Fibrosarcoma of the Mandible: A Diagnostic Embarrassment

Mostafa Md. Anisuzzaman¹, Mohammad Khursheed Alam¹, Safiqur Rahman Khan³, Mohammad Kamrujjaman¹

ABSTRACT

Background: Fibrosarcoma is a malignant mesenchymal neoplasm of fibroblasts that rarely affects oral cavity and can cause local recurrences or metastasis. The aetiologic factors are still unknown, but many authors have reported the radiation therapy history as an important aetiological factor, followed by trauma and underlying conditions like Paget’s disease, fibrous dysplasia or chronic osteomyelitis. Although fibrosarcomas are rare, they can occur anywhere in the body. The most common sites are in the retroperitoneum, thigh, knee, and distal extremities. Fibrosarcoma of mandible is rare, with an incidence which ranges from 0.6-6.1% of all primary fibrosarcomas of the bone. The prognosis for fibrosarcomas is poor with a five-year survival rate of 20-35%.

Case note: We report a rare presentation of gingival fibrosarcoma in a young boy, who presented with painless lump on lower right posterior segment of mandible.

Conclusion: Clinical follow-up of patients cannot provide any significant conclusions concerning prognosis, but the data undoubtedly show the malignant biological nature of these neoplasms and similar prognosis to that of other fibrosarcomas described in the oral cavity.

KEY WORDS
fibrosarcomas, mandible, neoplasm

INTRODUCTION

Fibrosarcoma (FS) is a malignant neoplasm of fibroblastic origin and may either arise in the soft tissue or be of primary intraosseous origin (20% of all cases)³,⁵. The latter origin has been debated since 1940, when Ewing⁶ established the initial entity, and is now generally accepted.

It is a rare tumor, accounting for approximately 5% of all malignant intraosseous tumors⁷,⁸, and especially affects the long bones. Its occurrence in the head and neck is about 10% of cases, of these the mandible being the commonest site⁹. More than 75 cases in the mandible have been reported in the English language literature¹⁰.

Histologically it is often difficult to distinguish fibrosarcomas from other soft tissue sarcomas and diagnosis is often achieved by exclusion. Differential diagnosis must consider other malignant tumors, i.e. monophasic fibrous synovial sarcoma, malignant fibrous histiocytoma, malignant nerve sheath tumor and liposarcoma, as well as benign tumors, i.e. benign fibrous histiocytoma, nodular fasciitis, fibroma and fibromatosis. The low-grade myxofibrosarcoma type, however, is often confused with the fibromyxoid sarcoma type, and morphological distinction is sometimes difficult and problematic¹¹. Given the diagnostic difficulty in differentiating between these different forms, immunohistochemical analysis is of considerable help in diagnosing fibrosarcomas.

We report two cases of primary mandibular fibrosarcoma together with clinical, histological and immunohistochemical findings, and discuss differential diagnosis of this rare tumor of the oral cavity.

CASE REPORT

A 15-year old male was referred to Department of oral and maxillofacial surgery with a dull painful swelling on the lower right posterior segment of mandible for 2 months. During intraoral examination the swelling was smooth surface, firm in consistency, no tender on palpation, fixed with under laying structure, over laying mucosa was normal in color which extend anteriorly posteriorly from distal to lower second molar to anterior border of ramus of mandible and mesiolaterally from right buccal vestibule to lingual vestibule (Figure 1). It is also associated with difficulty in chewing food. Extra orally there was no swelling on right side of face. The right submandibular lymph node was palpable and non-tender. Then incisional biopsy was taken first which reveals Capillary lobular haemangioma. But the result has no clinical correlation with this lesion. For that reason, we decided for biopsy again which reveals inflammation granulation tissue. Then we excised the soft tissue mass with extraction of impacted third molars under general anesthesia (Figure 1). The postoperative specimen was sending for histopathological examination. The report showed low-grade fibrosarcoma, which is very surprising to us. Then we send the patient to oncologist for further management. They advised him for immunohistochemistry, ultrasonography of the neck and some routine blood test. The immunohistochemistry showed (Figure 2):

1. Desmin: The tumor cells are Negative.
2. Smooth Muscle Actin (SMA): The tumor cells are Positive.
3. CD 34: The tumor cells are Negative.
4. Ki 67: Negative, percentage of positive nuclei < 10%
5. H caldesmon: The tumor cells are Positive.
Anisuzzaman M. M. et al.

Interpretation: The Findings favors low-grade myofibroblastic sarcoma by the process of exclusion. The ultrasonography of the left neck showed Negative for malignant cell. (Smear shows scanty cellular material containing a few polymorphs, lymphocytes and histiocytes in the background of blood). Finally oncologist and we are decided for only follow up due to respect of age, sex and available treatment facilities. Patient was evaluated in every 2 months by clinically and radiologically.

DISCUSSION

A fibrosarcoma is a malignant tumour that arises from fibroblasts. Fibrosarcomas can arise in soft tissues or within bones. Intra-osseous fibrosarcomas may develop endosteally or possibly periosteally, the latter affecting bones by spread from adjacent soft tissues. The mean age for the occurrence of fibrosarcomas is between the 2nd and 6th decades of life, with equal gender distribution. In the head and neck region, only 0.05 % cases have been reported, with mandible as a rare site of occurrence. Clinically, the tumour presents with a swelling which is associated with pain and paresthesia and occasionally with loss of teeth and ulceration of the overlying mucosa. Radiological imaging of fibrosarcomas has revealed radiolucent lesions with a geographical, moth-eaten or a permeative pattern of bone destruction.

The change of appearance in radiographic imaging in the present case was striking. The radiograph showed an impacted lower left third molar with radiolucency around the tooth, which gave an appearance of an odontogenic cyst or a tumour. Further histopathological and immunohistochemical investigations are necessary for making a final diagnosis. Microscopically, low grade fibrosarcoma has been characterized as uniform spindle shaped cells which are arranged in fascicles, having a herringbone growth pattern. There are mild degrees of nuclear pleomorphism and rare mitoses. High-grade lesions show intense nuclear pleomorphism, a greater cellularity and atypical mitoses.

In the present case, pleomorphic spindle shaped cells were arranged in a herringbone pattern, which were associated with collagen and mild mitosis, which are characteristic of fibrosarcoma characteristic of a fibrosarcoma. The histological appearance of a fibrosarcoma is similar to that of Solitary fibrous tumour, rhabdomyosarcoma and leiomyosarcoma.

To rule out a Solitary fibrous tumour, rhabdomyosarcoma and leiomyosarcoma from a fibrosarcoma, an Immunohistochemical (IHC) study was carried out for the following markers: desmin, Smooth muscle actin (SMA), CD 34, Ki 76, and H Caldesmon. IHC showed Smooth muscle actin -positive cells and Ki 76- positive less than 10% cell, while the other immunomarkers showed negativity. A fibrosarcoma is essentially a diagnosis of exclusion and by definition. The treatment choice of fibrosarcomas is radical surgery. Radiation therapy and chemotherapy can be used for inoperable cases or as a palliative treatment, as their role in treatment is still unclear. They are given for high grade tumours, as these tumours may present with subclinical or microscopic metastases. Prognosis of the tumour is dependent on histological grade, tumour size and adequate surgical treatment with disease free margins. The five year survival rate for this disease is poor, ranging from 20 to 35%.

In our cases we excised the mass with extraction of impacted lower right third molar and follow up. Oncologist decided this treatment protocol after proper evaluation of postoperative biopsy report, immunocyto-histochemistry report, USG of neck report, clinical feature.

CONCLUSION

This rare tumor, which generally affects the long bones and deep soft tissue, must be differentiated from other similarly rare forms of sarcoma that may involve the oral cavity. Immunohistochemical tests, such as desmin, smooth muscle actin, CD 34, Ki 76 and H caldesmon, as well as conventional clinicopathological features may be helpful to distinguish the various types. There is still a paucity of reports on fibrosarcomas in the head and neck region.

REFERENCES


