CASE REPORT

FIBROSARCOMA OF THE MANDIBLE -A CASE REPORT

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Abstract

Fibrosarcoma in the head and neck region is a rare entity. The advent of immuohistochemistry and electron microscopy has added a new dimension in its clinicopathological differentiation from other types of spindle cell lesions. Fibrosarcoma can be primary, or secondary due to other bony lesions like Paget's disease, Osteomyelitis etc. In this neoplasm there is malignant proliferation of fibroblast which can cause local recurrences or metastasis. Herring bone pattern of the pleomorphic fibroblasts can be seen histopathologically.

In this case report, a 70 year old patient presented with a large, diffuse, fleshy swelling involving the body of the mandible and the left side of the associated soft tissues. Immunohistochemically, the cells only showed immunoreactivity for vimentin and negativity for S-100 protein, pancytokeratin, desmin. Based on, clinical, radiological, histopathological and immunohistochemical findings the final diagnosis was given as fibrosarcoma.

Key Words: Fibrosarcoma, Herring bone pattern, Immuno histochemisry

INTRODUCTION

"Fibrosarcoma", a malignant mesenchymal neoplasm of the fibroblasts, rarely affects the oral and maxillofacial region, accounting for less than 1% of all the cancers. The etiology of the entity remains debatable; fibrosarcomas have been reported in association with several conditions such as Paget's disease, fibrous dysplasia, chronic osteomyelitis, bony infarcts and previously irradiated areas of bone in cases of malignancy. Clinically the lesion may present with an extensive, fleshy, sessile or pedunculated, exophytic, lobulated mass that may exhibit pain, paraesthesia, swelling, loosening of regional teeth along with ulceration of overlying mucosa. Classical presentations of nodal metastasis are usually not evident. [1]

Radiographically, an osteolytic lesion is usually present, with ill-defined borders; however, fibrosarcoma of the jaws cannot be distinguished from other destructive lesions of the bone. [2] The sarcomas involving bone and extremities often present with pain and swelling after a long duration of symptoms and pathological fracture in the extremities is evident.

Histopathologically, the lesion exhibits diverse characteristics. Fibrosarcomas can be graded, with low and high grade of malignancies. Low grade malignancies show spindle shaped fibroblasts, arranged in streaming fascicle with low to moderate cellularity and a herring bone pattern. High grade malignancies are characterized by their immense nuclear pleomorphism, greater cellularity and atypical mitotic figure along with presence of ovoid,

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Figure 1-Extraorally a diffuse, moderately large swelling involving the left lower third of face

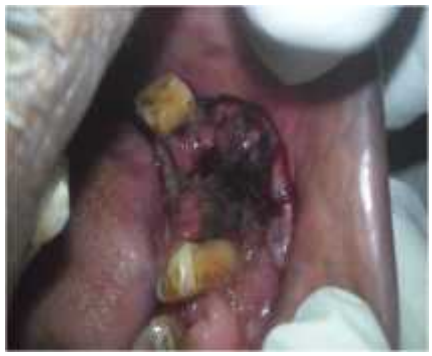

Figure 2-Intraorally, a diffuse ulceroproliferative fleshy growth involving the left lower alveolar ridge

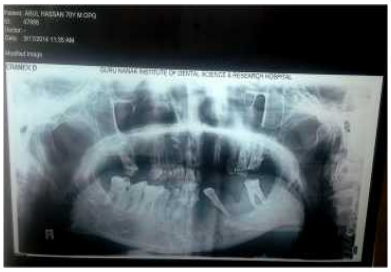

Figure 3 - OPG revealed presence of a diffuse osteolytic lesion involving the left mandibular body region extending from 33 to 38 regions along with floating on air appearance of 33 and 38.

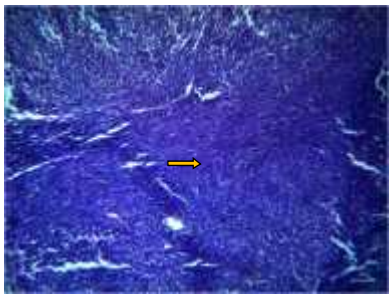

Figure 4- Histopathological evaluation revealed the presence of actively proliferating neoplastic cells showing pronounced cellular & nuclear pleomorphism within the fibrovascular connective tissue stroma

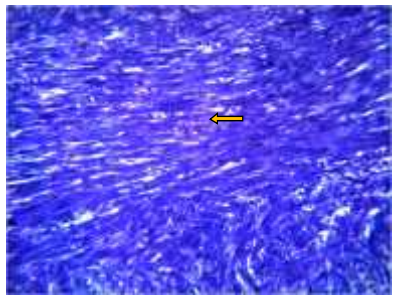

Figure 5- The tumor cells appear to be spindle shaped, being arranged in streaming fascicles and occasionally exhibiting ''herring bone'' pattern of distribution

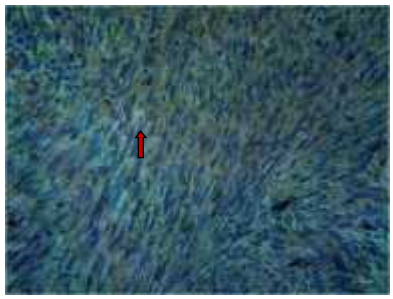

Figure 6 - IHC revealed Vimentin positivity

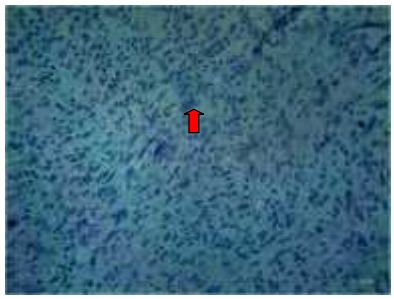

Fig 7a - Cytokeratin negativity on IHC involving the cytoplasm

Figure 7b-S100 negativity involving the nucleus and cytoplasm in IHC

Figure 7c-Desmin negativity on IHC

round or spindle shaped nuclei. [3]

Fibrosarcoma bears a strong histopathological resemblance to other lesions like malignant fibrous histiocytoma or liposarcoma. The destructive radiographic pattern and the fleshy to firm mass of fibrosarcoma could be suggestive of both malignant fibrous histiocytoma and osteosarcoma. Fibrosarcomas are distinct from malignant fibrous histiocytomas by virtue of its uniform spindle cell fibroblastic cell population arranged in parallel bands or a "herring bone " pattern. Osteosarcomas are primarily malignant tumor of osteoblasts (alkaline phosphatase positive), whereas fibrosarcomas are malignant tumors of fibroblasts (alkaline phosphatase negative). Electron microscopy or immunohistochemistry (IHC) play a vital role in differentiating these lesions. Electron microscopy has contributed to the identification of the fibroblastic phenotype in case of fibrosarcomas. The positive immune staining for vimentin along with pan keratin negativity, negativity towards muscle immune markers like desmin, S-100 help in diagnosing the lesion.[3,4]

The focus of treatment is a very wide local excision using 3 cm clinical margins in soft tissue and bone as well as assessing margins with frozen sections at the time of surgery. Lymph node dissection is not required. Like most sarcomas, fibrosarcomas are malignancies for which the prognosis correlates most closely with histologic grade. Using a four grade system of advancing histological grade, reported 5 year survival rates are 82% for grade I, 55% for grade II, 36% for grade III and 21% for grade IV tumors.[4] Gadolinium enhancement helps to improve the tissue characterization in case of fibrosarcomas, where the highly cellular zones are usually represented as homogenous or non-homogenous enhancement areas.[5]

Presented here is a case of a 70 year old male patient with diffuse swelling involving the left lower third of face which, was diagnosed as Fibrosarcoma based on its clinical, radiographical, histopathological and immunohistochemical features.

A 70 year old male patient, from a semi urban area of Agarpara, North 24 Parganas, reported to the Department of Oral & Maxillofacial Pathology, Guru Nanak Institute of Dental Sciences & Research, Kolkata, with the chief complaint of discomfort & pain involving lower left side of face for last 45 to 60 days. The swelling was relatively fast progressing, but without any symptoms initially. Later, he developed discomfort, mild pain and heaviness over the region. Frequent episodes of hemorrhage were also noted along with mobility and spontaneous exfoliation of regional teeth.

Extra oral clinical examination revealed a diffuse, relatively large swelling involving the left lower third of the face measuring about 3cm x 2cm and extending from left angle of mouth to ascending ramus and involving the left sub mandibular region.(Fig.-1). The overlying skin appeared stretched but without any secondary changes. Palpation revealed relatively firm, mildly tendered swelling with stretched overlying skin and localized rise of temperature. No skin fixity to the underlying structures was noted. Pronounced left submandibular lymphadenopathies were also noted. Past medical and surgical histories were non contributory. Intra oral clinical examination revealed the presence of a diffuse ulceroproliferative fleshy growth involving the left lower alveolar ridge extending from 33 to 38 regions. The lesion exhibited extensive areas of nodularity and hemorrhage with marked induration at the borders (Fig.2). The regional teeth (33, 38) revealed grade III mobility with absence of 36 and 37. There was no discharge or pulsation.

Orthopantomograph (OPG) revealed the presence of partially edentulous upper and lower jaws and a diffuse osteolytic lesion involving the left mandibular body region extending from 33 to 38 regions along with moth eaten appearance at the borders with floating in air appearance of 33 and 38 (Fig 3). No other abnormalities were detected and no other radiographs were performed also.

Thorough medical and hematological investigations revealed presence of anemia and raised ESR levels. Incisional biopsy was performed under local anesthesia.

Histopathological evaluation revealed the presence of actively proliferating neoplastic cells showing pronounced cellular & nuclear pleomorphism within the fibrovascular connective tissue stroma(Fig 4). The tumor cells appear to be spindle shaped, being arranged in streaming fascicles and occasionally exhibiting "herring bone" pattern of distribution (Fig 5). Occasional areas of atypical round to ovoid cells showing pronounced pleomorphism were also noted along with areas of intense non specific chronic inflammatory cell infiltration.

The case was provisionally diagnosed as an anaplastic spindle cell neoplasm and hence was sent for further immunohistochemical evaluation (IHC).

IHC revealed positivity for Vimentin (Fig 6), but was negative for Cytokeratin (Fig 7a), S-100(Fig 7b), and Desmin (Fig 7c). Hence the case was diagnosed as “Fibrosarcoma” and referred for further management to Chittaranjan National Cancer Institute (CNCI), Kolkata. The patient was also advised for periodic follow-ups but he did not turn up for the same.

DISCUSSION

Fibrosarcoma is a malignant mesenchymal tumor of fibroblastic origin with no established

epidemiological and etiological factors whatsoever. The behavior of the tumor may vary, depending on its occurrence in either soft tissue or bones, frequently involving the extremities.Fibrosarcomas are rarely seen involving the jaws especially the mandible. They may arise endosteally or periosteally, with the latter affecting the bone by spread from soft tissues. Erosion of involved teeth may occur.[6,7] In the present case, a fleshy, ulceroproliferative mass involved the left side of lower jaw with a history of spontaneous exfoliation of regional teeth and concomitant displacement of 35 and 38.

Radiographically, the tumors are usually osteolytic with relatively ill-defined peripheries, occasionally overlapping with features of chondrosarcoma and osteolytic osteogenic sarcomas. Our case presented likewise features.[8]

Histopathological analysis have not only opened up a plethora of differential diagnosis but also varying morphologies within the same entity –beginning with bland looking mass resembling fibromas, to herring bone arrangement of malignant cells at places bearing the classical appearance of fibrosarcomas to even anaplastic areas of challenging histomorphological diagnosis. With increasing grade of fibrosarcomas, there is also a corresponding increase in the metastatic potential and aggressiveness.[9]

Low grade malignancies show spindle cells in fascicular arrangement, low to moderate cellularity, and a few mitotic figures, with spindled, oval or rounded nuclei. High grade malignancies revealed high cellularity, increased number of atypical mitotic figures, with overlapping nuclei showing pleomorphism. The present case revealed areas of spindle cells, exhibiting pronounced nuclear and cellular pleomorphism, being arranged in 'herring bone" pattern at places along with areas of atypical round to ovoid cells. Non specific chronic inflammatory cell infiltrations were usually noted in the less abundant connective tissue matrix.[10]

Immunopositivity for vimentin, together with negativity for muscle markers help to diagnose fibrosarcoma. The present case also presented with vimentin positivity with negativity for cytokeratin and muscle markers such as desmin and S-100.

This lesion is to be differentiated from malignant fibrous histiocytoma, liposarcoma, synovial sarcoma and a plethora of other spindle cell and poorly differentiated neoplasm.[11]

The presented case had been sent for surgical management. Radiation and chemotherapy, in inoperable cases is followed by palliative treatment.[12]

Prognosis is related to the histopathologic grade and the margin status following surgical intervention.

SUMMARY AND CONCLUSION

Fibrosarcoma is a rare tumor, which generally affects the long bones and deep soft tissue. It must be differentiated from other similarly rare forms of sarcoma that may involve the oral cavity. Thorough clinical, radiographical, and histopathological protocols must be followed, along with that, relevant immunohistochemical (IHC) analyses, such as vimentin, S-100, desmin and cytokeratins, would point out to characteristic features-which may be helpful in distinguishing the lesion from various other types of malignant mesenchymal tumors.[13] There is a very fine distinction between low grade malignancies and other types of spindle cell sarcomas.

Hence, careful microscopic examination with accurate sampling, correct knowledge of pathology and practical experience of pathologist is required to render the correct diagnosis.[14]

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