



RECURRENT GIANT DERMATOFIBROSARCOMA PROTUBERANCE OF ANTERIOR ABDOMINAL WALL: A CASE REPORT

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ABSTRACT

Dermatofibrosarcoma protuberans is an uncommon, locally aggressive, cutaneous, soft tissue sarcoma of dermis. It is most commonly seen on the trunk and it frequently recurs locally after an incomplete excision, but distant metastasis is rare. Patients need post-operative local irradiation even with histopathological clear margins, following wide excision, owing to its high affinity for local recurrence. Here, we are reporting a case of giant recurrent dermatofibrosarcoma protuberans of the lower part of the abdomen in a 49 years female.

KEYWORDS: Dermatofibrosarcoma protuberans, Abdominal wall, Recurrence.

INTRODUCTION

Dermatofibrosarcoma protuberant (DFSP) is a rare locally aggressive mesenchymal tumor of the dermis, accounts for approximately 4% of all soft tissue sarcomas and 0.1% of all cutaneous malignancies with an incidence of one case per million per year.^[1] The tumor presents as an indolent growing plaque, usually localized on the trunk, the proximal extremities, and the head-neck region. DFSP is most often diagnosed in middle-aged adults, but it has been described in infants and the elderly as well. Most tumors are less than 5 cm, but untreated DFSP can attain massive dimensions^[2] as in our case. Wide local excision offers the best cure and prolonged follow up is indicated due to high recurrence. We are presenting a rare case of recurrent giant DFSP, managed by wide local excision with skin grafting and postoperative radiotherapy therapy.

CASE REPORT

A 49 years old female presented in surgical clinic with a history of progressively increasing growth over lower abdomen since two years with rapid increase in size for last three months. She also complained of ulcerations over the swelling and foul smelling discharge since last one month. There was no history of fever and weight loss. She had undergone surgery around five years back for a similar lesion, no relevant documents were available. On general examination, she was anemic with poor nutritional status. Local examination revealed a huge mass of size 30cm x25cm with multiple potato like projections over the lower abdominal wall extending to the bilateral inguinal region, the mons pubis upto anterior

part of vulva with multiple discharging ulcerations. Scar mark from the previous surgery was also seen below the umbilicus. (Fig.1) There was no inguinal lymphadenopathy and rest of abdominal examination did not reveal any abnormality.

Her blood investigations revealed Hemoglobin of 8gm% and rest, including biochemistry were all within normal limits. X-ray chest PA view and an ultrasonography of the abdomen did not reveal any abnormality. MRI abdomen revealed bulky heterogeneous mass lesion of skin-subcutaneous tissue not invading the underlying fascia or muscle. (Fig.2).

After correction of anaemia, she was underwent wide excision of the growth, with 3cm margin along with excision of the anterior rectus sheath. The defect was covered with split skin graft. Post-operative period was uneventful and she was discharged on 10th postoperative day. The histopathology of excised specimen revealed monomorphous spindle cells arranged in a storiform pattern; embedded in scarcely to moderately dense fibrous stroma (Fig.3). Immunohistochemical analysis with CD-34 was compatible with dermatofibrosarcoma protuberance. Surgical margins were free of tumor. In view of giant recurrent disease she underwent adjuvant radiotherapy and is on regular follow-up since 3 years without recurrence. (Fig.4)

LEGENDS



Fig-1: Image of Dermatofibrosarcoma Protuberance of lower anterior abdominal wall.



Fig-2: MRI abdomen shows lower abdominal wall growth arising from skin and sub cutaneous tissue.

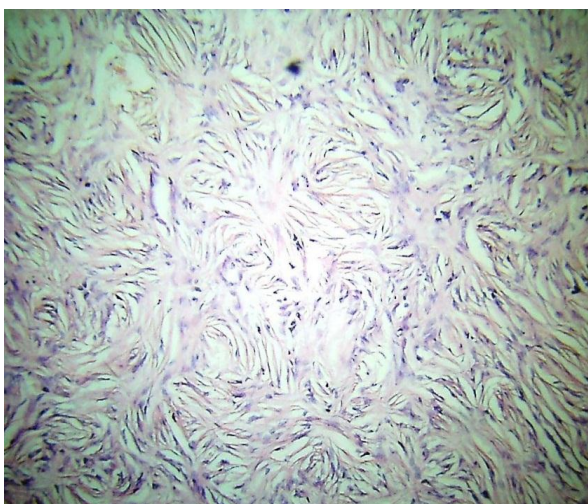


Fig. 3: Histological appearance showing proliferation of spindle cells arranged in storiform or cartwheel pattern (Haematoxylin and Eosin stain; original magnification $\times 400$).



Fig. 4: Clinical picture of post-op scar resulting from wide excision of tumor with split thickness skin graft.

DISCUSSION

DFSP was first described by Taylor in 1890. Darier and Ferrand described it as a gradual and recurrent cutaneous neoplasm in 1924.^[3] The term 'Dermatofibrosarcoma Protuberans' (DFSP) was coined by Hoffman in 1925.^[4] DFSP is a slow growing nodular, polypoidal neoplasm of the dermis. The lesions arise as pink or violet-red plaques, and the surrounding skin may be telangiectatic. Tumors, generally, are fixed to the dermis but move freely over deeper-lying tissue, but often fixed to more deep structures in advanced and/or recurrent cases.^[5] Moreover, DFSPs are superficial in 77%, and invaded deeper structures in only 22% of patients.^[6] The tumor occurs in patients of all ages, with the highest frequency occurring between the second and the fifth decades. Males affects slightly more common than in females and the male-to-female ratio is approximately 3:2.

CT scans or MR images are well suited to show the location, the relation of lesion to underlying structures and the distinct lobular or nodular architecture.^[7] Magnetic Resonance Imaging (MRI) is better when the tumor is large, particularly a large recurrence lesion. MRI shows well-defined lesion that had prolonged T1 and T2 relaxation times. On T1-weighted imaging, the tumor is isointense, slightly hypointense or hyperintense to skeletal muscle. Compared with that of fat, the tumor is of a high or intermediate signal, however a border can be hard to separate from fat on conventional T2-weighted images without fat saturation, but better depicted on short tau inversion recovery (STIR) imaging with the signal approaching water or blood.^[8] The differential, on imagings are dermatofibroma, neurogenic tumour, fibrosarcoma or malignant fibrous histiocytoma.^[7]

Diagnosis is usually established after excisional or punch biopsy which reveals characteristic uniform, slender, spindle shaped, fibroblast like cells, arranged in a typical storiform or cartwheel pattern. Immunohistochemistry using CD34 is a useful marker to confirm the diagnosis and also helpful in identifying tumour cell at the surgical

margins, especially in recurrent DFSP. Cytogenetically, DFSP commonly has translocation involving PDGF-beta & COL1A1.^[6]

Surgery is the mainstay of treatment. Wide excision with a safety margin of 3 cm including the underlying fascia is recommended. Emphasis is on histologically free margins for local control. Mohs micrographic surgery (1978) has the advantage of high oncologic effectiveness and maximal tissue saving and is increasingly accepted as the treatment of choice.^[9]

Chemotherapy is not useful. Adjuvant radiotherapy seems to be controversial, recommended in large low-grade (> 5 cm) or high-grade sarcomas, positive resection margins, where wide excision also may result in major cosmetic or functional deficits.^[5] Recently molecular targeted therapy using Imatinib mesylate has been used successfully to treat unrespectable and metastatic lesions as well as locally advanced primary and recurrent DFSP. It competitively inhibits the adenosine triphosphate-binding site of the platelet-derived growth factor B (PDGF -B) receptor tyrosine kinase in DFSP. Over activation of PDGF B receptor tyrosine kinase leads to cellular proliferation and tumor formation. When imatinib binds at the receptor site, kinase activity down regulates, resulting in growth inhibition and increased apoptosis.^[10]

DFSP is characterized by local invasion and recurrence. The likelihood of local recurrence after wide excision is less than 10%; it exceeds 50% when the final margins are positive.^[5] Although metastasis is rare, lung metastasis is most common, while lymph node metastasis is exceedingly rare. Poor prognosis is characterized by its late presentation, aggressive local invasion, regional nodal involvement and distant metastasis. Moreover, histologic poor prognostic indicators are; high number of mitotic figures, increased cellularity, DNA aneuploidy, TP53 gene over expression and fibrosarcomatous change. Regular follow-up is an important as the tumour has a high chances for recurrence.

CONCLUSION

DFSP is a rare tumour of dermis. Painless, cutaneous, and multilobulated lesions should arouse the suspicion of this tumour. Core or incision biopsy helps in pre-operative diagnosis. MR imaging allows accurate preoperative assessment and aids in the diagnosis. Surgery is the main stay of treatment. Wide excision should be ensured. Chemotherapy is not useful and radiotherapy has limited role. Follow-up is important as the tumour has a high chances for recurrence.

Conflict of Interest

None.

Funds

None.

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REFERENCES

1. Mc Peak CJ, Cruz T, Nicastrì AD. Dermatofibrosarcoma Protuberans: An analysis of 86 cases- five with metastases. *Ann. Surg.* 1967; 166: 803-16. PMID:4964386.
2. Mendenhall WM, Zlotecki RA, Scarborough MT. Dermatofibrosarcoma protuberans. *Cancer.*, 2004; 101: 2503-8. DOI: 10.1002/cncr.20678.
3. Darier S, Ferrand M. Dermatofibrosarcomes progressifs et récidivantes on fibrosarcomes de la peau. *Ann Dermatol Veneréol*, 1924; 5: 545-562.
4. Hoffmann E. Ueber das knollentribende Fibrosarkom der Haut (dermatofibrosarcoma protuberans). *Dermatol Z*, 1925; 43: 1-28. DOI:10.1159/000250699.
5. Lindner NJ, Scarborough MT, Powell GJ, Spanier S, Enneking WF: Revision surgery in dermatofibrosarcoma protuberans of the trunk and extremities. *Eur J Surg Oncol.* 1999; 25: 392-397. <http://dx.doi.org/10.1053/ejso.1999.0663>.
6. Bowne WB, Antonescu CR, Leung DH, Katz SC, Hawkins WG, Woodruff JM, Brennan MF, Lewis JJ: Dermatofibrosarcoma protuberans. A clinicopathologic analysis of patients treated and followed at a single institution. *Cancer*, 2000; 88: 2711-2720. doi:10.1002/1097-0142(20000615)88:12<27::AID-CNCR>9.0.CO;2M.
7. Shrimali R, Garg L, Setia V, Jain, Dermatofibrosarcoma protuberans (DFSP)-CT findings in DFSP-a rare skin tumour. *Indian J Radiol Imaging*, 2002; 12(3): 357-8. <http://www.ijri.org/text.asp?2002/12/3/357/28482>.
8. Torreggiani WC, Al-Ismail K, Munk PL, Nicolaou S, O'Connell JX, Knowling MA: Dermatofibrosarcoma protuberans: MR imaging features. *AJR*, 2002; 178: 989-993. PMID:11906888.
9. Van deweyer E, De Saint ASN, Gebhart M. Dermatofibrosarcoma protuberans: how wide is wide in surgical excision. *Acta Chir Belg*, 2002; 102: 455-458. PMID:12561153.
10. Piotr Rutkowski, Agnieszka Wozniak, and Tomasz Switaj. Advances in molecular characterization and targeted therapy in dermatofibrosarcoma protuberans. *Sarcoma*, 2011; 1-6. doi.org/10.1155/2011/959132.