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CASE REPORT OF DERMATOFIBROSARCOMA PROTUBERANS OF AN ABDOMINAL LUMP

General Surgery

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ABSTRACT

We would like to share with you a rare instance of Dermatofibrosarcoma Protuberans, which was identified by a histological analysis of an abdominal mass. Having a lump in the center and on the left side of his lower abdomen, the 48-year-old male patient was brought into our hospital. abdomen CT revealed that the anterior and left lateral abdominal walls had a neoplastic etiology with an exophytic component and pre-peritoneal extension?A sarcomatous lesion?subcutaneous or cutaneous cancer. A tumor emerging from the anterior and posterior rectus sheath was discovered during surgical examination. Dermatofibrosarcoma Protuberans has histological characteristics that point to a fibrosarcomatous transformation. By histopathologically examining an abdominal mass originating from the anterior and posterior rectus sheath, a rare case of Dermatofibrosarcoma Protuberans was identified in this study.

KEYWORDS

INTRODUCTION

The uncommon and slowly growing cutaneous tumor known as dermatofibrosarcoma protuberans (DFSP) has a significant potential for invasion and recurrence locally. The diagnosis of DFSP is frequently postponed because of the slow-growing and indolent characteristics. A significant parietal defect caused by the treatment's intensive surgical technique may be difficult to correct. We describe a case of a middle-aged man who had a DFSP of the anterior abdominal wall treated with wide local excision (WLE) and primary closure. Through this report, we addressed the challenges of the diagnosis, the unique aspects of the abdominal localization's treatment strategy, and the pertinent research.

CASE REPORT

An abdominal bulge (over the middle of the abdomen and on the left lower side of the abdomen) that had been persistent for four years was the main complaint of the 48-year-old patient. The swelling began as a tiny cutaneous lesion across the front belly and grew bigger over time. An oval mass with regular boundaries and a polylobed surface was found during a physical examination; it was positioned in the middle of the abdomen of size 18×8×4 cm3 and 4×3 ×2 cm3 in size on the left lower side of the belly.

A firm, permanent swelling on the skin was discernible upon probing. Regarding the deeper musculoaponeurotic plane, it is still freely mobile. Black patches with ulcerated areas and seropurulent, foul-smelling, copious discharge were visible on the swelling's surface; however, the discharge eventually lessened. Large lobulated soft tissue density mass lesion measuring 16 x 6.7 x 6 cm3 was found in the umbilical and infraumbilical regions of the abdomen according to the CT scan result.

The underlying rectus musculature borders the lesion. Another tumor of a similar nature, measuring 6 ×1.9× 2.5 cm3 and infiltrating the abdominal musculature with an exophytic component, was observed in the left lateral abdominal wall. There were no metastatic lesions visible on CT. With 3 cm of minimum macroscopic safety margins around the tumor's palpable and visible boundaries, the patient underwent WLE.

Primary closure was used to close the resulting defect by moving the nearby tissue. In fact, the skin and subcutaneous tissue beneath the wound's caudal edge was lifted off the anterior abdominal aponeurosis, allowing for wound-free-tension coverage and flapping advancement. For the left lower side of the abdomen mass and the midline abdominal mass, closure was performed in two stages with two months between each.

The specimen's histopathology showed that it had spindle cells in a storiform arrangement, neoplastic spindle cells that showed nuclear pleomorphism with eosinophilic cytoplasm, and the presence of fibrocollagenous and adipose tissue. This is a characteristic of DFSP appearance (Grade 1 Fibrosarcomatous Transformation of Dermatofibrosarcoma Protuberans). The margins of the removed skin were clear of any tumor cells.

DISCUSSION

Less than 2% of all soft tissue sarcomas are DFSPs, an uncommon mesenchymal tumor that develops in the dermis. Like in the case of our patient, DFSP mainly affects young adults between the ages of 20 and 50. It is well recognized that the trunk and extremities are most typically affected by DFSP. The abdominal localization of DFSP is still one of the uncommon malignant tumors of the abdominal wall's soft tissues, which are predominately desmoid tumors.

The most typical DFSP presentation is an indurated plaque that is asymptomatic and slowly grows over months to years. Skin-colored, sclerodermiform or telangiectatic atrophic skin covers the tumors. It gets upraised, hard, and nodular if left untreated. Despite frequently being anchored to the dermis, the nodule has the ability to locally infiltrate deeply into the fascia, the muscle, and other underlying structures. Despite the delayed diagnosis, most tumors are superficial and smaller than 5 cm.

The lesion's history and its location in the dermis are the key supporting evidence for the indicated diagnosis. In fact, the delay in diagnosis is caused by indolent growth combined with the rarity of DFSP and the variety in clinical appearance. Therefore, DFSP should be recommended in any persistent, painless, asymptomatic parietal swelling to reduce diagnosis delay and eventual mismanagement. Histological results serve as the basis for the ultimate diagnosis. Therefore, DFSP can be easily identified by the growth of fibrohistocytic cells that express CD34.

The size and tumor history both affect tumor stage. Physical examination can really be used to quickly determine the extent and mobility of the tumor as well as a localized node in the event of small and superficial DFSP. However, imaging technologies like ultrasound and CT scan are frequently employed in normal clinical work, allowing to confirm the extension and offer objective lesion data for the surgical planning. Soft tissue attenuation, post-contrast agent enhancement, and a single, subcutaneous lobular-nodular architecture are all seen on a CT scan. Additionally, a CT scan can be used to assess distant metastatic patterns.Magnetic resonance imaging (MRI) is essential in determining the size and extent of a large tumor or a recurring primary tumor, as well as how it connects to nearby bone and neuromuscular tissues.

For DFSP, surgery is the primary form of treatment. It should consider any potential infra-clinical tumor expansions that might be the cause of

the local recurrence. WLE is the normal course of treatment. There is debate over the ideal resection margin width. However, most authorities advise using margins between 2-4 cm.

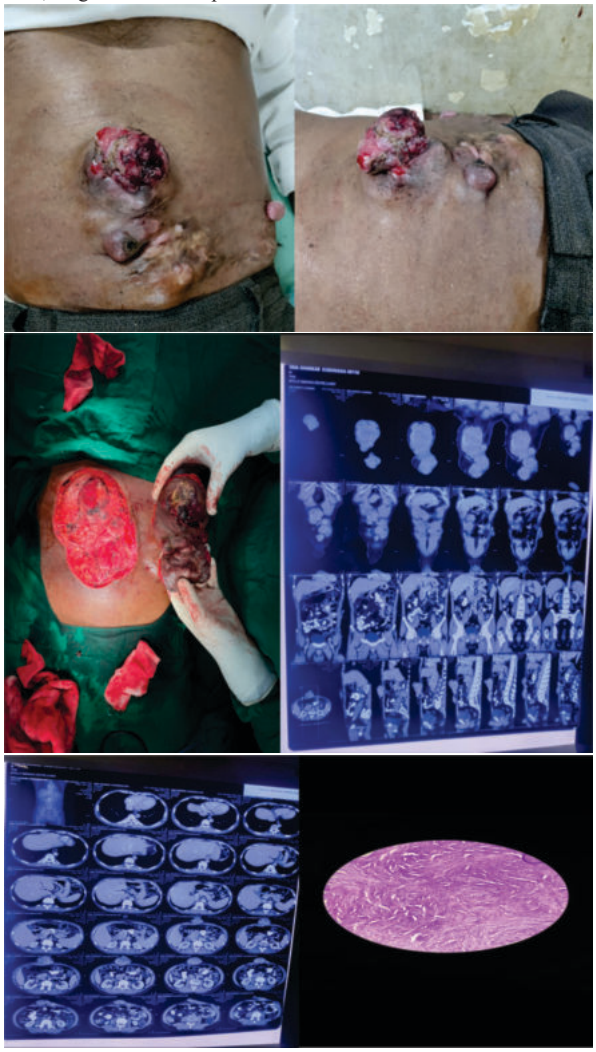
The size and thickness of the wound left behind after removing a tumor affects how those faults should be managed and repaired. In fact, lesions that are tiny and superficial—which account for the majority of DFSP in abdominal locations—are frequently treated with WLE and primary closure rather than the necessary resection of the entire thickness of the abdominal wall. The primary closure was possible in our patient for two main reasons.

The first flaw was spreading aponeurosis and incomplete thickness. Second, the abdominal wall's flexibility enables tension-free closing. However, a massive full-thickness excision is advised in cases of big tumors with deep penetration or recurrent tumors, and reconstruction is necessary.

The strategy for post-treatment follow-up has not been adequately established. The majority of local recurrences manifest within 3 years, while 30% take five years or more to manifest. Therefore, it has been recommended that patients undergo examinations every six months for the first three years, followed by annual exams moving forward.

CONCLUSION

A uncommon soft-tissue malignant tumor, DFSP has a significant potential for local invasion and recurrence but a low risk of metastasis. Clinical awareness of this condition is essential for an early diagnosis and effective treatment. The preferred method of treatment is wide local excision (WLE), which guarantees microscopically disease-free margins. Surgery should give careful consideration to an effective repair of the ensuing anterior abdominal wall defect. To determine any potential risk of recurrence that might happen over a long period of time, long-term follow-up is advised.



REFERENCES

1) Atoini F, Zidane A, Arsalane A, Elkaoui H, Kabiri EH. Tumeur de Darrier-Ferrand de la paroi thoracique antérieure. *Rev Pneumol Clin* 2008;64:265–6. [PubMed] [Google Scholar]
2) Kreicher KL, Kurlander DE, Gittleman HR, Barnholtz-Sloan JS, Bordeaux JS. Incidence and survival of primary dermatofibrosarcoma protuberans in the United States. *Dermatol Surg* 2016;42:S24–31. [PubMed] [Google Scholar]
3) Ortonne N. *Sarcomes cutanés*. www.em-premium.com/data/traites/de2/98-69434/ [Internet]. 2018. Available from: https://www-em-premium-com.scd-rproxy.u-strasbg.fr/article/1263799/resultatrecherche/5 (6 January 2019, date last accessed).
4) Stojadinovic A. Soft tissue tumors of the abdominal wall: analysis of disease patterns and treatment. *Arch Surg* 2001;136:70. [PubMed] [Google Scholar]
5) Mendenhall WM, Mark T S, Franklin PF. *Dermatofibrosarcoma Protuberans: Epidemiology, Pathogenesis, Clinical Presentation, Diagnosis, and Staging* [Internet]. UpToDate. 2018. Available from: https://uptodatefree.ir/topic. htm?path= dermatofibrosarcoma-protuberans-epidemiology-pathogenesis-clinical-presentation-diagnosis-and-staging (17 April 2022, date last accessed).