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Case Report

Anterior abdominal wall dermatofibrosarcoma protuberans: A case report

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Abstract

Introduction: Dermatofibrosarcoma protuberans is a rare low-grade fibrohistiocytic tumor ,common in age group of 20 to 50 years. Histologically, it is diagnosed on the basis of a spindle-cell tumor arranged in small bundles in a characteristic cartwheel pattern. Though local recurrence rate is high, the treatment of choice is surgical resection with negative margins. We describe a case of a 34-year-old lady with dermatofibrosarcoma protuberans.

Key words: dermatofibrosarcoma protuberans, soft tissue neoplasm.

Introduction

Dermatofibrosarcoma protuberans (DFSP) is a rare soft tissue neoplasm with low-grade malignancy. It is a low-grade fibrohistiocytic tumor characterized by a high rate of local recurrence although metastasis is rarely seen.It commonly occurs between the second and fifth decades of life and affects all races. [1, 2]

We report a case of a 34-year-old lady with DFSP, which was managed surgically.

Case Report

A thirty-four year lady presented with supra-umbilical painless mass. On examination, it was 3.5 cm by 2 cm in size, firm, fixed to abdominal wall. There was no history of discharge from the lesion, fever and significant weight loss. She had history of excision done for same lesion three months before with provisional diagnosis of sebaceous cyst, which recurred. Histopathology report revealed features suggestive of Abdominal Fibromatoses. The chances of local recurrences being high, wide excision was advised and a close follow up of patient were also recommended.

After six weeks of excision, patient presented with hypertrophic scar around previous incision site. Again, excision with wide margin was done and histopathology examination showed features suggestive of dermatofibrosarcoma protuberans or Cellular benign fibrous histocytoma and Immunohistochemistry of CD34 was advised. Report of CD34 was positive which confirmed Dermatofibrosarcoma protuberans. Patient did well during post-operative period with no recurrence after six months.

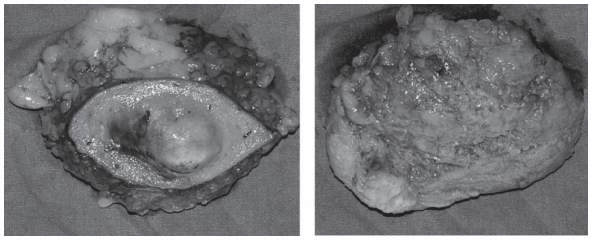

Figure 1: Specimen after resection

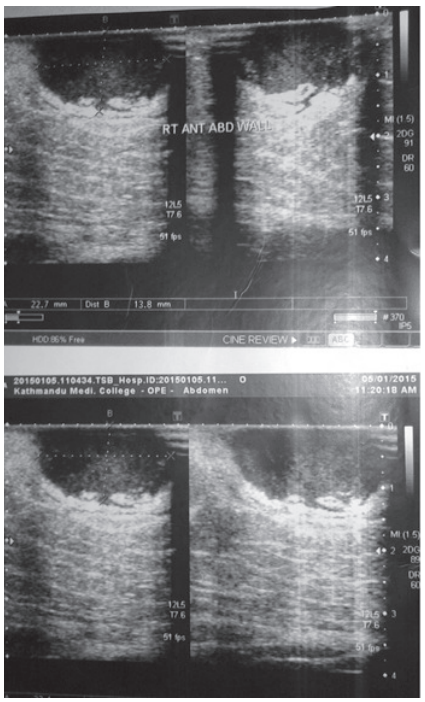

Figure 2:USG Showing the lesion regarding its depth

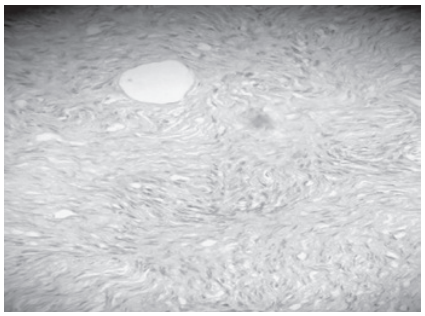

Figure 3: Histological slide showing monomorphic and mitotic activity of the cells

Discussion

Darier and Ferrand first described this condition in 1924 with the name "progressive and recurrent dermatofibroma." Later, Hoffman proposed this rare tumor as the term dermatofibrosarcoma protuberans.[3] It is a low-grade fibrohistiocytic tumor. This condition presents between 20 and 50 years of age usually over the trunk.[2]

This condition is rare with incidence of 0.8 cases per million people per year. [4]

Most common soft tissue tumor of abdominal wall is the desmoid tumor and the least frequent is Dermatofibrosarcoma Protuberans (DFSP).[5]It is locally infiltrative and metastasis rarely occurs.[4]

DFSP is diagnosed based on histology in which tumor cell has characteristic feature spindle-shaped with an elongated nucleus and are arranged in short bundles. The pattern is one of infiltrative growth that generally extends to the deep.[6]

This lesion should be differentiated from dermatofibroma, keloid and hypertrophic scar, malignant melanoma, epidermal inclusion cyst and metastatic carcinoma of skin. Exclusion can be done with histopathology of excised mass and immunohistochemistry. [8]

For confirmation of DFSP, Immunohistochemical studies are highly sensitive. Immunohistochemical marker- CD34 is used to differentiate DFSP tumour cells from normal stromal cells and dermatofibroma. In our case, DFSP was diagnosed on the basis of histology and confirmation on immunohistochemistry, as the tumour cells were diffusely immunoreactive for CD34.[3]

Treatment of this condition is wide resection of the tumor with a 2-3 cm margin. In the case of incomplete resections or large tumors radiotherapy is reserved as adjuvant therapy. [9, 10]

The rate of recurrence to less than 2% can be achieved with Mohs micrographic surgery and is considered the treatment of choice.[11]

Metastases can occur in DFSP but is extremely rare with very poor prognosis.In every 3 to 6 months, clinical follow-up is recommended during the first 3 years following surgery and every year thereafter.[1,6]

Conclusion

DFSP has potential for aggressive local invasion and the high rate of recurrence. Diagnosis should be made early and plan for surgical resection with wide margin.

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