

## Case Report

# Dermatofibrosarcoma protuberans of perianal region: a rare case report

Subbiah Shanmugam\*, Radha Siva Bharath

Department of Surgical Oncology, Government Kilpauk Medical College, Royapettah Government Hospital, Chennai, Tamil Nadu, India

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### \*Correspondence:

Dr. Subbiah Shanmugam,

E-mail: [radha.bharath2@gmail.com](mailto:radha.bharath2@gmail.com)

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## ABSTRACT

Dermatofibrosarcoma protuberans (DFSP) is a rare locally aggressive slow growing soft tissue sarcoma with very low metastatic potential with high local recurrence rates. We presented a rare case of DFSP of left perianal region ulcerating into anal canal, we did wide excision with 3 cm margin along with abdominoperineal excision of rectum and permanent colostomy, defect is reconstructed with left pedicled antero-lateral thigh flap. DFSP has high chance of recurrence, it should be excised with at least 2-3 cm 3D margin and patient should be followed up for a long time.

**Keywords:** DFSP, Abdominoperineal, Soft tissue sarcoma

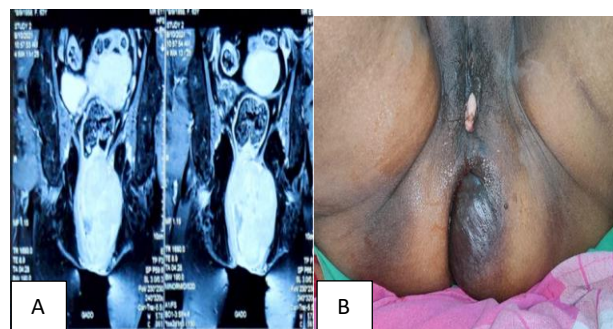
## INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) is a rare and slow-growing fibrohistiocytic neoplasm. DFSP is a low-grade soft tissue sarcomas which is locally aggressive and very low metastatic potential.<sup>1</sup> Most frequently it is seen at trunk, proximal extremities, head and neck (90%). Incidence of perianal <1%.<sup>4</sup> It affects adults between 2<sup>nd</sup> and 5<sup>th</sup> decade of life. Local recurrence after incomplete resection is common. In histopathology, tumor is located in dermis and consists of uniform fusiform cells, densely grouped with elongated nucleus, showing strong positive reaction to CD34 and negative to S-100 and desmin.<sup>6</sup> This DFSP case reported for its rarity of site.

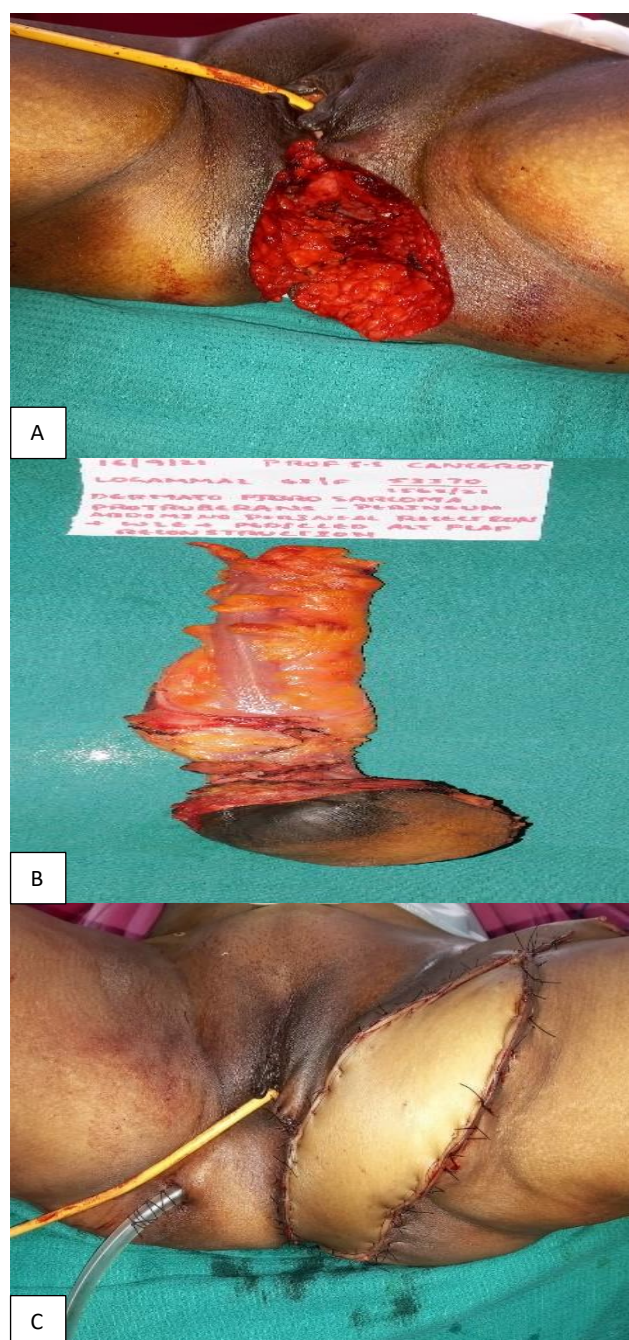
## CASE REPORT

We report a case of 63-year-old women presented with a perianal lump that had gradually progressed in size over 6 months and attained present size, she complains of pain at site of swelling. On examination patient had a 10×10 cm lobulated mass lesion at left perianal region leading to pressure ulcer into anal canal 1 cm from anal verge on

left side. Upper border felt at 5 cm from anal verge. Histopathology showed monomorphic fusiform cell neoplasm, in a 'swirled' focal arrangement, infiltrative of dermal tissue, compatible with DFSP. MRI of pelvis showed clinical exam. Patient underwent excision of lesion with circum-ferential margins of 3 cm along with abdomino-perineal resection of rectum through laparotomy, permanent colostomy. Defect reconstructed with left pedicled antero-lateral thigh flap. Histology confirmed DFSP (grade 2). All margins were free.



**Figure 1 (A and B): MRI showing DFSP at left perianal region and clinical picture.**



**Figure 2 (A-C): Defect after resection, resected specimen and clinical picture after reconstruction.**

## DISCUSSION

DFSP is a rare and locally aggressive dermal mesenchymal tumor. It is a sarcoma of cutaneous origin corresponding to less than 1% of all soft tissue sarcomas, with an annual incidence of 0.8-4.5 cases per million individuals.<sup>1</sup> It is a slow growing tumor, with a high recurrence rate due to its strong capacity to infiltrate subcutaneous tissue, fascia and underlying muscle in the form of pseudopods.<sup>1</sup> It presents as asymptomatic multinodular bluish or brownish erythematous plaque, developing over years, with its typical 'protuberant' aspect.<sup>1</sup> It can also present as an atrophic plaque,

resembling morphea, and is often misdiagnosed as such. In 1983 Frierson and Cooper reported a case of the rare DFSP myxoid variant.<sup>2</sup> Hong et al also reported similar case of this clinical form which is rarely documented in literature, with prominent myxoid stromal alterations.<sup>3</sup>

The most common DFSP sites are the trunk and extremities. It is unusual above the neck.<sup>4</sup> Perianal DFSP are very rare and only few cases are documented in the literature (less than 1%). The proportion of involvement between men and women is 1:1. DFSP mainly occurs between the 2<sup>nd</sup> and 5<sup>th</sup> decade of life.<sup>4</sup> There are reports of lesions arising in areas that has suffered previous trauma or in patients who underwent previous surgeries.<sup>5</sup> Our patient had no such history of trauma or surgery. In general, the tumor is located in the dermis, but it can sometimes present as infiltrative growth in the subcutaneous fatty tissue, forming a pastry pattern (in 60% of cases; neoplastic cell bands parallel to the epidermis) or a honeycomb pattern (where delimitation of adipocyte between the tumoral tissue).<sup>1</sup> Its low potential for metastasis is typical, with less than 2% presenting with regional or distant metastases, most often metastasis restricted to lungs and less frequently to lymph nodes.<sup>4</sup>

Differential diagnoses can be recurrent dermato-fibroma, hypertrophic scars, skin manifestations of myofibroblastoma, keloid, metaplastic carcinoma, fibromatosis.<sup>1</sup>

Histopathology reveals uniform densely grouped fusiform cells, with elongated nuclei without much cytologic pleomorphism arranged in characteristic storiform pattern.<sup>3</sup> The degree of nuclear atypia is higher in nodular lesions than in plates. Fibrosarcomatous focal alterations are occasionally observed in DFSP.<sup>4</sup> Immunohistochemical findings are positivity to CD34 in 84-100% and vimentin (related to the fibroblastic nature of the tumor), and negative to markers such as S-100, HMB45, desmin and actin. The characteristic pattern is positive for CD34 and negative for factor XIIIa, Stromelysin-3 (dermatofibroma marker), CD117.<sup>6</sup>

The standard treatment for a localized disease is wide local excision with recommended margins of 2-3 cm, a three-dimensional resection including skin, subcutaneous tissue and underlying fascia is needed to prevent recurrence.<sup>4</sup> In our case in order to achieve clearance we did abdomino perineal resection of rectum as lesion is ulcerating into the anal canal with involvement of sphincter. The local recurrence rate will decrease with the increase of surgical margins.<sup>4</sup> This procedure can generate cosmetic deformities and even functional loss. In our case we reconstructed the defect with left pedicled antero lateral thigh flap. The factors associated with high rates of recurrence are histological subtype, size, location (difficulty to give good margin), cellularity and high mitotic rate.<sup>6,7</sup>

There are current reports about use of Mohs micrographic surgery as a first-line therapeutic measure in cases of

limited tumors for tissue preservation and reduction of recurrence rate.<sup>8-11</sup> The 90% of DFSP cases present a translocation in chromosomes 17 and 22, leading to changes in the quantity of platelet-derived chain growth factor, resulting in the activation of platelet-derived growth factor receptor and act as a stimulus for tumor growth. Imatinib mesylate is a selective inhibitor of tyrosine kinases, which include platelet derived growth factor receptor. It inhibits the growth of tumor cells and the transformation of fibroblasts. Clinical studies have shown a positive response in 65% of patients who used the drug, so it can be considered as an alternative for patients with non-resectable, recurrent/ metastatic lesions.<sup>4,8,12</sup> There is need to study the role of imatinib in neo adjuvant setting where location of DFSP is difficult to get a 3D R0 resection without functional loss.

Radiation therapy can be used as adjuvant therapy in cases where adequate surgical margins are not easily reached, or in cases of positive margins, even after maximum resection. It is also indicated for patients with inoperable macroscopic lesions.<sup>4,12</sup> Postoperative radiotherapy has been associated with a cure rate of more than 85%.<sup>13</sup> However, it increases risks of subsequent development of other skin tumors.<sup>12</sup>

Since the tumor projects in multiple directions, reaching deep structures, not even wide excision can prevent residual tumor in single or multiple focus, and this would explain lesion recurrence.<sup>12</sup> Recurrence mostly appears within 3 years, with half of them occurring in the first year after resection. New lesions appear even after 5 years of completion of treatment, which justifies the need for biannual clinical and patient monitoring for an extended period after surgery.<sup>4,12</sup>

## CONCLUSION

Rarity of DFSP made us to review the most important aspects besides presenting as an interesting and atypical case with ulceration into anal canal. Narrow confines of perineum may not allow three-dimensional wide excision or compartment excision. One must understand about high chance recurrence even when excised with wide margins. For this reason, these patients must be observed periodically after surgery for a long time.

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## REFERENCES

1. Pérez OG, Righetti R, Woscoff A, Amante H. Case for diagnosis. Dermatofibrosarcoma protuberans. A Bras Dermatol. 2010;85:245-7.

2. Frierson HF, Cooper PH. Myxoid variant of dermatofibrosarcoma protuberans. Am J Surg Pathol. 1983;7:445-50.
3. Hong YJ, Choi YW, Myung KB, Choi HY. A case of myxoid dermatofibrosarcoma protuberans. Ann Dermatol. 2011;23:379-81.
4. Dragoumis DM, Katsohi LA, Amlianitis IK, Tsiftoglou AP. Late local recurrence of dermatofibrosarcoma protuberans in the skin of female breast. World J Surg Oncol. 2010;8:48.
5. McKee PH, Calonje JE, Granter S. Pathology of the Skin, with Clinical Correlations. Philadelphia, Elsevier-Mosby. 2005;2.
6. DuBay D, Cimmino V, Lowe L, Johnson TM, Sondak VK. Low recurrence rate after surgery for dermatofibrosarcoma protuberans: a multidisciplinary approach from a single institution. Cancer. 2004;100:1008-16.
7. Goldblum JR, Tuthill RJ. CD34 and factor-XIIIa immunoreactivity in dermatofibrosarcoma protuberans and dermatofibroma. Am J Dermatopathol. 1997;19:147-53.
8. Rutkowski P, Wozniak A, Switaj T. Advances in molecular characterization and targeted therapy in dermatofibrosarcoma protuberans. Sarcoma. 2011;2011:959132.
9. Snow SN, Gordon EM, Larson PO, Bagheri MM, Bentz ML, Sable DB. Dermatofibrosarcoma protuberans: a report on 29 patients treated by Mohs micrographic surgery with long-term follow-up and review of the literature. Cancer. 2004;101:28-38.
10. Xiang Q, Lu W, He X, Li H. Surgical treatment of dermatofibrosarcoma protuberans using wide local excision combined with Mohs micrographic surgery. Zhongguo Xiu Fu Chong Jian Wai Ke Za Zhi. 2011;25:1350-3.
11. Galimberti G, Montañó AP, Kowalczyk A, Ferrario D, Galimberti R. Outcomes in 11 patients with dermatofibrosarcoma protuberans treated with Mohs micrographic surgery. Int J Dermatol. 2012;51:89-93.
12. Bhambri S, Desai A, Del Rosso JQ, Mobini N. Dermatofibrosarcoma protuberans: a case report and review of the literature. J Clin Aesthet Dermatol. 2008;1:34-6.
13. Mendenhall WM, Zlotecki RA, Scarborough MT. Dermatofibrosarcoma protuberans. Cancer. 2004;101:2503-8.

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