

Case Report

Unusually Large Dermatofibrosarcoma Protuberans Involving Entire Abdominal Wall Reconstructed with Bilateral Large TFL and Groin Flaps: A Case Report

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ABSTRACT

Dermatofibrosarcoma Protuberans (DFSP) is a rare low grade soft tissue tumor with high recurrence rates. DFSP represents 1% of all soft tissue tumors. Recurrent DFSP requiring an entire abdominal wall reconstruction poses a challenge to the reconstructive surgeon.

Case Report: Here we report a large recurrent anterior abdominal wall DFSP in a teenage girl involving the anterior rectus sheath and the deep fascia of the abdominal wall (20 x 17 x 9 cm) posing difficult reconstructive challenge. The reconstruction was done by B/l tensor fascia lata flap and extended right groin flap.

Discussion: A large composite anterior abdominal wall defects are usually reconstructed by free flaps, but the more common local flaps can also be equally effective.

Conclusion: Complete surgical excision for DFSP of the abdominal wall, which result in defects of entire abdominal wall can be reconstructed with simpler local pedicled flaps.

Key Words: Dermatofibrosarcoma, Abdominal wall Reconstruction, Tensor Fascia Flap, Groin Flap.

INTRODUCTION

Dermatofibrosarcoma Protuberans (DFSP) a low grade soft tissue tumor of fibroblast origin, it is a locally aggressive tumor with high recurrence rates. [1] DFSP annual incidence of 0.8 to 4.5 per million worldwide. It accounts for less than 5% of soft tissue tumors and 0.1% of malignancies. [2,3]

DFSP most commonly affects trunk and proximal extremities, [4] it is more common in men than in women in the third decade but rarely occurs in childhood. [5] Darrier and Fernand were the first to recognize DFSP as a clinicopathological entity and Hoffman was the first to introduce the term "DFSP". [6,7]

CASE REPORT

A 17yr old female presented with history of recurrent swelling of anterior abdominal wall which was previously excised twice previously; reports not available. On clinical examination multiple lobulated lesions noted in the anterior abdominal wall, three of which were of significant size largest measuring 20 X 17 X 9 cms, with ulceration of the overlying skin. Multiple small swelling noted extending from umbilical region to suprapubic and to bilateral anterior superior iliac spines. The swellings were freely mobile over deep fascia. No lymphadenopathy noted. [fig - 1]



Figure 1: Pre-Operative picture.

On MRI intensely enhancing well defined lobulated mass lesions along the subcutaneous plane of anterior abdominal wall was noted. Trucut biopsy revealed features suggestive of DFSP.

Considering the size of the swelling, young age and the expected defect; plan was made to harvest bilateral tensor fascia lata (TFL) flap and right groin flap, as the size of the expected defect would be 30 X 20cms delay of the left TFL (17X25cms) and right groin(15X12cms) flaps were done.[fig – 2,3]



Figure 2: Delayed Right Groin Flap.



Figure 3: Delayed Left Tensor Fascia Lata Flap.

One week later patient was taken up for definitive surgery where the deep fascia showed signs of involvement hence the entire anterior abdominal wall up till the muscular layer was resected [fig – 4].

Bilateral groin flaps measuring 17X25 cms left and 12 X 20cms on the right were raised along with the right groin flap measuring 15 X 12 cms to cover the defect. [fig – 5,6,7]



Figure 4: Intra-Operative Picture after excision.



Figure 5: Intra-Operative picture after inset of flaps



Figure 6: Intra-Operative picture, right lateral view.



Figure 7: Intra-Operative picture, left lateral view.

Histopathology revealed neoplastic lesion in dermal and subdermal layers having spindle to round cells having large pleomorphic spindled to round nucleus, and cells arranged in fascicles and whorls. The finding is consistent with DFSP. Patient had epidermal necrosis of the tip of left TFL flap which was graft with STSG and 6months post op patient had complete healing of all the wounds without any recurrences or break down of wounds.[fig 8]



Figure 8: 6 months Post-Operative picture

DISCUSSION

Abdominal wall sarcomas are rare entity and are difficult to treat due to the need of varied reconstructive measures after wide local resection. 2 to 3cms wide negative margins are thought to be adequate for resection; failure to achieve negative margin leads to recurrence rates as high as 60%.^[8]

DFSP are usually small < 5 cms but Bowne et al. reported 3% with large tumors (≥ 10 cm).^[9] in our case we are reporting a large DFSP involving umbilical, paraumbilical, iliac and suprapubic regions measuring 20 x 17 x 9cms. Various methods have been described for abdominal wall reconstruction including skin grafts, abdominoplasty technique,^[10] ALT free flap,^[11] mesh reconstruction, etc.

Here we have reconstructed unusually large abdominal wall DFSP with extended ALT and groin flaps, proving that well known and long practiced principles of plastic surgery can be applied to solve most complex of problems in easy and efficient way; even in times where microsurgical techniques predominate.

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