

A Case of Primary Dermatofibrosarcoma Protuberans of the Lower Anterior Abdominal Wall Treated by Abdominoplasty

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Abstract

Dermatofibrosarcoma protuberans is a rare skin neoplasm with a high recurrence rate and a local invasive behavior. Excisional surgery is the mainstay of treatment of such tumors. Obtaining an adequate surgical safety margin is essential to ensure radical excision, however this may result in extensive skin and soft tissue defects that may require resurfacing. In this clinical case report, we describe a case of a large DFSP of the lower anterior abdominal wall, treated by abdominoplasty, we also perform a relevant scoping literature review on the topic.

Keywords: Dermatofibrosarcoma protuberans; Abdominoplasty.

Introduction

Dermatofibrosarcoma protuberans (DFSP) comprises less than 5% of all soft tissue sarcomatous neoplasms, but accounts for a fifth of all cutaneous sarcomas, coming second only to Kaposi's sarcoma. It has an incidence of less than 2.5 per million per year [1,2]. Although rare, DFSP is the third most common abdominal wall soft tissue malignancy; being exceeded only by Desmoid tumors and soft tissue sarcoma (STS) [3,4].

The lesions usually start as discolored cutaneous plaques or nodules that enlarge slowly over time and can attain large sizes. The tumors metastasize in less than 5% of all cases but have a challenging Loco-regional behavior due to their peculiar growth pattern, which is characterized by a tentacular, frond – like projections into surrounding tissue that extend variable distances from the main tumoral mass [5-7]. Such growth pattern is often indiscernible clinically or on imaging

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and necessitate a careful histological assessment of margins to ensure adequate oncological excision [5].

Excisional surgery remains the mainstay of treatment and loco-regional recurrence represent the most common form of treatment failure [8-10]. Risk of recurrence has been found to be inversely Proportional to the safety margins distance. A minimum of 2-3 cm safety margin is recommended. Wide local excision of DFSP usually results in a significant tissue loss and disfigurement that necessitate reconstructive procedures [10].

We treated a patient who presented with a sizable DFSP in the infra-umbilical part of the anterior abdominal wall by performing abdominoplasty. Such procedure is versatile in design and can be modified and employed to both provide an adequate excisional margin and an esthetically pleasing immediate reconstruction for anterior abdominal wall lesions.

Case report

A 51-year-old Bangladeshi male carpenter was referred to our unit with an asymptomatic anterior abdominal wall cutaneous lump. The patient first noticed the mass 3 years earlier while taking a shower. The mass was pea sized, with slightly darker covering skin. It was progressive in size, remained asymptomatic, but was associated with color variegation, bosselation, and skin peeling. There were no similar lesions elsewhere or erstwhile. The patient volunteered no other complaints and didn't link the lesion to any antecedents. Symptoms review elicited no Loco-regional, Radicular, Systemic, constitutional or ALARMS symptoms. There

was no significant past medical history, and the family history was unremarkable. The patient was not on any medication and denied any drug allergies.

Examination was only significant for a solitary cutaneous protuberant mass, located 3 cm below the umbilicus and 1 cm to the left of the midline. It had a circular dark perimeter with a visible diameter of 11 cm and a summit at 4 cm from the base. The mass was bosselated with three obvious hypopigmented bosses. The skin cover of the mass was color variegated, thinned at the bosses, crepey and peeling. The surrounding skin, however, was unremarkable.

On palpation the lesion was isothermal to its surrounding, non-tender, firm and incompressible. It was inseparable from its covering but moved over the underlying tensed abdominal musculature in all meridians.

Subcutaneous satellite or tentacular spread was not detected and there was no involvement of the draining lymphatic basins. No evidence of distant spread was found on clinical examination.

The patient was consented for medical photography and an image of the lesion was uploaded in a reverse image search engine. The majority of the matched images were of DFSP lesions, which matched the most probable diagnosis on our differential diagnoses list.

The patient was informed of the clinical findings and the possible diagnoses. A diagnostic plan was offered, and the patient consented to proceed with it.

An abdominopelvic CT scan revealed a solitary superficial protuberant mass concordant in morphology and topography with the clinical findings. The mass was found inseparable from the overlying overstretched skin, but the underlying abdominal musculature appeared to be uninvolved by the tumor. There were no other abnormalities detected by this study, or by a complementary chest CT.

A tru-cut biopsy of the lesion revealed spindle cells in storiform pattern in keeping with DFSP, not otherwise specified.

The case was discussed in the MDT meeting and a consensus on WLE with a safety margin of 3 cm, as the best treatment option was reached. The options for resurfacing the resulting skin and tissue defects were reviewed and discussed. In view of the topography of the lesion, abdominoplasty was considered to be well suited in terms of providing both adequate excision and esthetically pleasing reconstruction.

The patient was informed of the diagnostic findings and counseled regarding his therapeutic options in light of the MDT meeting recommendations. He consented for the procedure and was admitted after completing preoperative anesthesia evaluation.

Preoperative skin marking was performed taking into consideration the safety margin requirements and in line with classic anterior abdominoplasty operative principles. The inferior incision line was marked 3 cm above the pubic symphysis and extended laterally in a handle-bar configuration. The superior incision line was elected 2 cm above the umbilicus as dictated by the estimated stretchability of the supra-umbilical flap. It was extended laterally to the point of intersection with the inferior incision marking. Symmetrization of incisions markings was ensured by using reference grid lines in the supine and the standing positions (Figure 1).



Figure 1: Pre-operation marking of the lesion.

The histopathology was confirmatory of the preoperative diagnosis of DFSP (Figure 2), and all margins were free. The posterior margin was the closest with a distance of 1 cm, however the excised underlying rectus sheath was unremarkable. All other margins were more than 3 cm.

The tumor bearing fasciocutaneous pannus bounded by the incision markings was excised. The posterior excision limit was the anterior rectus sheath and external oblique aponeurosis. The Part of the rectus sheath

immediately posterior to the tumor was excised, despite the lack of apparent radiological or operative involvement, as a precautionary measure.

The umbilicus was preserved on its cutaneous pedicle, and the supra-umbilical flap raised to the costal margin. The superior flap edge was brought to the inferior incision margin with minimal tension. No muscle plication was deemed necessary. The wound was closed over two redivac® Drains.

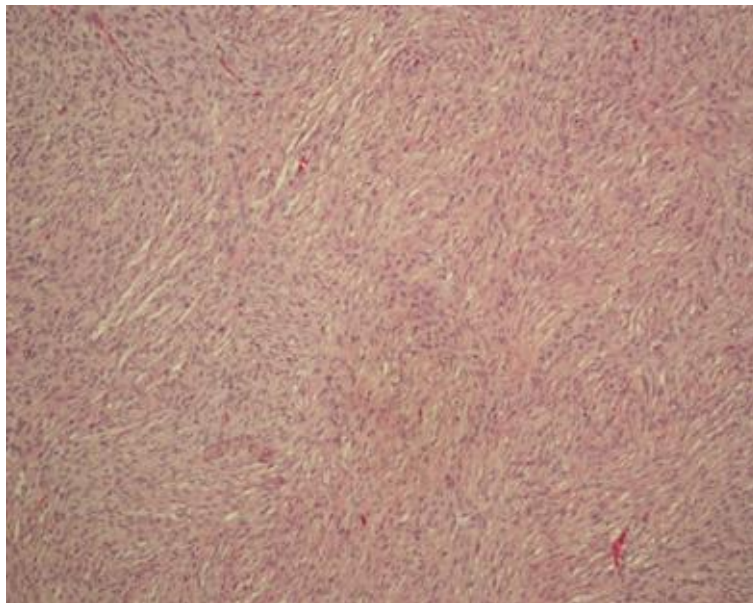


Figure 2: Spindle-cells arranged in storiform pattern and have eosinophilic cytoplasm in general, no necrosis or significant nuclear atypia.

The postoperative course was uneventful and managed in accordance with enhanced recovery after surgery (ERAS) principles. The patient was discharged on the 3rd postoperative day and was followed in the outpatient departments.

Operative and pathological data was reviewed in the MDT meeting, in which the need for adjuvant local or Systemic treatment was

deemed unnecessary due to the lack of pathological markers of neoplastic aggressiveness or marginal involvement. Enrollment of the patient in the sarcoma surveillance program was advised. The patient reports high esthetic satisfaction and has remained disease free at one year postoperatively with a well-healed incision as seen in Figure 3.



Figure 3: Post operative result at 1 year.

Discussion

Anterior abdominal wall is defined as the area bounded by the costal margin and the inguinal ligaments cranio-caudally, and the anterior Axillary lines dextrosinistrally.¹⁰ It is a common site for DFSP; being the site in 25-46 % of cited cases in published case series. The hypogastric location is twice as common as the epigastric one. Border tumors extending to the costal margins, groins and flanks represent a small proportion of anterior abdominal wall lesions [11-13].

Delay in the diagnosis of DFSP is common, this maybe attributed to the indolent nature of this neoplasm and the fact that most patients report no symptoms. This delay in diagnosis is more common in truncal DFSP compared to other locations, which may explain the relatively larger sizes of the truncal, including abdominal, tumors at presentation [4]. This, together with the need to achieve a three-dimensional excision with a minimal safety margin of 2-3 cm, usually

results in a significant skin and soft tissue loss that requires resurfacing [10,11].

The topography of DFSP has an important therapeutic and prognostic implications. Cervical location is associated with the worse prognosis as it has the highest recurrence rate, this has been linked to a higher rate of fascial infiltration at presentation, on the other hand anterior abdominal wall lesions rarely infiltrate down to the deep fascia [7]. The variation in the rate of fascial involvement is inversely Proportional to the thickness of Subcutaneous superficial fasciae. Topography also dictates the preferred reconstructive strategy of the resultant post excisional defects [7,11,13].

The micrographic surgical serial excision (MS) has been employed with the aim of tissue conservation, this technique may obviate the need for complex reconstruction and facilitate primary closure of defects [14-16]. However, this technique has several limitations which include its availability, complexity and questionable suitability to

large lesions (lesions with diameter >10cm), in which significant tissue loss and disfigurement may still be faced and reconstruction required, even with MS employed [10,17].

Reconstructive options are dictated by many factors, but ideally any chosen method should aim for maximizing esthetic and functional gains for the patient and minimizing possible complications, complexity and multiplicity of procedures [11,18].

Abdominoplasty is a relatively simple, single site, synchronous and immediate reconstructive procedure in the context of anterior abdominal wall soft tissue tumors in general and DFSP in particular [19,20]. The coverage it provides matches the tissue loss and is more durable than split thickness skin grafts [20].

The covering flap is contiguous to the defect, thus obviating the need for a donor site management. The procedure is versatile in design and is modifiable and extensible.20 Reversed abdominoplasty can address epigastric soft tissue tumors in a similar fashion to classical abdominoplasty for infra umbilical ones [18,21].

Hybrid abdominoplasties can be planned for borders lesions. Defects resulting from the excision of upper abdominal wall lesions crossing the costal margins can be resurfaced by reversed abdominoplasty and bilateral thoraco-epigastric flaps [20,21]. On the other hand, in defects from lesions extending down to the groin combining classical abdominoplasty with pedicled thigh flaps may be possible [19,22].

Conclusion

Early diagnosis of DFSP and the employment of MS may minimize excisional tissue loss, thus allowing for primary closure of resultant excisional defects. However, excision of large lesions, especially in the abdominal wall results in significant tissue loss regardless of the excisional technique. For anterior abdominal wall lesion, abdominoplasty should be considered for resurfacing resultant defects; the procedure is versatile, extensible and can be combined with other reconstructive procedures for a durable and esthetically satisfying results.

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Declaration of Conflicting Interests

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Ethics approval

Our institution does not require ethical approval for reporting individual cases or case series.

Consent

A verbal and a written consent were taken from the patient directly prior to publishing information or images.

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