

CASE REPORT

Fibromyxoid Sarcoma With Localised Destruction Of Surrounding Tissue: A Case Report And Review Of Literature

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ABSTRACT

Introduction: Fibromyxoid sarcoma of the oral cavity is a very rare, highly vascular, unencapsulated and locally invasive tumor. It should be operated soon. However it can recur depending on the depth of the tumour.

Case report: Case report with complete review of other studies along with comparative findings and different management approaches during the study period was taken in account. The presentation and procedures to differentiate the sarcoma from other diseases was done. It was found that local resection of the tumour and post-operative radiotherapy was a better plan of management. However, in our case the patient party did not consent for surgery. The patient is still under follow up.

Conclusion: Local resection of the tumour and post-operative radiotherapy is a better plan of management.

Keywords: Fibromyxoid Sarcoma, mandible, swelling

How to cite this article: Hiranya Prova Saikia, Pradip Kumar Tiwari. Fibromyxoid sarcoma with localised destruction of surrounding tissue: A case report and review of literature. *International Journal of Contemporary Medical Research*. 2015;2(2):405-407

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Source of Support: Nil

Conflict of Interest: None

INTRODUCTION

Fibrosarcoma (FS) is a malignant mesenchymal neoplasm of the fibroblasts that rarely affects the oral cavity. 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).^{1,2} The latter origin has been debated since 1940, when Ewing established the initial entity, and is now generally accepted.³ It is a rare tumor, accounting for approximately 5% of all malignant intraosseous tumors,⁴⁻⁶ 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 site.⁷ Clinically, in the oral cavity the major symptoms are pain, swelling, and sometimes loosening of the teeth,^{8,9} paresthesia and occasionally ulceration of the overlying mucosa.¹⁰ Microscopically there is proliferation of fibroblasts with variable amounts of collagen and reticulin fiber formation.¹¹ FS sometimes contains hyalinized collagen that cannot be differentiated from osteoid substance. Characteristically, the cells are uniform spindle-shaped (spindle cell sarcoma), multipolar, with elongated oval or round hyperchromatic nuclei and vary little in size and shape. These cells are arranged in interlocking bands or fascicles that run in different directions, and may be arranged in a herringbone pattern with areas of myxomatous, pseudomyxomatous and cartilaginous changes.¹² Its histological grading is based on the degree of cellularity, degree of cellular differentiation, mitotic activity, the amount of collagen produced by the tumor cells and the extent of necrosis.

CASE REPORT

A 14 year old female presented to the out patient

department, Department of Otolaryngology and Head and Neck Surgery, Assam Medical College, Dibrugarh, Assam, India, with a chief complain of swelling of the cheek and upper jaw with difficulty in chewing and pain over the swelling (Figure-1). The patient also complained of occasional pain over the ears and difficulty in vision. Her HPE (Figure-2) from the tissue revealed clusters of atypical spindle shaped cells embedded in myxoid matrix leading a diagnosis towards fibromyxoid sarcoma. Her NECT and CECT scan of faciomaxillary region (Figure-3) showed a well-defined heterogenous, predominantly isodense lesion, measuring approximately 9.6 cm x 5.9 cm x 6.8 cm, showing heterogenous enhancement on post-contrast study involving the left maxillary sinus and superior alveolar arch. The lesion caused destruction of all the walls of the left maxillary sinus, left orbital floor, lamina of the left pterygoid, superior alveolar process and zygoma. There was involvement of the masticator muscle on the left. The lesion extended to the oral cavity and left nasal cavity and exerting mass effect and causing mild right sided deviation of the nasal septum. There was also evidence of mild left sided proptosis. The lesion displaced the left upper pre-molars and molars. Mucosal thickening was noted in the left sphenoid, bilateral frontal and left sided ethmoidal air cells. Further she was planned for resection and radiotherapy. The patient is under continuous follow up.

DISCUSSION

Fibrosarcoma (FS) is a malignant mesenchymal neoplasm of the fibroblasts that rarely affects the oral cavity. Different histological types of this tumor exist, one of which is myxofibrosar-

coma, which was initially described by Angervall in 1977.¹³ It is characterized by low cellularity composed of spindle-shaped cells with minimal cytological atypia, and with cells deposited in a variably fibrous and myxoid stroma, usually appearing more myxoid than fibrous. Myxofibrosarcoma has been defined as a malignant fibroblastic lesion in which at least 50% of the entire tumor displays a highly vascularized and myxoid stroma with distinctive curvilinear vessels.¹¹ 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 problematic. The treatment of choice is surgical resection with a wide margin.^{14,15} The need for adjuvant radiotherapy and/or chemotherapy is still unclear, and is normally indicated in high-grade tumors because these may present subclinical or microscopic metastases at the time of diagnosis. The 5-year survival rate is 60% for oral soft tissue fibrosarcomas.¹⁶ The potential to recur and to spread of the myxofibrosarcoma type seems also to be related to the anatomical depth of the primary tumor and the histological grade. Although histologically this tumour is relatively bland (low-grade), it does carry a significant risk of recurrence and quite often progresses to a higher grade lesion. The local recurrence rate of the low-grade type is as high (50-60%) as that of the high-grade type, with an overall risk of approximately 20% to 25% of metastasis;^{17,18}

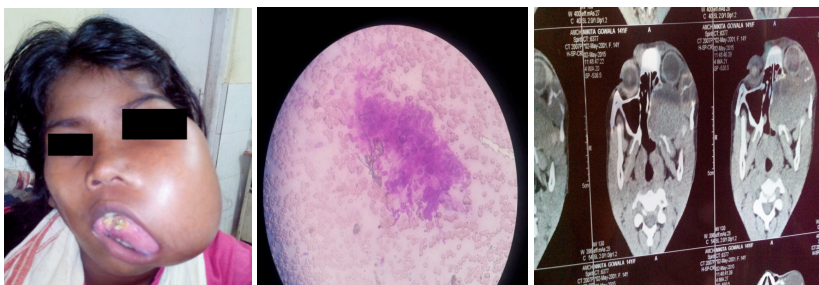


Figure-1: Fibromyxoid sarcoma of oral cavity;
Figure-2: HPE showing fibromyxoid sarcoma;
Figure-3: NECT and CECT scan of faciomaxillary region

thus, it is obvious that wide surgical treatment is necessary to suppress the risk of local recurrence and the risk of histological progression.¹⁹

CONCLUSION

This rare tumour should always be differentiated from other tumours of same lineage and should be soon operated and post-operative radiotherapy be given. We have tried to present a neglected case of fibromyxoid sarcoma and also given our view for its differentiation and treatment.

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