Case Reports

Giant Cell Fibroblastoma of Hand in an Adult: An Uncommon Presentation

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Giant cell fibroblastoma is a rare intermediate-grade fibrohistiocytic tumor of childhood and even rarer in adults. It is seen in the first decade of life. It is known for its recurrence, but its metastasis has never been reported. Giant cell fibroblastoma requires attention as it was previously misdiagnosed as sarcoma. So, histology is mandatory and remains the gold standard for its diagnosis. We present a case of 21-year-old female who presented with complaints of a painless lesion over the dorsal aspect of left hand.

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Introduction

Giant cell fibroblastoma is an unusual fibrohistiocytic tumor, first described by Schmookler and Enzinger in 1982.[1] It is usually seen in males during the first decade of life.[2] Giant cell fibroblastoma requires attention as it is uncommon in adults and known for its recurrence.[3] So we are presenting a case of adult female who presented with a painless lesion over the dorsal aspect of left hand.

Case Presentation

21 year old adult female presented to the surgical OPD with complaints of painless swelling over the dorsal aspect of the left hand since 2 years. There was no history of trauma in the past. The lesion was not associated with fever and anorexia. A soft pin head size reddish-coloured lesion measuring 1 x 0.7 x 0.3 cm was seen on local examination. The overlying skin didn't show any ulceration. The surrounding skin was normal. A clinical diagnosis of sclerosing haemangioma was made. Routine hematological workups were within normal limits. No radiological workup and cytology were performed, as it was a superficial tiny lesion. The excision was performed under local anesthesia and excised tissue was sent for histology to the department of pathology.

Grossly a single greyish-white skin-covered tissue measuring 0.7 x 0.5 x 0.2 cm was received. On cut it appears whitish. Microscopically, the section showed a hyperkeratotic, hyperplastic epidermis (Fig. 1a) with underlying dermis showing fibroblast and floret-like giant cells arranged in vague fascicles on a collagenous stroma (Fig. 1b). Cells are elongated and stellate shaped having hyperchromatic nuclei with scant eosinophilic cytoplasm. The giant cells contain multiple round to oval-shaped hyperchromatic irregular nuclei with abundant amphophilic cytoplasm. Also seen are pseudo vascular spaces lined by fibroblastic cells (Fig 1c). No evidence of necrosis and mitosis were seen. Immunohistochemistry showed strong Vimentin positivity and CD34 negativity. The patient is on regular follow up.

Discussion

Giant cell fibroblastoma was first described by Shmookler and Enzinger in 1982.[1] It clinically presents as a small, slow-growing, painless swelling involving back, thigh and trunk.[3] Other uncommon sites include the shoulder, perineum and extremities.[4,5] It predominantly affects infants and children and is rarely seen in adults aged range 6mths to 62 years.[6] Grossly it is an encapsulated greyish-white gelatinous lesion found under the skin in the dermis. Histology shows variable cellularity with fibro myxoid stroma containing bland spindled and

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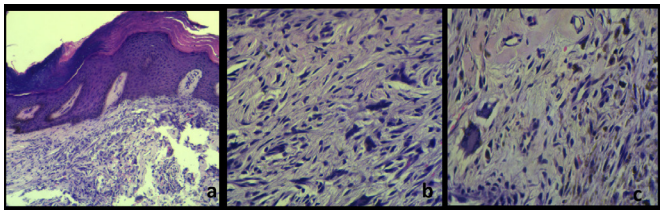

Figure 1: Light microscopy in H and E stain a: Hyperkeratotic hyperplastic epidermis, x200 B: Fibroblast and floret like giant cells, x400 C: Pseudovascular spaces lined by fibroblast and giant cells, x400

multinucleated floret like giant cells. These spindle cells are mildly to moderately pleomorphic and are arranged diffusely or in vague fascicles. These spindle cells have vesicular to hyperchromatic nuclei and scant eosinophilic cytoplasm. The giant cells contain a variable number of round to oval-shaped vesicular nuclei with abundant amphophilic cytoplasm and irregular cytoplasmic contours. Slit like sinusoidal spaces are characteristic and are lined by multinucleated giant and tumor cells.[1,7]

Previously, giant cell fibroblastoma was considered a juvenile variant of dermato fibrosarcoma portubrens. However, recent clinical, morphological, immunophenotypical and molecular studies confirm that these two entities are of the same spectrum.[1,6] Morphologically, both giant cell fibroblastoma and dermatofibrosarcoma protuberant are dermal or subcutaneous in origin, but rarely involve superficial skeletal muscle. Both show honeycomb and parallel growth patterns with uninvolved adnexa, myxoid changes and prominent vasculature.[6]

Immunohistochemistry of both lesions shows negative reactions for keratin, S-100, HMB-45, smooth muscle actin and desmin. Cytogenetically, t(17;22) translocation are seen in both giant cell fibroblastoma and dermatofibrosarcoma protuberans.[6] Although both giant cell fibroblastoma and dermatofibrosarcoma protuberans show similar clinical and morphological features, but histopathology examination still remains the gold standard for differentiating them. The presence of pseudovascular spaces lined by giant cells, solid areas with stromal giant cells, consistent hemorrhage, perivascular and onion skin-like chronic inflammation without storiform pattern favors the diagnosis of giant cell fibroblastoma.[6]

Vimentin remains the consistent immunohistochemical marker for giant cell fibroblastoma.[7] Wide local excision remains the treatment of choice and these patients do not require any form of chemotherapy.[8]

The differential diagnoses of giant cell fibroblastoma includes liposarcoma, myxofibrosarcoma, malignant fibrous histiocytoma and papillary intralymphatic angioendothelioma. Liposarcoma shows the presence of lipoblast, absence of slit-like sinusoidal spaces, and occurrence of this lesion in adulthood. Myxofibrosarcoma are seen in deeper location and occurrence in older patients. Malignant fibrous histiocytoma is characterized by markedly atypical mesenchymal cells, mitotic figures and absence of sinusoidal spaces. Papillary intralymphatic angioendothelioma can be differentiated by positive vascular markers and absence of myxoid and cellular areas in this tumor.[1,6]

However, our case was of adult female with painless lesion over the hand and showing similar morphological features of giant cell fibroblastoma with immunohistochemical positivity of Vimentin and negativity of CD34.

Conclusion

Giant cell fibroblastoma remains a rarity in adults, very few cases have been reported in females and histopathology examination along with immunohistochemistry remains the gold standard for its diagnosis.

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None

Conflict of interest

None

Ethical Approval

all procedures were in accordance with the ethical standards at the authors institution and with the 1964, Helsinki declaration and it s later amendments or comparable ethical standards.

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