rapidly enlarging, hemorrhagic mass similar in clinical appearance to an ulcerated pyogenic granuloma and peripheral giant cell granuloma.[4] The intraosseous fibrosarcoma is symptom-free until it reaches a remarcable size, symptoms begin such as pain, swelling, paresthesia, loose teeth, and ulceration of the overlying mucosa.[5] In our case the patient was asymptomatic with classical clinical findings.

Radiographically, the tumor appears like an osteolytic lesion, with ill-defined borders, thinning and disruption of the cortex, resulting in soft tissue invasion.[5,6] as we have seen in the our case. 3-D CT finding suggest Destructive bony lesion, frequently associated with extra osseous soft tissue mass and MRI finding Low to intermediate signal intensity with inhomogeneous enhancement[7]

Histologically fibrosarcoma consist of largely interlacing fascicles of fibroblasts. The classical “Herrring bone” patterns of interlacing bundles are arranged perpendicular to each other and is very pronounced in some cases. The tumor divided in to well and poorly differentiated types. In our case, the tumor cells were large & spindle shaped with elongated & hyperchromatic nuclei. The cytoplasm was scanty and it showed mitotic figures in large numbers exhibiting the characteristics of Mild grade Fibrosarcoma. The five year survival rate for soft tissue fibrosarcoma was approximately 60%[8] as against the fibrosarcoma occurring in bone which has a five year survival rate of 31.7%[9].

The diagnosis can occasionally be difficult to distinguish fibrosarcoma from other malignant neoplasms such as malignant fibrous histiocytoma, fibroblastic osteosarcoma, leiomyosarcomas, lymphomas and metastatic disease. This cognitive state of diagnostic dilemma demands immunohistochemistry, that adds to the pathologist's diagnostic techniques when trying to distinguish morphologically similar tumors. Fibrosarcoma stains strongly positive for the intermediate filament vimentin. Markers for muscle (desmin, smooth muscle actin, HHF-35), human osteoblasts (osteocalcin), macrophages (CD-68), leukocyte common antigen (LCA), neural tissue (s100, neuron specific enolase), melanoma (HMB-45), neutrophils (CD-31), hematopoietic cells (CD-34), epithelial tissue (cytokeratin, epithelial membrane antigen), and CD-99 will be absent. Thus, fibrosarcoma is essentially a diagnosis of exclusion[10-14].

The treatment of choice was wide resection with free margins[15]. Radiotheraphy is beneficial in inoperable tumor and chemotherapy is indicated for palliative treatment. Local recurrence is most common but metastasis is rare. The prognosis however is poor but is favorable than most other malignant neoplasm occurring in this region[16].

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