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Case Report

Dermatofibrosarcoma Protuberans- A Case Report

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Abstract

Dermatofibrosarcoma Protuberans (DFSP) is an uncommon soft tissue tumor that involves the dermis, subcutaneous fat, and in rare cases, muscle and fascia. It represents less than 0.1% of all tumors, but is considered to be the most common skin sarcoma. We present one such case where histopathology and immunohistochemistry helped clinch the diagnosis .DFSP has a high rate of recurrence but a low rate of metastasis. Wide surgical excision is the cornerstone of treatment for DFSP.

Keywords: Fibrosarcoma, protuberans, storiform, sarcomatous, dermis, spindle shape, recurrence.

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INTRODUCTION

Dermatofibrosarcoma Protuberans is a superficial, low-grade, locally aggressive, spindle, fibroblastic, neoplastic lesion which accounts for ~1% of all soft tissue sarcomas (Paramythiotis D *et al.*,). It is commonly found on the trunk (42%-72%), however, it can also develop in the extremities, head or neck (Amjadzadeh M *et al.*,). Slow growth and low aggressiveness are its main characteristics. Its incidence is 0.8-4.1 per million people annually and the disease is seen more in women than in men in the age group being 20-50 years (Amjadzadeh M *et al.*,).

CASE REPORT/MATERIAL AND METHOD

A 42 year old female presented with complaints of a lump in the right thigh for 3 years and pus discharge from it for 15 days. On Examination, the lump was seen on the medial aspect of the right thigh. It had a broad base with denuded ulcerated skin and there was a pus discharge seen from the center of the lesion. CT scan showed a large well defined heterogeneous enhancing exophytic mass lesion in the upper thigh skin. Total excision of the lump was performed with a good aesthetic result.

Grossly we received a soft tissue mass measuring 6 * 5.5 * 4.5 cm. The external Surface was covered with skin on the medial side which shows

ulceration. Deep resected margin shows adipose tissue. Cut Surface showed a large gray-white tumor with specs of hemorrhage measuring 5 *5.5*4 cm.

Microscopy showed epidermis with ulceration and granulation tissue and a diffusely infiltrating tumor centered on lower dermis extending into subcutis. Tumor cells were arranged in a storiform pattern and were composed of uniform spindle tumor cells containing plump to elongated wavy nuclei. The collagenous stroma contained small blood vessels .Deep resected margin was free from tumor. Circumferential margin was free from tumor. Based on the above histopathological features, а diagnosis of Dermatofibrosarcoma Protuberans was made. Dermal Fibrous Histiocytoma, Desmoplastic Melanoma and Adult fibrosarcoma were close differential diagnosis. Immunohistochemistry was done and the final diagnosis panel showed positivity for CD34 (Immunoreactive, score 4+in neoplastic cells) and were negative or non-immunoreactive for desmin, SMA, S 100. Based on the clinical features, microscopic finding (spindle shaped cells arranged in storiform pattern with no mitosis and necrosis) and immunohistochemistry positivity), a diagnosis findings (CD 34 of Dermatofibrosarcoma Protuberans was made and confirmed.

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Figure 1: External surface covered with ulcerated skin on the medial side



Figure 2: Cut surface showed a gray-white tumor with specs of hemorrhage and calcification



Figure 3: H&E, 10X cells are arranged in storiform pattern



Figure 4: H&E, 40 X displaying spindle shape tumor cells having plump nuclei

DISCUSSION

Dermatofibrosarcoma protuberans (DFSP) is a superficial fibroblastic neoplasm with storiform architecture. It usually presents as a painless, firm slowgrowing plaque or nodule. (Massi D et al.,). With time, the tumor evolves into multiple "protuberant" nodules that infiltrate the subcutaneous tissue, fascia, muscles and even bone (Amjadzadeh M et al.,) In early stages; it should be differentiated from other entities clinically like lipoma, epidermal cysts, keloids, dermato-fibroma, and nodular fasciitis. In the later stages, the differential diagnosis should be pyogenic granuloma, Kaposi sarcoma, and other soft tissue sarcomas (Paramythiotis D et al.,). On ultrasound, DFSPs have been found to be mostly hypoechoic or mixed hyperechoic, with mostly well-defined margins or irregular, with projections similar to pseudopodia. Histological examination is the definitive diagnostic method. (Paramythiotis D et al.,).

Microscopically, tumor is located in the dermis, but it can sometimes show infiltrative growth in the subcutaneous fatty tissue, forming a pastry pattern (60% of cases; neoplastic cell bands parallel to the epidermis) or a honeycomb pattern (delimitation of adipocyte islets between the tumoral tissues). Atypia is minimal, and mitoses are rare (Chan I L et al) Mitotic count. necrosis, and areas of fibrosarcomatous change should be mentioned in the report as they have been shown to be correlated with aggressive clinical behavior and lower overall survival (Paramythiotis D et al.,). In addition to the classical form characterized by a storiform pattern of tumor cells, the pigmented (Bednar's tumor), myxoid types, and DFSP with sarcomatous areas can be observed (Paramythiotis D et al.,). The Differential Diagnosis for DFSP includes Dermal Fibrous Histocytoma which is usually <1cm with haphazard arrangement and CD 34 negative whereas in our case it was more than 6 cm with storiform arrangement and CD 34 was positive, second differential was desmoplastic melanoma but they lack the storiform pattern, the other differential was adult fibrosarcoma which was differentiated from DFSP as they were CD 34 negative (Amjadzadeh M et al.,).

The characteristic cytogenetic features of DFSP are a supernumerary ring chromosome and a reciprocal chromosomal translocation t(17;22)(q22;q13), causing a fusion of the plateletderived growth factor β -chain (*PDGFB*) gene at 22q13 and the collagen type 1 α 1 (*COL1A1*) at 17q22 which results in the activation of the PDGF. *PDGFB* copy number status may become a useful diagnostic marker since the gene is a potential target of treatment in patients with DFSP (Thway K *et al.*,). In our case the patient was advised molecular studies, but as he didn't return for follow up, his reports could not be retrieved. The standard treatment is wide local resection with at least a 2-cm margin (Paramythiotis D *et al.*,).

CONCLUSION

Thus Dermatofibrosarcoma protuberans is a borderline tumor which can progress to malignant fibrous histocytoma. As it is locally aggressive and has a low rate of metastasis, histopathology and immunohistochemistry can help in early detection and guide appropriate management.

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