Open Access Original Research Article

MedPeer Publisher
Abbreviated Key Title: MedPeer
ISSN : 3066-2737
homepage: https://www.medpeerpublishers.com

Perianal protuberant dermatofibrosarcoma: a clinical case

DOI: 10.70780/medpeer.000QGS3

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ABSTRACT

Dermatofibrosarcoma protuberanta (DFSP), or Darier-Ferrand tumor, is a rare, intermediate-grade skin tumor characterized by slow but locally aggressive growth [1] We report the case of a 50-year-old woman with a subcutaneous mass in the gluteal cleft that had been present for five years. The patient received six months of neoadjuvant imatinib treatment, resulting in a significant reduction in tumor volume, before undergoing wide surgical excision. Pelvic MRI showed a subcutaneous lesion without invasion of deeper tissues, associated with bilateral iliac lymphadenopathy. The diagnosis was confirmed by histopathological and immunohistochemical analysis. This case illustrates the value of a multidisciplinary approach combining medical treatment and surgery in the management of locally advanced subgluteal cleft lymphomas.

KEYWORDS :

Perianal dermatofibrosarcoma – Darier and Ferrand tumor – Imatinib – excision recurrence_ radiotherapy.

MAIN ARTICLE

INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) is a rare fibroblastic skin tumor, representing less than 1% of soft tissue sarcomas. **[2]** occurs mainly in young or middle-aged adults and is most often located in the trunk. Although its metastatic potential is low, DFSP is locally aggressive, with a high risk of recurrence in cases of incomplete excision. The frequent identification of the COL1A1–PDGFB translocation has led to the development of targeted therapies using tyrosine kinase inhibitors, such as imatinib, used particularly in the neoadjuvant setting to reduce tumor volume and facilitate surgical excision in anatomically complex locations, such as the gluteal cleft **[3]**

CLINICAL OBSERVATION:

This is a 50-year-old female patient, originally from and residing in Meknes, a housewife, married with three children, and the wife of a retired member of the Royal Armed Forces. She has been treated for diabetes with insulin therapy for 9 years and for hypertension with amlodipine 5 mg/day for 3 years. The patient was exposed to secondhand smoke and had no significant family history of diabetes.

The history of the disease

It dated back five years to the appearance of a painless subcutaneous nodule the size of a coffee bean in the intergluteal cleft, which grew slowly with a progressive increase in size. This progression led the patient to consult several doctors before finally being referred to the general surgery department of the Moulay Ismail Military Hospital for specialized care.

On admission, the clinical examination revealed a clinically stable and healthy patient. Skin examination revealed a firm, painless mass, initially 7 cm in its longest dimension, which decreased in size after treatment. The overlying skin was healthy, without ulceration, fistulization, or signs of inflammation, and no palpable peripheral lymphadenopathy was found on examination of the lymph node areas.

Figure 1: Cutaneous projection of the tumor mass in the intergluteal cleft

The pelvic MRI revealed a posterior median intergluteal subcutaneous lesion measuring 75.2 × 65.4 × 35 mm, without involvement of deep muscle layers or adjacent bone structures.

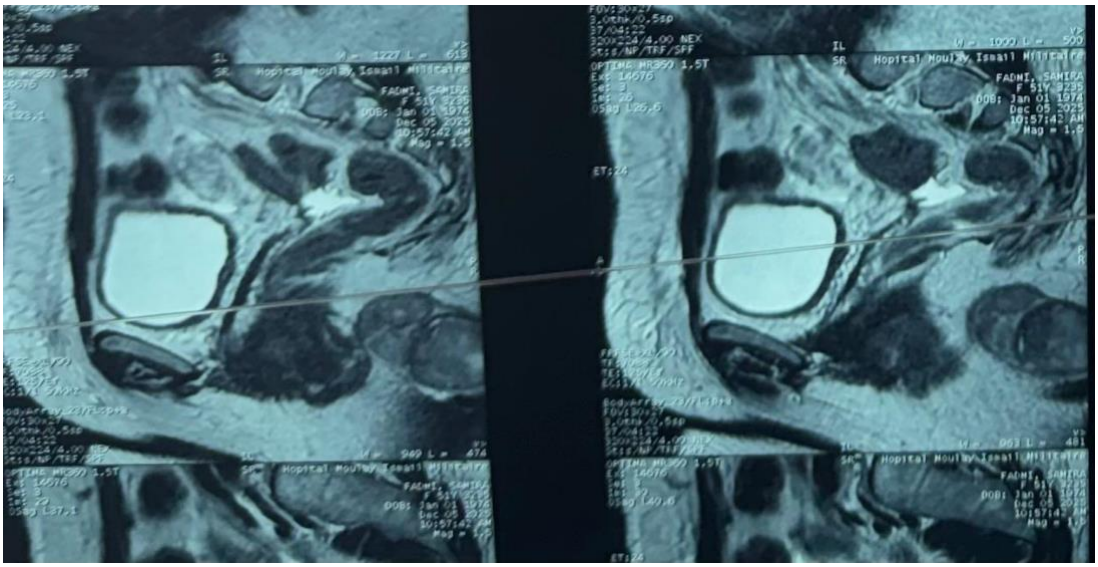

Figure No. 2 Coronal section in T2 sequence showing the arrangement of the tumor

Given the presence of this mass, a percutaneous trocar biopsy was performed. Histopathological examination showed a proliferation of spindle cells arranged in a scrofula pattern, infiltrating the dermis and hypodermis, consistent with a protruding dermatofibrosarcoma. Immunohistochemical analysis revealed strong positivity for CD34, thus confirming the diagnosis of dermatofibrosarcoma.

Support provided:

The body of histological and radiological evidence confirmed the diagnosis of protruding dermatofibrosarcoma of the intergluteal cleft. After discussion in a multidisciplinary tumor board meeting and considering the intergluteal location, the tumor size, and the functional risk associated with immediate excision, it was decided to initiate neoadjuvant treatment with imatinib for six months in order to reduce the tumor volume and facilitate complete surgical excision while preserving the sphincter structures.

A follow-up MRI, performed six months after the start of treatment with imatinib, revealed a significant volume reduction of the tumor, with dimensions decreasing to 62 × 45 × 35 mm, representing a reduction of approximately 43% of the initial volume, indicating a favorable partial response and allowing planning for wide excision surgery.

Following tumor reduction, a complete pre-operative assessment was performed, including a pre-anesthetic evaluation. The assessment was deemed satisfactory, and the patient received the anesthesiologist's approval for surgery. As part of the surgical preparation, bowel preparation with Fortrans was initiated 72 hours prior to the procedure to reduce the risk of bacterial contamination in the surgical area. This preparation optimized surgical exposure and decreased the risk of postoperative infection, particularly given the lesion's proximity to the intergluteal cleft and the anal canal.

Operative report:

The patient was positioned prone under general anesthesia, with a cushion placed under her hips to optimize exposure. After marking and measuring the circular excision margins around the mass according to the pre-established plan, a skin incision was made according to the preoperative drawing, allowing for en bloc excision of the lesion down to the sphincter bundle. The two bundles, superficial and internal, of the anal sphincter were identified and sutured to preserve continence. Closure was performed layer by layer, a Redon-type suction drain was inserted, and a sterile dressing was applied. The immediate postoperative course was uneventful, without local complications or sphincter dysfunction. The properly oriented excised specimen was sent to the pathology laboratory.

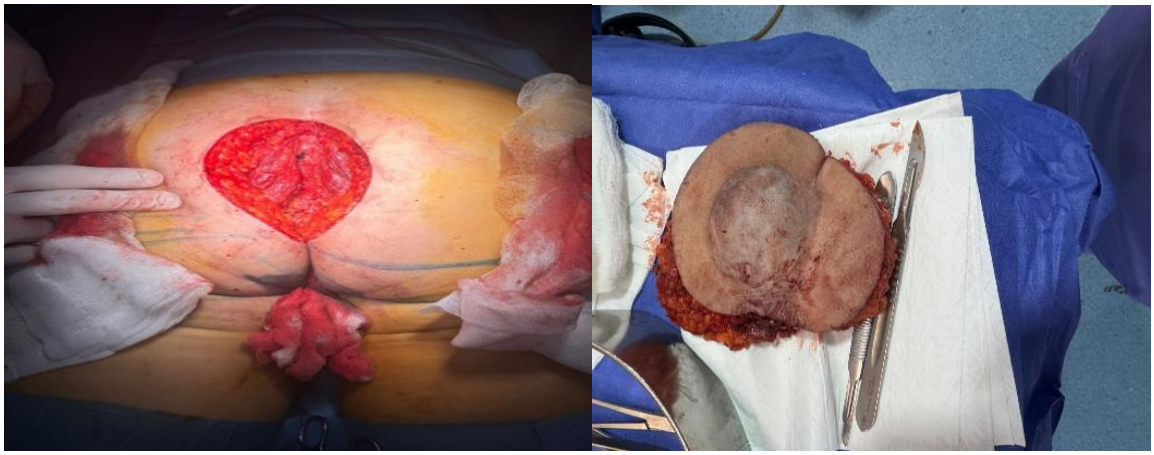

Figure 2: Tumor excision time while respecting safety margins

Post-operative care:

To prevent wound infection, ensure complete healing, and reduce the risk of recurrence, a rigorous postoperative monitoring protocol was implemented. On day 1, the patient was stable, with a clean dressing and 150 cc of serosanguineous fluid drained. On day 2, her clinical condition remained satisfactory, the dressing was still clean, and the drain yielded 50 cc of serosanguineous fluid. On day 3, given the good local progress, the drain was removed. On day 15, the dressing was clean, and the sutures were removed.

The anatomopathological examination

The excision was complete, with lateral margins of 2 cm less than 1 cm and a deep margin of 0.4 cm, all clear. Immunohistochemistry showed diffuse positivity of CD34, confirming the diagnosis of dermatofibrosarcoma protuberant.

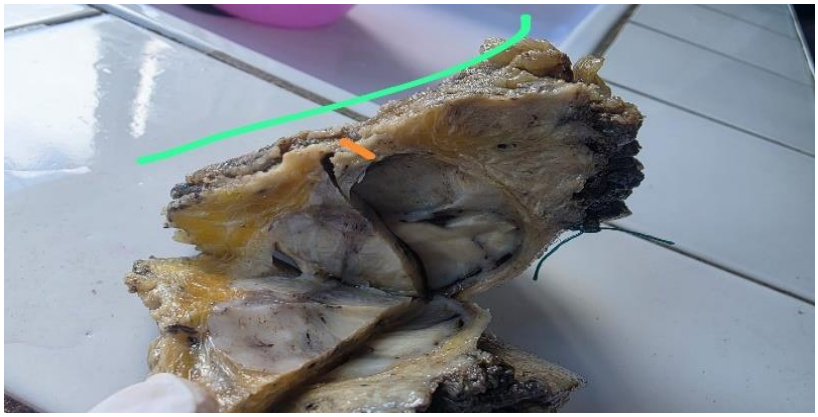

Figure No. 3: Macroscopic appearance of the tumor after fixation, showing the excision margins.

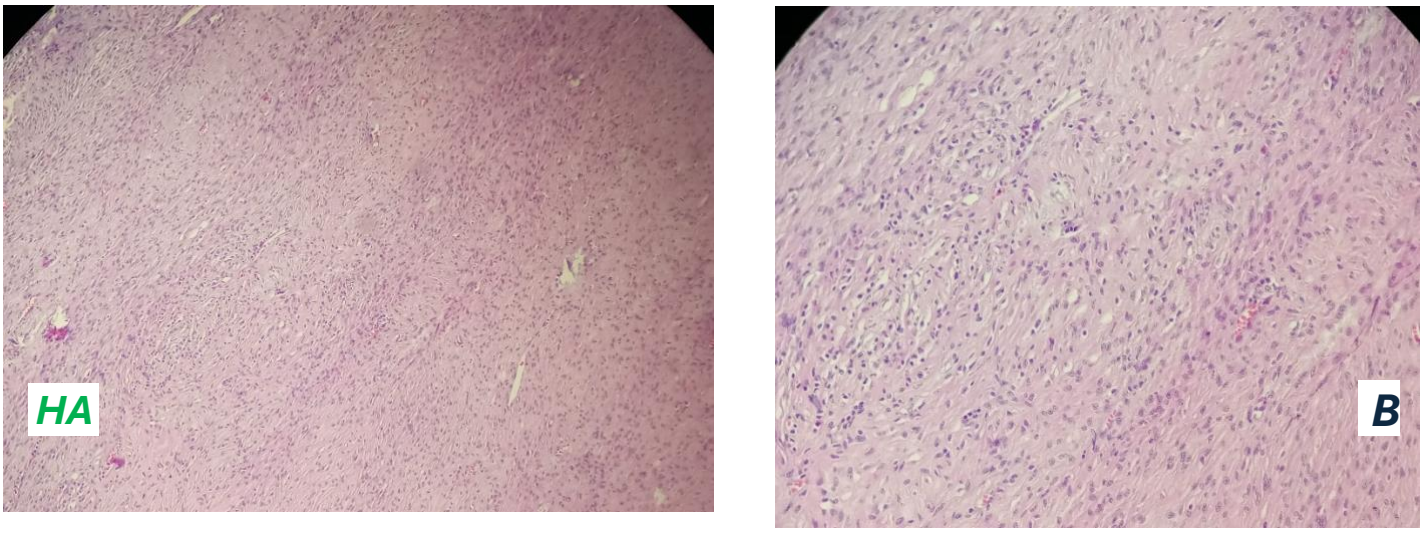

Figure No. 4:Histological sections taken from the tumor mass

A) Spindle cell proliferation in a storiform arrangement (H&E, ×100)

B) Expression of CD34 confirming the diagnosis of DFS

Figure 3: Appearance of the surgical scar after suture removal on day 15

Long-term monitoring;

Regular follow-up was initiated. One year later, clinical examination combined with digital rectal examination revealed no signs of local recurrence. The overlying skin was healthy. The anal canal was supple, patent, without narrowing or stenosis.
After obtaining the anatomopathological results confirming the diagnosis of protruding dermatofibrosarcoma and given the proximity of the deep safety margin (0.3 cm), it was decided to carry out targeted adjuvant radiotherapy at the level of the area with a narrow margin.

This additional radiation therapy aimed to eliminate any remaining microscopic tumor cells and reduce the risk of local recurrence.

DISCUSSION:

Dermatofibrosarcoma protuberans (DFSP) is a rare mesenchymal tumor characterized by slow growth but marked local aggressiveness. In our case, the disease history dated back several years, beginning with a small, painless subcutaneous nodule that gradually evolved, a pattern supported by several studies and often explaining the delay in diagnosis. **[4,5]**

The initial staging workup was negative, with no evidence of secondary lesions. Although DFS is characterized by marked local aggressiveness, its metastatic potential remains low, reported in the literature at between 3 and 5% **[6]**
Metastatic forms, although rare, are associated with a poor prognosis. In our observation, no metastases were observed, but strict and prolonged clinical monitoring is still warranted.

Intergluteal location presents a unique situation, making surgical management more complex due to the proximity of sphincter structures and the need to preserve anal function. This anatomical constraint necessitates rigorous surgical planning to ensure satisfactory oncological resection while minimizing functional morbidity.

In our case, the use of neoadjuvant imatinib was prompted by the increased tumor volume. This therapeutic strategy, now well-established in cases that are initially unresectable or have a high functional risk, resulted in significant tumor reduction, facilitating wide resection with clear margins. Surgery remains the gold standard treatment for DFS, and the lateral and deep

margins recommended in the literature generally range from 2 to 5 cm, depending on local anatomical constraints.

In our case, preserving healthy anatomical barriers, particularly the gluteus maximus aponeurosis, allowed for complete excision while limiting the extent of the surgical procedure. This approach is essential to reduce the risk of local recurrence, which remains high in DFSP when excision is incomplete.

Bowel preparation was performed, along with weaning from oral feeding and the initiation of hydration and parenteral nutrition for 7 days. This management resulted in a stool-free anus, thus promoting optimal healing.

In our case, no lymph node dissection was indicated. DFS is characterized by primarily hematogenous dissemination, with lymph node involvement being exceptional. Thus, in the absence of suspicious lymphadenopathy, routine lymph node dissection is not recommended **[7]**

Prolonged follow-up is essential, given the risk of late local recurrence, which can occur several years after initial treatment. In contrast, lymph node or distant metastases remain exceptional in DFS. Therefore, the presence of iliac lymphadenopathy observed in our case was not considered a sign of metastatic dissemination, in the absence of suggestive histological or radiological evidence.

CONCLUSION :

Protruding dermatofibrosarcoma of the intergluteal cleft is a rare and delicate location due to its proximity to the anal sphincter. In our case, management combined neoadjuvant imatinib therapy, surgical excision with 3 cm margins reduced to 1 cm at the sphincter to preserve function, and postoperative radiotherapy. This multimodal strategy optimizes local control while maintaining continence, provided that prolonged clinical follow-up is maintained to detect any recurrence.

ACKNOWLEDGEMENTS

The authors declare that they have no conflicts of interest.

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