CASE REPORT

Fibrosarcomatous dermatofibrosarcoma protuberans: A case report of an aggressive soft tissue sarcoma

Raghunath Prabhu,¹ Neha Kumar, Sakshi Sadhu, Rajgopal Shenoy

Kasturba Medical College, Manipal, Manipal University India, ¹University Hospital of North Staffordshire, London

Correspondence address: Dr. Raghunath Prabhu, Kasturba Medical College, Manipal, Manipal University, India. E-mail: drraghu81@yahoo.co.in

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ABSTRACT

Dermatofibrosarcoma protuberans is a rare soft tissue neoplasm arising from mesenchymal cells, making up about 6% of all soft tissue tumours. Often found on the scalp, neck, trunk and extremities, it has an intermediate-to-low-grade malignancy potential except the fibrosarcomatous variant (high-grade), which is more likely to metastasize than other types. Here we present the case of a 46-year-old male who presented with an erythematosus swelling on his upper back. MRI scan suggested a mesenchymal mass, which was then excised. Histopathology was reported as fibrosarcomatous variant of dermatofibrosarcoma protuberans. He is receiving radiation at present.

Key words: Back, dermatofibrosarcoma protuberans, excision, sarcoma

INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) is a soft tissue malignancy arising from the skin dermis, making up about 6% of all soft tissue tumours¹ and can spread deeper into fat, fascia, muscle and bone. It usually recurs in the same location post excision and has a 5% chance of metastasis.² Histopathological classification of this mesenchymal tumour is made on its composition. The fibrosarcomatous variant is a rare type of DFSP (FS-DFSP), which tends to metastasize due to its aggressive nature. It expresses the markers CD34 and apolipoprotein-D, although how the fibrosarcomatous nature of this tumour influences prognosis is a topic of debate. Treating this malignancy is often a challenge for clinicians due to its unpredictable and recurrent nature. Our case is a rare occurrence as these high-grade tumours have a very low incidence.

CASE REPORT

A 46-year-old diabetic male presented to the outpatients department at a large teaching hospital in Southern India with a large swelling on his upper left back. The patient reported he has had this swelling for 10 years and slowly growing. The swelling then started to bleed (duration of 1 day) which prompted him to seek medical advice. He is a known insulin dependent diabetic. On physical examination, all his vitals were stable. He was of an average build and was not cachetic. On examination of the back, there was a 15 x 12 x 5 cm non-tender, round swelling on the left upper back [Figure 1]. It was firm in consistency, immobile with a lobulated surface and showed clearly defined borders. The skin overlying it appeared erythematosus with a superficial ulcer. There was no local rise of temperature. On admission, his blood results showed decreased sodium at 130.0 mmol/L, raised serum urea 29mg/dL, HbA1c 13%, all other parameters were normal. He had significant glycosuria as well. A fine needle aspiration of the swelling revealed stromal fragments and endothelial atypical cells with pleomorphic nuclei and prominent nucleoli but no definite diagnosis was established. An MRI scan of the spine showed a mesenchymal tumour in the subcutaneous plane of the left posterior chest wall.
There was no obvious infiltration into the underlying muscles or intraspinal/thoracic extension. An ultrasound of the abdomen showed hepatomegaly with grade 2 fatty infiltration. Subsequently the patient underwent a wide local excision under general anesthesia. A vacuum assisted dressing was applied on the wound and the patient made an uneventful recovery, following which he underwent split skin grafting.

Figure 1: A 15 x 12 x 5 cm non-tender, round swelling on the left upper back.

The specimen sent for histopathology was 1.12 kg and measured 21 x 21 x 9.5 cm, the margins were uninvolved. It was positive for CD34 and CD10. Based on the histopathological report, a diagnosis of sarcoma arising in the DFSP (FS-DFSP) was made [Figure 2].

Figure 2: Histopathological examination showing fibrosarcomatous variant of dermatofibrosarcoma protuberans (A-dermatofibrosarcoma protuberans showing storiform pattern, B-fibrosarcomatous differentiation with herring bone-like growth pattern, C-tumour infiltrating between lobules of fat, D-tumour with area of necrosis).

DISCUSSION

Darier and Ferrand first described DFSP in 1924[3], but further work by Taylor and Helwig allocated a typical microscopic appearance to this clinical entity in 1962.[4] Several variants of DFSP have since been described, from myxoid and atrophic types to sclerosing DFSP and a granular cell variant. Most of these display similar characteristics to each other. The only exception to this fact is the FS-DFSP. These are relatively rare and tend to be more aggressive. These appear as >5% of the lesion consisting of undifferentiated pathology. FS-DFSP is characterized by spindle cells seen in a herringbone-like growth pattern. About 50% of these lesions test positive for the tumor marker CD34 (still less than in cases of DFSP). Apolipoprotein D (Apo D) is a glycoprotein expressed in DFSP and can also be used an indicator of disease.[5]

DFSP and its variants are usually problematic solely due to the morbidity caused by repeated operations, it rarely metastasizes (about 5% of the cases). FS-DFSP is much more aggressive than other types and many reports have suggested that both local recurrences and distant metastases are relatively higher in this variant.[6] However, this is thought to be dependent on the status of the excised margins. If wide excisions are one with free margins, local recurrences tend to be much rarer along with distant metastases (about 22%).[7] Nonetheless, the effect of the presence of fibrosarcomatous changes in FS-DFSP on the prognosis remains a topic of wide debate, with some authors advocating that the changes lead to poorer prognosis in themselves, regardless of the margin status. In our case, the margins were clear of tumor, hence, it will be interesting to see how this patient progresses and if he develops any recurrence. Another point of difference is FS-DFSP usually has a female preponderance compared to other types of DFSP which tend to affect males more.

Our patient was positive for both the tumor markers, Apo D and CD34, but a study by Palmerini et al suggests that these are expressed less in FS-DFSP than DFSP.[8] Surgical excision is the only curative option currently available with the Mohs micrographic surgery being employed quite often these days.[8] Radiotherapy and chemotherapy
(particularly in FS-DFSP) have also shown to play a role in controlling local recurrence.\textsuperscript{[9]}

Local recurrence rates with FS-DFSP are 75-90%, with distant metastases occurring in 15-65% cases.\textsuperscript{[10]} It has a significantly poorer prognosis than classical DFSP. The presence of myoid differentiation in DFSP or FS-DFSP is also rarely observed, not seen in our patient. Overall it is thought that patients with DFSP have a better prognosis than those with FS-DFSP, therefore our patient’s prognosis is likely to be dismal and he has to be on regular follow-up.

**CONCLUSION**

FS-DFSP is a rare form of DFSP, which can lead to significant morbidity and a reduced survival rate. The tumor’s aggressive nature combined with its presence in patients for a long duration contributes towards this. Currently surgical excision is the only modality with a curative intent, with the role of adjuvant chemotherapy being studied. Our case is one such rare occurrence and we hope to build on existing reports of this condition to help better educate clinicians on this soft tissue neoplasm and its management.

**REFERENCES**


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