

Case Report

Aggressive Fibromatosis versus Low Grade Fibrosarcoma - a Diagnostic Dilemma

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Abstract

The fibromatosis are a group of lesions that can infiltrate widely, replacing muscle, fat and bone with fibrous tissue of varying cellularity. They do not develop distant metastases, however, locally they show an aggressive and infiltrative behaviour. The major challenge in the diagnosis lies in not over or under diagnose of fibrosarcoma and an underdiagnosis of reactive fibrosis. But this histologic differentiation is necessary owing to different treatment strategies.

Keywords: Aggressive Fibromatosis, Fibrosarcoma.

Introduction

Fibrous tumors and tumor like lesions form a heterogeneous group of distinct entities differing in biologic behaviour but being histologically very similar and thus presenting considerable difficulty in pathologic diagnosis. Examination of various classifications reveal many diverse lesions with confusing and overlapping nomenclature.¹

The term fibromatosis refers to a group of fibrous tumors or tumor like lesions of soft tissues that share similar microscopic characteristics and possess an intermediate biologic potential between benign and malignant lesions.² Fibromatosis are non metastasizing but may exhibit both rapid growth and visceral involvement. Spontaneous regression has been described but rare tumors mimic a malignancy in their tendency to occur locally.³

Pathologically, fibromatosis has a deceptively bland appearance. It is however associated with an infiltrative growth pattern that results in difficulty in complete excision and propensity for recurrence.⁴

Although, clinical features would also be useful in the categorization of these lesions as benign or malignant, the mentioned literature, among the jaw

lesions, today, suggests that the reliability of clinical parameters would not be suitable for such connective tissue neoplasms.⁵

Here, we report a case of fibromatosis of the maxilla. The histopathological diversity in all the areas was analyzed in great detail and an attempt was made to separate the lesion from other lesions of same origin on the basis of definite histopathological parameters.

Case Report

A 21 year old male reported to the Department of Oral Pathology, I.T.S – C.D.S.R, with the chief complaint of swelling and pain on left side of the face since one month. Past history revealed that the pain subsided on intake of medicines prescribed by a local practitioner but the swelling did not subside. Patient had a history of pus discharge for the last 2 days. Intraoral examination revealed swelling of size 3 x 3 cm in the left anterior maxillary region with ill defined borders, pink color and soft consistency. No lymph node involvement was observed. On radiological examination, an osteolytic lesion with irregular margins was seen in the upper occlusal radiograph. The CT scan revealed soft tissue mass perforating anterior wall of maxilla and palate. The provisional diagnosis was of Central Giant Cell Granuloma.

An incisional biopsy was performed and gross examination showed four brownish tissue bits with one larger bit measuring 3 x 1.5 cm and three equal approximately equal bits measuring 1x 1 cm. Microscopic examination revealed a connective tissue proliferation of predominantly fibroblastic cells in a background of moderate amount of collagen tissue.

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The overall pattern was generally ranging from sheets, storiform pattern and to a certain extent, in a fascicular pattern (Figure I).

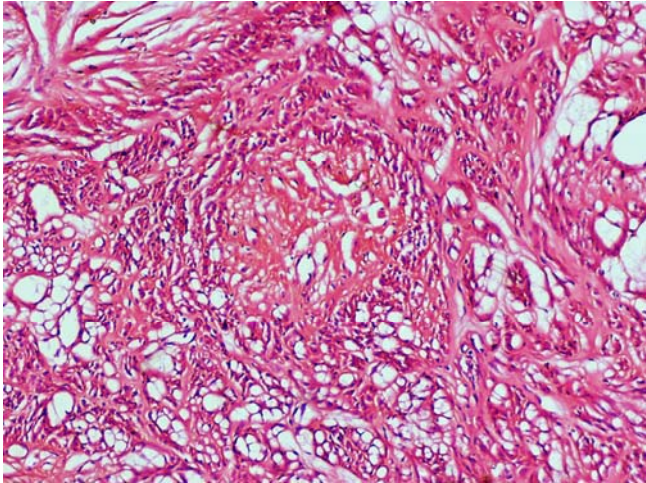


Figure I: Photomicrograph showing fasciculated arrangement of cells (H/E X 10)

The cells were large, oval and fibroblastic in nature but pleomorphism was abundant. Pleomorphic cells(Figure II showed) a range from elongated flattened

nuclei to a large giant cell like elongated nuclei. Cells showed moderate to abundant cytoplasm. A few areas of the section were predominantly spindle and wavy in nature(Figure III).

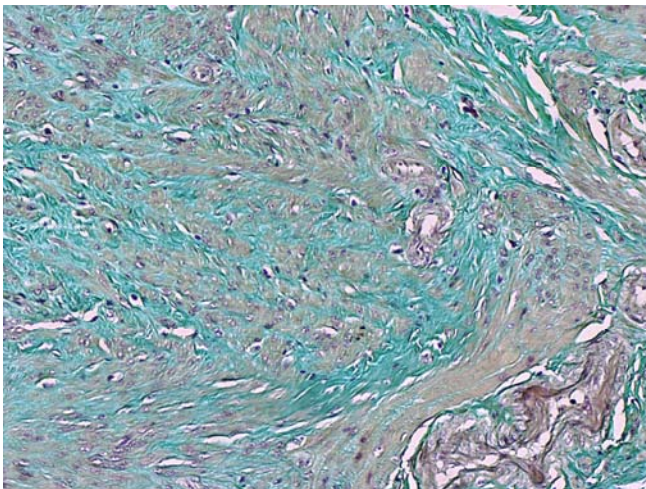


Figure II: Photomicrograph showing cellular and nuclear pleomorphism (H/E X 40)

The blood vessels seemed to be constricted due to the high proliferative nature of the cells around. Nucleus of the tumor cells showed a clear nuclear membrane but in some areas, condensation of the

nuclear material and 2 or more nucleoli were seen. Mitotic figures were present but were not seen uniformly throughout the section (Figure IV). The final diagnosis given was that of Aggressive fibromatosis.

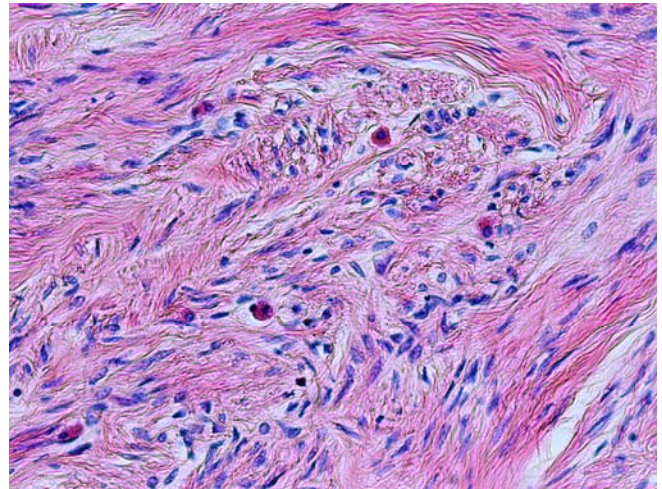


Figure III: Photomicrograph showing bland spindle shaped cells (H/E X 40)

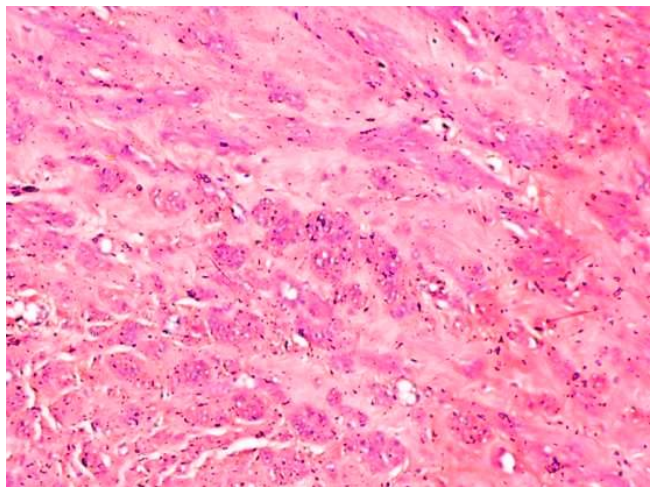


Figure IV: Photomicrograph showing two typical mitotic figures visible in the centre (H/E X 40)

Discussion

The fibromatosis constitute part of a spectrum of poorly understood proliferative lesions whose histologic features overlap to such an extent that the pathologist may be more influenced by the anatomic location of the lesion, sex and clinical behavior than by the histologic appearance in rendering his or her diagnosis. Wherever they occur, the diagnosis and management of fibromatosis are always source of concern.¹

It has been defined as a non neoplastic spindle cell proliferation of childhood which may be locally aggressive but has no metastatic potential. The natural history is of initial rapid growth and local aggression.³ Enzinger and Weiss divided the fibromatosis into 2 broad categories: superficial and deep. The fibromatosis that occur in the head and neck including those that involve oral and paraoral structures are considered under the heading of extraabdominal fibromatosis. Infantile fibromatosis is the childhood counterpart of extraabdominal fibromatosis.²

The histopathologic differentiation between aggressive fibromatosis and other closely related spindle cell lesions like fibrosarcoma, neurofibroma, nodular fasciitis, fibrous histiocytoma and infantile myofibromatosis is a challenge to the pathologist as it requires expertise to differentiate the finer details. But the major challenge in dealing with the lesions of fibromatosis is to avoid an overdiagnosis of fibrosarcoma and an underdiagnosis of reactive fibrosis. Fibromatosis has a more uniform growth pattern, more mature cells and a paucity of mitosis compared with fibrosarcoma. Reactive fibrosis such as that following injury or trauma has a more variable growth pattern than fibromatosis and may show areas of focal hemorrhages or hemosiderin deposition.²

The grade I fibrosarcoma is usually discernable from fibromatosis by the presence of occasional larger nuclei with ominous chromatin clumping, greater cellularity, greater mitotic activity and thin rather than thick collagen bundles.¹ Immunohistochemistry is of little help in differential diagnosis because positive immunostaining against vimentin can be observed in all fibrous connective tissue tumors.⁶ Mitotic figures are rare and the finding of more than one mitotic figure per high power field or atypical mitotic figures should raise the suspicion of fibrosarcoma. Since on rare occasions, features of fibromatosis and fibrosarcoma are found together in the same neoplasm, careful sampling of the tumor is mandatory for a reliable diagnosis. Clinical considerations are of little help in distinction of fibromatosis and fibrosarcoma because both tumors may occur at the same location and in the same age group. Also, it is notoriously difficult to separate fibromatosis from well differentiated fibrosarcomas especially in infants and juveniles when fibromatosis is characterized by higher mitotic rates than in adults. Indeed, doubts have been expressed to whether this distinction can be made at all.¹

The storiform pleomorphic variant of malignant fibrous histiocytoma also shows frequent transitions from storiform to pleomorphic pattern. In its classic form, a lesion of malignant fibrous histiocytoma consists of plump spindle cells arranged in short

fascicles in a cartwheel or storiform pattern around slit like vessels. These differ from other similar lesions by the presence of occasional plump histiocytic cells, numerous typical and atypical mitotic figures and secondary elements including xanthoma cells and modest number of chronic inflammatory cells. Another characteristic feature is the presence of large number of giant cells with multiple hyperchromatic irregular nuclei.⁷

Authors have propounded that since soft tissue and intraosseous lesions are histologically indistinguishable and since in the maxilla and mandible, origin in bone or soft tissue is uncertain, the term Desmoplastic should not be used in the area of head and neck but all the lesions should be termed desmoid fibromatosis.¹

A small panel of antibodies to include S- 100 protein, smooth muscle actin, desmin and vimentin would in most cases help in establishing the diagnosis. Fibromatosis is generally positive for vimentin. However, it should be pointed out that immunohistochemical studies have shown myofibroblastic differentiation in some cases of fibromatosis. Thus, in such cases SMA would also be positive along with vimentin. A case of fibrous histiocytoma is positive for vimentin and can be variably positive for actin.² Histopathologically, our differential diagnosis narrowed down to Aggressive fibromatosis and low grade fibrosarcoma. So in our case immunohistochemical aid was not utilized since vimentin is positive for both the lesions.

The final diagnosis of aggressive fibromatosis was based on a number of factors which included spindle shaped monotonous population of fibroblasts arranged in a whorl like and fasciculated pattern and the presence of collagen. The cells were mature in appearance and the presence of few typical mitotic figures was noticed.

Conclusion

There is a very fine distinction between fibromatosis and a low grade fibrosarcoma and careful microscopic examination with accurate sampling is required to render the correct diagnosis. An accurate diagnosis is imperative since it changes the line of treatment drastically.

References

1. Vally IM, Altini M. Fibromatosis of the oral and paraoral soft tissue and jaws- Review of literature and report of 12 new cases. *Oral Surg Oral Med Oral Pathol* 1990; 69: 191 -98.

2. Fowler CB, Hartman KS, Brannon RB. Fibromatosis of the oral and paraoral region. *Oral Surg Oral Med Oral Pathol* 1994; 77: 373 -86.
3. Baerg J, Murphy JJ, Magee JF. Fibromatosis: Clinical and pathological features suggestive of recurrence. *J Pediatr Surg* 1999; 34: 1112-44.
4. Tse GM, Chan KF, Ahuja AT, King AD, Pang PCW, To EWH. Fibromatosis of the head and neck region. *Otolaryngol Head Neck Surg* 2001; 125: 516 - 19.
5. Watzinger F, Turhani D, Wutzl A, Fock N, Sinko K, Sulzbacher I. Aggressive fibromatosis of the mandible: A case report. *Int. J. Oral Maxillofac Surg* 2005; 34: 211-13.
6. Seper L, Burger H, Vormoor J, Joos U, Kleinheinz J. Aggressive fibromatosis involving the mandible - Case Report and review of literature. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 2005; 99: 30-8.
7. Enzinger FM, Weiss SW. Malignant fibrous Histiocytic Tumors. In, *Soft tissue tumors, 2nd edition*. New Delhi, B.I Publication Pvt. Ltd, 1988, 357-59.

Corrigendum

Due to miscommunication the article titled” **Is quality of sputum sample a major determinant in the overall positivity of AFB smears?**“ by Shamim Mumtaz Col Abdul Bari, Ahsan Ahmad Alvi, Hameeda Akhtar and Sajid Hussain Shah which was published in another journal as well was published in IJP 7 (2) pp 80-83. The article should be deemed unpublished in IJP and therefore not included in IJP website and author and subject indices. The authors have extensively apologized for this miscommunication for this mistake. The IJP editorial board regrets this mishap.