WORLD JOURNAL OF PHARMACEUTICAL RESEARCH
SJIF Impact Factor 8.084
Volume 13, Issue 2, 1006-1009. Case Study ISSN 2277–7105

DERMATOFIBROSARCOMA PROTUBERANS - CASE STUDY

***[1]Dr. Menka Changle and [2]Dr. Dhanraj Gaikwad**

[1]PG. Scholar, Dept. of Shalya Tantra, TAMV, Pune.

[2]Associate Professor, Dept. of Shalya Tantra, TAMV, Pune.

Article Received on
04 December 2023,

Revised on 24 Dec. 2023,
Accepted on 14 Jan. 2024

DOI: 10.20959/wjpr20242-31060

***Corresponding Author**
Dr. Menka Changle
PG. Scholar, Dept. of Shalya Tantra, TAMV, Pune.

INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) is a rare soft tissue tumor that involves the dermis, subcutaneous fat, and in rare cases, muscle and fascia. The tumor typically presents as a slowly growing, firm plaque mainly on the trunk of young adults. The cause of DFSP is not clearly understood. Studies have implicated a chromosomal translocation, resulting in the fusion protein COL1A1-PDGFB, which promotes tumor growth through the overproduction of platelet-derived growth factor (PDGF). Diagnosis is made via skin biopsy. DFSP is considered an intermediate-grade malignancy with a low likelihood of metastasis but a high local recurrence rate. Given its propensity for a subclinical extension, the optimal treatment modality for DFSP is Mohs micrographic surgery (MMS), a surgical technique that allows complete margin assessment and tissue preservation. Alternatively, DFSP can be treated with wide local excision, chemotherapy or radiation therapy.

AIM

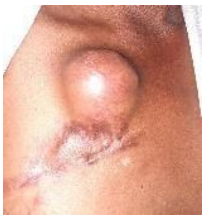
- Review the typical clinical presentation of a patient with dermatofibrosarcoma protuberans.

OBJECTIVE

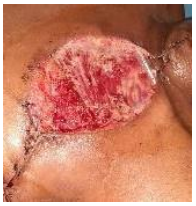
- Outline the treatment strategy for a patient with dermatofibrosarcoma protuberans.

Presenting complaints & medical history

A 55-year-old man was admitted to our department with a large protuberant mass located at the right side of his chest wall. Thepatient reported an increasing size of the mass during the preceding six months. The patient denied any recent weightloss, fever, night sweats or chills.

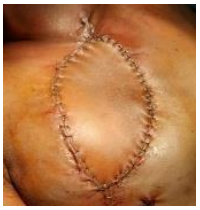


On physical examination, a large, firm, painless mass adherent to underlying & skin was found with no sign of localized heat or redness. There were no palpable cervical or axillary lymph nodes. There was neither personal nor familial history of malignancy.



Patient had previously operated for a mass 13yrs ago presented sameon the right side of Chest wall, Histopath s/o Fibrous Histocytocytic Tumor of soft tissue (benign).

The patient underwent wide excision under general anaesthesia, with 3cm lateral and deep resection margins & sent for Histopath s/o Multifocal Dermatofibrosarcoma Protuberans (DFSP). Nearest skin cut margin is free of tumor. Base of excision is involved by tumor. Patient additionally Underwent wide local excision combined with subsequent Latissimus Dorsi flap reconstruction.



RESULT

Based on the histological and immunohistochemical findings, the diagnosis of DFSP wasmade.

Treatment & Management

The treatment of dermatofibrosarcoma protuberans is surgical removal, ideally with Mohs micrographic surgery (MMS), a surgical technique that ensures complete histopathologic margin control at the time of surgery.

Wide local excision may be performed with 2- to 4 cm margins. However, local recurrence is relatively common with wide local excision even with clear surgical margins.

Chemotherapy agent imatinib mesylate, an oral tyrosine kinase inhibitor, can be used for recurrent, unresectable, and metastatic dermatofibrosarcoma protuberans in adults. The duration of therapy varies. Some sources recommend 6 months of therapy, but this may be extended if needed.

Radiation therapy may also be used for unresectable or recurrent tumors, and adjuvant radiation may decrease the risk of local recurrence.

DISCUSSION

DFSP is a rare, slow-growing malignant fibroblastic mesenchymal skin tumor which constitutes less than 0.1% of all malignant neoplasms and 1% of all soft tissue sarcomas. In the early stages, DFSP should be differentiated from lipomas, epidermal cysts, keloids, dermatofibroma. In the later stages, the differential diagnosis should consider pyogenic granuloma, Kaposi sarcoma, and other soft tissue sarcomas.

CONCLUSION

DFSP is a rare disease, Pathological examinations are required in patients with suspected DFSP, with the aim of minimizing the misdiagnosis rate. Once diagnosed, DFSP requires prompt treatment by extended tumor resection, followed by an increased follow-up frequency. Combined treatment requires consideration to reduce the recurrence rate in unresectable cases or in patients with repeated recurrence following resection.

REFERENCES

1. Neff R, Collins R, Backes F. Dermatofibrosarcoma protuberans: A rare and devastating tumor of the vulva. Gynecol Oncol Rep, 2019 May; 28: 9-11.
2. Gladdy RA, Wunder JS. Risk-stratified surveillance in dermatofibrosarcomaprotuberans: Less is more. Cancer, 2019 Mar 01; 125(5): 670-672.
3. Robbins pathologic basis of disease.
4. Baily & love short practice of surgery 24th edition.
5. Sabiston textbook of surgery 18th edition.
6. Schwartz's principles of surgery 8th edition.
7. www.medscape.com.

8. British Journal of cancer Accepted, 9 September 2002.