

GIANT CELL FIBROBLASTOMA - A RARE SOFT TISSUE TUMOR IN AN ELDERLY FEMALE

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ABSTRACT:

Giant cell fibroblastoma (GCF) is a rare soft tissue tumor of intermediate group occurring most commonly in children. Unusually it has been known to occur in adults and very rarely to involve patients older than 60 years of age. We report a rare case of giant cell fibroblastoma in a 72 year old female patient. Pathologist need to be aware of the occurrence of this rare entity in advanced age and its morphology in order not to mistake it for a high grade sarcoma.

Keywords: *Giant cell, fibroblastoma, sarcoma*

INTRODUCTION

Giant cell fibroblastoma is a rare soft tissue tumor of fibroblastic origin. The earliest description dates back to 1983, when Schmooker and Enzinger first described 20 cases of GCF in their formative publication.^[1] Closely related to dermatofibrosarcoma protuberance, both share common demography, biology and molecular profile. Though morphology of the two tumors can overlap in primary tumor and in recurrences as well, nevertheless GCF has a rather distinct histomorphology.^[2,3] GCF shows characteristic pseudovascular spaces which are lined by spindle cells and giant cells some of which are atypical. The atypia is a major caveat and leads to the mistaken diagnosis of a high grade sarcoma.

GCF is a childhood tumor and rarely does it occur in adults. There are very few reported cases of GCF in adults. In a large series of 86 cases of GCF there were only 10 cases occurring above fourth decade of life.^[4]

Herein we report a rare soft tissue tumor occurring at an unusually advanced age, with a brief review of the histomorphology.

CASE REPORT

A 72 year old female presented with a swelling over the lateral aspect of the thigh of three months duration. There was no history of pain or rapid increase in the size of swelling. There was no history of fever, loss of appetite or weight loss. Rest of the clinical and personal history was

unremarkable. On local examination the swelling was 3×3×3cm, well circumscribed, soft to firm, present in the subcutaneous plane. It was non tender and the overlying skin was unremarkable. Inguinal lymph nodes were not palpable. Patient was a non-hypertensive and non-diabetic. A clinical diagnosis of soft tissue sarcoma was provisionally rendered. A wide surgical excision was performed.

Gross: Specimen consisted of a grey brown soft tissue measuring 9×4×3cm. Cut surface yielded a well circumscribed tumor measuring 3×3 cm. It was solid, firm, grey yellow, mucoid lesion.

Histopathology: Sections studied showed a well circumscribed tumor comprising of tumor cells arranged in short fascicles as well as in non-descript patterns. Individual cells were spindle to polygonal with moderate eosinophilic to foamy cytoplasm. Nuclei showed modest degree of pleomorphism with vesicular chromatin and prominent nucleoli. Plenty of vascular channels were seen lined by endothelial cells. Also seen were pseudo vascular spaces which were focally lined by giant atypical cells. Many multinucleate giant cells were seen scattered throughout the tumor and also lining the pseudo vascular spaces. Extensive areas of hyalinization were noted. There was no evidence of mitosis, necrosis or infiltration into the surrounding tissue. Final diagnosis of giant cell fibroblastoma was made. At immunohistochemistry there was diffuse positivity for vimentin and CD 34.

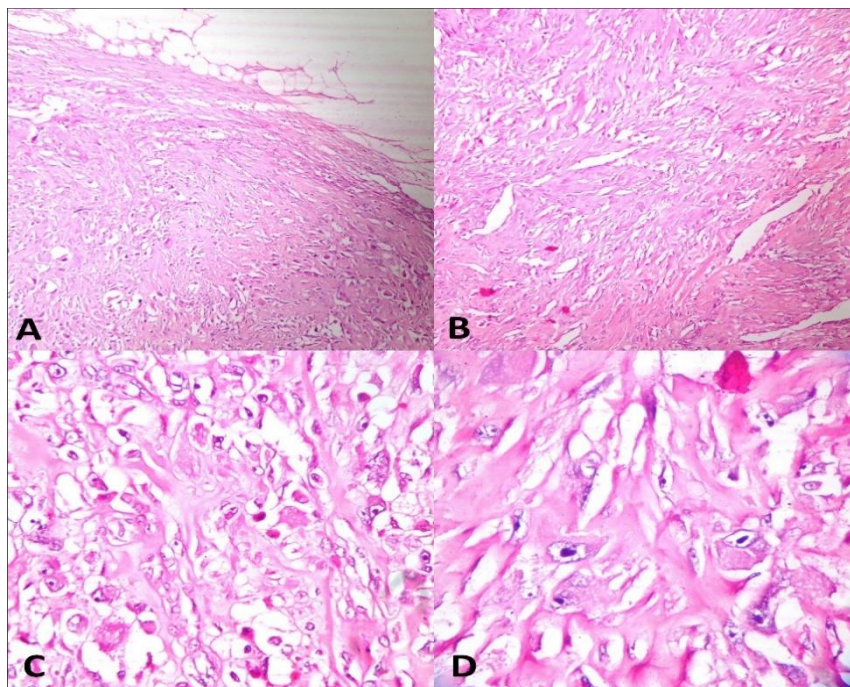


Fig. 1: (A) Section shows tumor which is well circumscribed. [H & E, x 40]
 (B) Section showing tumor cells arranged in short fascicles as well as in non-descript patterns. [H & E, x 40]
 (C) Section depicts tumor cells with modest degree of pleomorphism in the nuclei with vesicular chromatin and prominent nucleoli. [H & E, x 100]
 (D) High power view of Figure 1 C. [H & E, x 400]

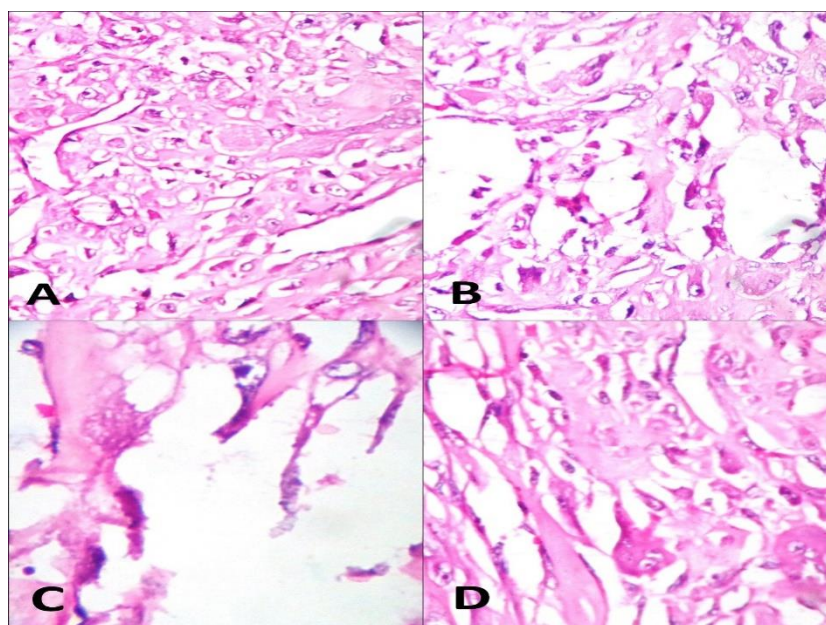


Fig. 2: (A) Section depicting vascular spaces. [H & E, x 100],
 (B) Section depicting pseudo vascular spaces. [H & E, x 100],
 (C) Section depicting pseudo vascular spaces lined by atypical nuclei. [H & E, x 400],
 (D) Section shows atypical nuclei and multinucleate giant cells. [H & E, x 400]

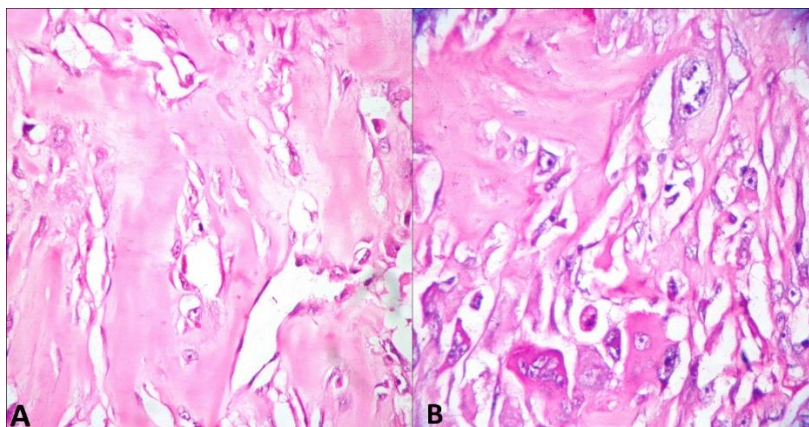


Fig. 3: (A) Section shows hyalinized areas. [H & E, x 100], (B) Section showing atypical and multinucleate giant cells. [H & E, x 400]

DISCUSSION

GCF is a rare fibrohistiocytic tumor of intermediate group. Schموoker and Enzinger are credited with the first account of this entity in 1983.^[1] Ever since then, this tumor has been proposed/considered as juvenile counterpart of dermatofibrosarcoma protuberance.

Literature review depicts these two tumors as different expressions of same neoplasm. Points embracing this notion include - common site of occurrence, superficial location and focal areas resembling DFSP appearing in GCF and vice versa. In addition even recurrences of each may show histology of the other tumoral counterpart. Testimony to this connotation is privileged by the occurrence of similar chromosomal abnormalities i.e., t(17,22) q(22,q13) translocation in both the tumors.^[3] In spite of all these associations, the fact that recognizing GCF as a separate entity holds importance in view that it is easily mistaken for a high grade sarcoma in 40% of the cases.

The characteristic histology of GCF consist of spindle cells and giant cells arranged in a non-descript pattern intermixed with areas of hyalinized and occasionally myxoid stroma. The clinching/hallmark features are the presence of angiectoid / pseudo vascular spaces lined by atypical or giant cells. The giant cells in the GCF are basically derived from the spindle cells a fact substantiated by molecular cytogenetic studies which demonstrated similar fusion gene COL1A1-PDGFB in both the components and by electron microscopy.^[2,5] Mitosis may be present but can be dismissed if all the clinching features are present. The pleomorphism and atypia are implicated caveat in misdiagnosis of a high grade sarcoma. In a series of 28 cases of GCF from AFIP, 14 cases were erroneously diagnosed as sarcoma.^[6] The common sarcomas with which GCF has been mistaken include myxoid liposarcoma, myxoid malignant fibrous histiocytoma and myxofibro sarcoma. Nevertheless, absence of

plexiform chicken wire blood vessels and characteristic lipoblast facilitate rule out a liposarcoma. Likewise inflammatory component, xanthoma cells, touton giant cells and storiform pattern are seen in myxoid MFH and not in GCF. It is difficult to distinguish on morphology low grade myxofibrosarcoma from GCF. Even at immunohistochemistry both the tumors demonstrate CD 34 positivity.^[7] However in all the above conditions, the distinctive pseudo vascular spaces, multinucleate giant cells and atypical spindle cells lying scattered in the stroma and lining the angiectoid spaces, seen in GCF, are absent. Occasionally pseudo vascular spaces are scanty requiring complete embedding of the tissue. Also there may be close association of tumor cells with lymphocytes simulating emperipolesis.^[7] Rarely GCF can demonstrate myoid whorls, perivascular lymphocyte cuffing and intralesional hemorrhage.^[4]

Clinically the tumor tend to occur in childhood with the median age being 6 years. Rarely does it occur in adults. In a series of 86 new cases of GCF only 10 cases were older than 40 years. There are only 14 reported cases of GCF in adults, out of which 10 occurring over fourth decade and 4 older than 55 years of age, as reported in a review of all the GCF cases diagnosed from the files of AFIP till 2007.^[4] Later in 2008, another case of GCF in a 62 year old male was reported. GCF predominantly occurs in males.^[7] Common sites of predilection are back of thigh, inguinal region, chest wall and extremities. An unusual site includes vulva, where a GCF arose at a site of previous fibro epithelial polyp.^[8] On gross it demonstrates a grey yellow mucoid appearance as was seen in our case.

50% of GCF recur commanding wide local excision as treatment of choice. Any type of conservative excision demands conscientious follow up. In their series of seven cases, Dymock et al reported low recurrence rate probably due to the short

follow up period.^[5] Till date no report of metastasis exists.^[9]

CONCLUSION

Though a childhood tumor, GCF can occur in adults. Awareness of the occurrence of this rare entity, its unusual presentation at an advanced age and familiarity with the morphology is mandatory for a Pathologist and surgeon for accurate diagnosis and planning proper management.

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