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Dermatofibrosarcoma Protuberans - Delayed diagnosis in a Young Male

Tarteel Siddiqui, Komal Faheem, Mahrukh Shafiq, Nausheen Ferozuddin, Pirhay Fatma

ABSTRACT:

The dermatofibrosarcoma Protuberans is a cutaneous tumor that grows slowly. It is commonly misdiagnosed as other common abdominal swellings, such as fibromas or lipomas. Recurrence is frequently seen. A male patient, 27, presented with a complaint of Right iliac fossa (RIF) swelling that had persisted for 17 years. A lumpectomy was performed on the patient because of its growing size. Histopathology verified the presence of Dermatofibrosarcoma Protuberans (DFSP). A Wide Local Excision (WLE) with mesh repair was performed, biopsy revealed elongated monomorphic spindle cells that extended in a storiform pattern into the deeper subcutaneous fat with negative margins. Immunohistochemistry verified that it was DFSP. A 17-year diagnosis delay is highlighted in the current report. Delays are further exacerbated by the asymptomatic nature and lack of knowledge among community physicians. For any chronic, asymptomatic, indolent parietal wall swelling, local physicians should take DFSP into consideration as a differential diagnosis.

KEYWORDS*:* CD34 Antigen, Dermatofibrosarcoma Protuberans, Soft Tissue Neoplasms, Local Excision, Surgical Mesh

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INTRODUCTION:

The cutaneous soft tissue sarcoma known as Dermatofibrosarcoma Protuberans (DFSP) is a rare, low to moderate-grade tumor that originates from the dermis. It is a locally aggressive skin tumor that rarely spreads and tends to recur after removal.[1] Previously thought to be of fibroblastic origin, recent immunohistochemical evidence suggests it may originate from dendritic cells in the skin.

The overall incidence is 6.25 per million with significantly higher rates in darker-colored individuals.[2] The trunk region accounts for 40–60% of cases of DFSP, followed by the proximal extremities (20–30%) and the head and neck (up to 15%). Nonetheless, unusual sites including the scalp, breast, and toes have been the site of incidents. Similar to other soft tissue sarcomas, DFSP shows gender predominance, with a higher correlation with the male gender.[3] As the name suggests, DFSP can grow to enormous proportions, resulting in large, protuberant nodules, causing significant local damage that needs complex tissue reconstruction.[4]

A young male patient with dermatofibrosarcoma of the right iliac fossa is described in this article. Incorporating a review of relevant research, it seeks to highlight the difficulties in managing this disease in situations with limited resources. The case report is consistent with SCARE 2023 criteria.[5]

CASE REPORT:

A male patient, age 27, was referred to the outpatient clinic due to a 17-year-old mass in his right iliac fossa. At the age of ten, the patient first became aware of it. It was indolent, measuring about 7 cm x 10 cm. For a long time, he did not seek medical attention. It was asymptomatic, but within the past three years, it has grown to a size of around 10 × 10 cm, which was uncomfortable. (Figure 1)

Upon physical examination, there was no sign of localized heat or redness and a big, hard, painless, multinodular, mobile mass with irregular margins was noted. No inguinal lymph nodes could be palpated. The patient was sent to the general surgeon since the lump became increasingly uncomfortable. Baseline and Ultrasound was done which revealed hypoechoic mass with peripheral projections and increased vascularity. An excisional lumpectomy was done. The post-operative phase went smoothly, and there was no surgical site infection.

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The removed mass was submitted for histology which showed subcutaneous tissue exhibiting cellular dermal proliferation of spindle cells extending into subcutaneous fat arranged in a storiform pattern with no atypia or mitoses. The sample was determined to be Dermatofibrosarcoma, and a wide local excision (WLE) was intended to achieve definite, clean margins due to the uncommon diagnosis. Although metastasis is a rare possibility, to be sure there were no underlying secondary masses, CT scans of the chest, abdomen, and pelvis were conducted that were deemed unremarkable. During the WLE, a 5 cm tumor-free margin was ensured through frozen section, which was effective in achieving a distinct perimeter around the resected tissue in all directions (Figure. 2). In the same procedure, and a 15 x15 cm prolene mesh was inserted for reinforcement of anterior abdominal wall. Immunohistochemistry of the sample demonstrated that the cells were CD34 positive further confirming the diagnosis of DFSP. Post-op recovery was unremarkable.

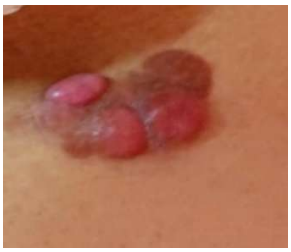

Figure 1: RIF Mass before Lumpectomy.

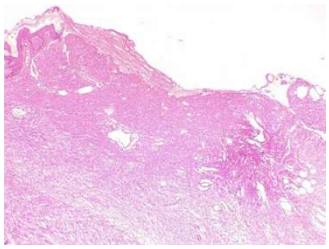

Figure 2: DFSP cells extending from dermis up to epidermis

DISCUSSION:

Prevalence in patients with DFSP were reported as 67.6% were Caucasian, 14.5% were Africans, 4.2% were Asian, 11.2% were Hispanic, and 2.4% were others. It is noteworthy that there was no difference in tumor size, treatment regimen, initial tumor location, or gender between the racial groups when compared to all other demographic factors. One protective factor against metastasis was a higher socioeconomic position.[2,6] There were no appreciable differences in tumor size or the types of treatment (chemotherapy, radiation, or surgery) across the various racial and ethnic groups. Surgery alone was performed on the majority of patients (75.7%), followed by surgery plus radiation (13.4%).[6]

DFSP typically begins as a tiny, painless, flesh-colored or brown nodule or papule. It can progress to either a sluggish nodular phase or a plaque phase, which can result in sclerosis or atrophy. It often moves about and becomes adherent to the skin above but not the tissues underneath. It may gradually penetrate adipose tissue and affix itself to deeper tissues like muscle and fascia. Telangiectasias may be seen at the edges or on the surface. A history of different traumas is frequently linked to the development of DFSP. This covers scars from surgery, injuries, burns, radiodermatitis, vaccination sites, central venous line placements, and bug bites.[7] Cachexia, like with other neoplasms, is rare. Early-stage lipomas, dermatofibroma, keloid, epidermal cysts, and nodular fasciitis should all be included in the differential analysis. Similar to the protuberant stage, additional soft tissue sarcomas such as Kaposi Sarcoma and Pyogenic Granuloma may be taken into consideration.[8]

Microscopically, it is often a well-differentiated sarcoma with large, elongated nuclei, modest mitotic activity, and a uniformly dense proliferation of spindle cells with CD34 positive marker. In addition to cells arrayed in the traditional storiform pattern, the stroma may have varying amounts of collagen and capillaries. [9] (Figure 3) A cartwheel-like arrangement of the cells may be formed by their radial arrangement around a central fibrous core. Areas with high-grade fibrosarcomatous change (FS-DFSP) may coexist with the traditional DFSP. When examined at low magnification, the fibrosarcomatous zones are distinguished by a fascicular, herringbone pattern of development. These regions usually exhibit mitotic figures, cytologic atypia, and enhanced cellularity. (Figure 4).

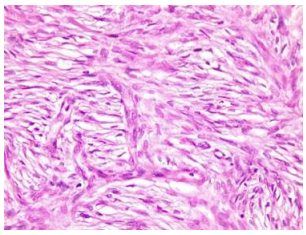

Figure 3: Classic Storiform Pattern seen in DFSP.

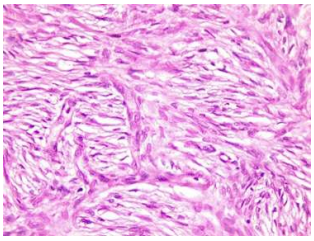

Figure 4: Herringbone Pattern at low magnification.

DFSP that exhibit symptoms of fibrosarcomatous transformation, a complete investigation is typically necessary even though distant metastases are uncommon. An ultrasound of the lymph nodes, a chest radiograph, and an abdomen ultrasound or CT scan are required for the diagnosis of metastatic illness. Although MRI and ultrasound may not guide about tissue infiltration, they can be helpful for the surgical plan, especially for individuals who have significant recurring lesions. The American Musculoskeletal Tumor Society (MSTS) approach, which assesses tumor grade and compartmentalization, is used to stage DFSPs and DFSP-FSs. Stage IA indicates no extension beyond subcutaneous compartment, Stage IB indicates involvement of the underlying fascia or muscle.[10]

Table 1: Musculoskeletal Tumor Society Surgical Staging

Stage	Grade	Local Extent	Metastasis
I-A	Low	Intercompartmental	-
I-B	Low	Extracompartmental	-
II-A	High	Intercompartmental	-
II-B	High	Extracompartmental	-
III	Any	Any	Present

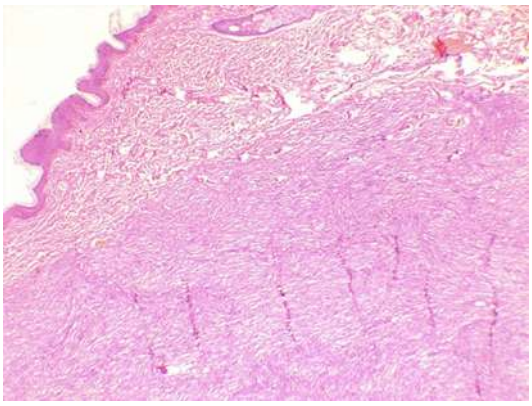

Figure 5: Dermal-based tumor with uniform, spindle-shaped cell

After partial excision, the recurrence rate is quite high. Thorough planning is necessary for the surgical treatment of dermatofibrosarcoma protuberans (DFSP), taking into account variables such tumor size, margin control techniques, location, and cosmetic results. The initial excision is the most hopeful for a cure since the likelihood of establishing primary wound closure decreases with each further excision unless there are efficient reconstructive solutions available. Wide local excision (WLE) with surgical margins of 1–5 cm of healthy, uninvolved skin is the standard of care. In order to identify and eliminate any "tumor pseudopodia" at the margins and reduce the chance of recurrence, a comprehensive pathological analysis of the material is essential. In certain situations, radiotherapy in surgical beds can reduce recurrence.[12]

CONCLUSION:

A delayed diagnosis and thus, a delayed treatment action may result from DFSP's asymptomatic, inactive character. The tumor often spreads to the periphery after originating in the dermis. But in this case, the tumor only spread to the skin and subcutaneous tissue. There may be a correlation between a higher risk of recurrence and certain histological variations of DFSP. Mohs micrographic surgery may be the preferred treatment with the advantages of maximum tissue preservation, improved functional and cosmetic results, and accelerated recovery. However, wide local excision is the unquestionable treatment that may be coupled with radiation to minimize recurrence.

Authors Contribution:
Tarteel Siddiqui: Conception and design of work, acquisition analysis drafting of work
Komal: Drafting of work, final approval of version to be published
Mahrukh Shafiq: Design and acquisition of work
Nausheen Ferozuddin: Drafting of work
Pirhay Fatma: Interpretation of data for work

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