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Case report

Dermatofibrosarcoma- An uncommon entity, commonly mismanaged: a case report

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ABSTRACT

Introduction & importance: Dermatofibrosarcoma protuberans (DFS) is a slow-growing, recurrent, cutaneous soft tissue sarcoma with low metastatic potential. It is not uncommon for DFS to get misdiagnosed and treated like commoner parietal wall swellings, viz., epidermal cysts, lipomas and fibromas. Suboptimal management often leads to recurrence, which may be difficult to manage.

Case report: A 33-year male was referred to us with a lump in his lower abdomen for 15 years, without any symptoms. The patient underwent medical management for 6 years, followed by excision. The swelling recurred four years later and progressed in size till the next 5 years. At our centre, the patient underwent imaging and aspiration cytology, which established diagnosis of DFS, followed by wide local excision. Biopsy was conclusive and margins negative.

Discussion: DFS presents commonly as an asymptomatic indurated plaque that slowly enlarges over months to years. Untreated, DFS can attain massive dimensions, producing the large "protuberant" nodules, and hence the name. In the current report, 15 years elapsed before the correct diagnosis was established. In this case, recurrence first appeared after four years of excision. The possibility of primary wound closure after wide excision decreases with every subsequent excision, and reconstructive options may not be readily available.

Conclusion: The current report highlights a diagnostic delay of 15 years in a case of DFS. Asymptomatic, indolent nature combined with low awareness among community doctors contribute to delay in timely diagnosis. Community doctors should consider DFS as a differential in any long-standing, indolent, asymptomatic parietal wall swelling, especially with a history of recurrence.

1. Introduction

Dermatofibrosarcoma protuberans (DFS) is a cutaneous soft tissue sarcoma. It is uncommon yet significant due to the propensity for local invasion and recurrence [1]. It most commonly presents as a firm, dermal swelling. Slow growth, high recurrence, and low metastatic potential characterize DFS [2]. Low prevalence compounded by lack of awareness among physicians often leads to DFS misdiagnoses and mismanagement on lines of commoner parietal wall swellings, viz. lipoma, epidermal cysts, and fibromas. Such treatment, however, is fraught and often followed with recurrence, leading to patient and doctor frustration. We share a similar case, where the correct diagnoses took fifteen years, and the patient had to undergo re-excision due to recurrence. The report is consistent with SCARE 2020 criteria [3].

2. Case report

A 33-year man was referred to our surgical outpatient clinic with a lump in lower abdomen for 15 years. The patient noticed it initially at the age of 18 years. It was asymptomatic, around 3 by 3 cm, and indolent. He took medicines from multiple physicians for the same, but the lump did not regress. It remained asymptomatic, yet grew to the size of approximately 8 * 8 cm in the ensuing six years. A local practitioner excised the lump surgically at this point. Post-operative period was uneventful. However, the patient noticed multiple nodules at the surgical stitch line four years after excision. Drug history and family history were unremarkable. The lesions were asymptomatic, increased in size progressively, coalesced, and evolved into a single lobulated swelling over the subsequent five years (Fig. 1). A tissue biopsy had not followed the previous excision. Fine needle aspiration cytology at our center suggested DFS. On imaging and intraoperatively, the lump was found to

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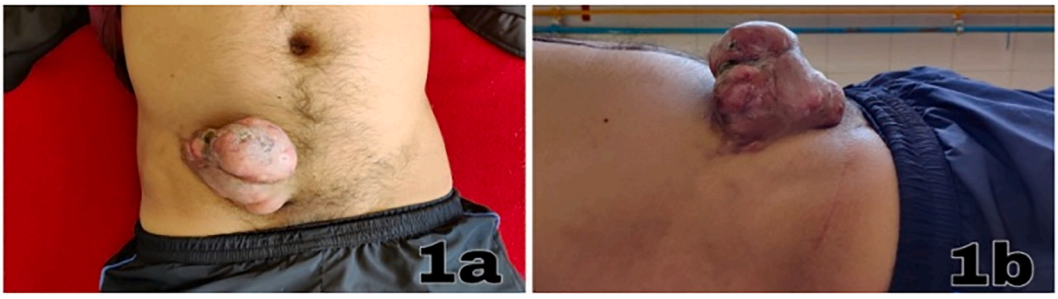

Fig. 1. Lobulated swelling in the right groin region: Recurred DFS post excision.

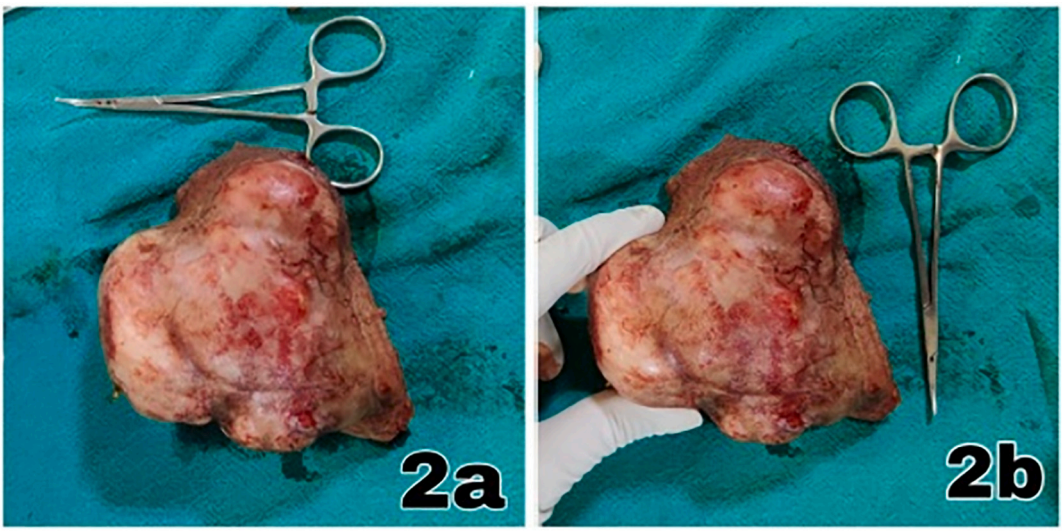

Fig. 2. Excised Tumor. (Size of mosquito forceps 5*3.5 inches).

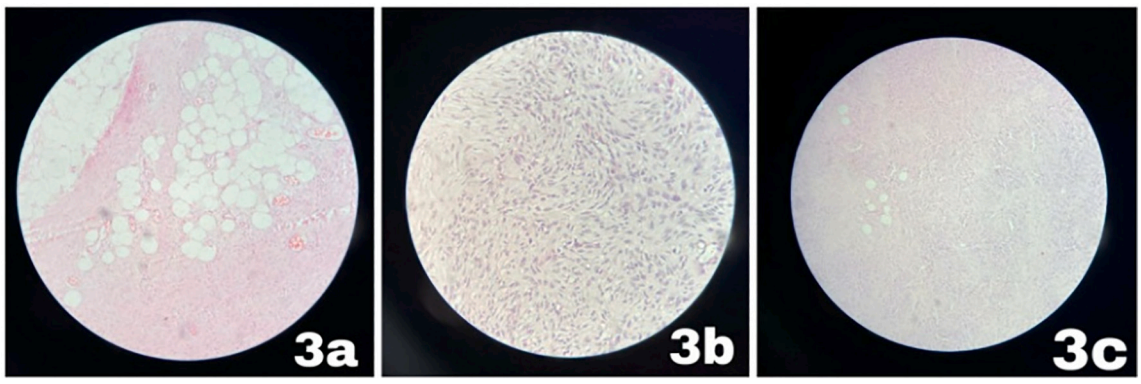

Fig. 3. DFS: Microscopy, showing, Infiltration in fat cells (a), mitosis (b) & storey form pattern (c).

be limited to supra-fascial planes. The patient underwent re-excision of the mass with five centimeter margins. (Fig. 2) The pathology report confirmed it to be DFS (CD34 positive) with negative margins. (Fig. 3) The patient continues to be in follow-up on an outpatient basis for six months without any recurrence.

3. Discussion

DFS is a mesenchymal tumor which is rare, representing less than 2% of all soft tissue sarcomas and less than 1% of all cancers [4]. It commonly presents between third and sixth decades of life [5]. The common sites affected by DFS are the trunk (42–72%), proximal extremities (16–30%), and head & neck area (16%) [6]. It presents commonly as an asymptomatic indurated plaque that slowly enlarges over months to years. The overlying skin may be tinged (brown-yellow, red), sclerodermiform, or thinned due to stretching. The Bednar pigmented variant has an irregular surface and typically contains brown pigmentation [7]. As the tumor slowly enlarges, it becomes raised, firm, and nodular; surrounding telangiectasia is not uncommon [8]. Untreated, DFSP can attain massive dimensions, producing the large "protuberant" nodules, and therefore, the name.

Diagnostic delay is not uncommon in DFS, partly due to low prevalence [9]. In the current report, 15 years elapsed before the correct diagnosis was established, which highlights the lack of awareness about

DFS among community doctors. Asymptomatic lump with indolent nature prevents patients from seeking secondary/tertiary care opinion. This is more likely to happen due to financial constraints among LMIC residents. This also increases the chances of misdiagnosis and subsequent mismanagement. In one series, more than three-fourths of lesions were superficial and less than 5 cm at diagnosis [10]. However, this report was from a developed country. In our setup, it's not infrequent to come across DFS that have attained massive dimentions at diagnosis.

Despite the delayed diagnosis, distant metastasis are rare (<2–4%); 5-year disease-specific survival rates approach 100% [10,11]. However, the local recurrence in DFS is a dreaded outcome. Local recurrence may happen in up to 50% of patients within three years [12]. In our case, recurrence first appeared after four years of excision. Margin-positive resection is supposedly the primary reason for such high recurrence. Inadequate margins due to presumptive alternative diagnosis is a common reason. The possibility of primary wound closure after wide excision decreases with every subsequent excision, unless reconstructive options are readily available. Therefore, the first excision stands the best chance of cure. Thorough pathologic examination of the specimen to rule out 'tumor pseudopodia' at margins is also vital to minimize recurrence. Timely radiotherapy to surgical beds can decrease recurrence in many cases [13].

4. Conclusion

Asymptomatic, indolent nature combined with low awareness among community doctors contribute to delay in the timely diagnosis of DFS. Excision on lines of commoner parietal wall swellings is fraught with a high incidence of recurrence. Although chances of metastasis are low, untreated DFS can lead to significant local destruction, warranting complex reconstruction of tissues. The aphorism 'a stitch in time, saves nine' is exemplified no better than DFS. Community doctors should consider DFS as a differential in any long-standing, indolent, asymptomatic parietal wall swelling, especially with a history of recurrence.

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Observational case studies are exempted from Ethical approval in MMMC & H.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Author contribution

Paran Tanwar: Design, First Draft
Amandeep Singh: Data, First Draft
Shaurya Pratap: Design, First Draft
Amulya Rattan: Design, Interpretation, Critical revision
Satinder Singh Minhas: Design, Interpretation, Critical revision

Registration of research studies

None.

Guarantor

Dr. Amulya Rattan.

Declaration of competing interest

- The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.
- Observational case studies are exempted from Ethical approval in MMMC & H.

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