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Case Series

Dermatofibrosarcoma protuberans: case series and literature review at the rivers state university teaching hospital, port Harcourt, Nigeria

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ABSTRACT

Dermatofibrosarcoma protuberans, first described by Jean Darier and Marcel Ferrand in 1924, can affect various body regions, including the head and neck, trunk and limbs. This case series aimed to document the occurrence of this rare dermatofibrosarcoma protuberans as seen at the Rivers State University Teaching Hospital and the neighbouring referring private clinics over five years (2019-2024). The clinical case notes of patients with dermatofibrosarcoma protuberans were retrieved and data were collected and analysed for the study. Eight dermatofibrosarcoma protuberans cases were observed in the study period. The average age of the patients was 43.9 years, with five females and three males. Most of the patients presented with large, locally advanced, ulcerated masses. All but one underwent wide local excision with a minimum excision margin of 3 cm, followed by chemotherapy. A few dermatofibrosarcoma protuberans were recorded at our centre within the period under study. Wide local excision and adjuvant chemotherapy were offered and adjuvant radiotherapy was recommended but not done.

Keywords: Case series, Dermatofibrosarcoma protuberans, Literature review, Nigeria, Port Harcourt

INTRODUCTION

Dermatofibrosarcoma protuberans is a soft tissue sarcoma arising from the dermis of the skin.[1] It is typically slow-growing, protuberant, with a nodular appearance. It exhibits a tendency for local recurrence and accounts for 1% of all malignancies and 1% of soft tissue sarcomas.[2] Additionally, it is regarded as the most common cutaneous sarcoma with an incidence of 0.8–4.5 cases per one million persons.[2,3]

Although the tumour was first recognized as a separate entity in 1890 by Sherwell and Taylor, it was first formally described in 1924 by Jean Darier and Marcel Ferrand and named "progressive and recurring dermatofibroma".[4,5] It is hence often referred to as darier ferrand dermatofibrosarcoma (DFSP), the same acronym for Dermatofibrosarcoma protuberans.[6,7] The latter terminology dermatofibrosarcoma protuberans was, coined in 1925 by Hoffmann, to reflect the clinical presentation of the disease and this tumour can occur in any region of the body where there is skin.[8]

The primary cell of origin of DFPS are probably fibroblasts or histiocytes of the dermis of the skin soft-tissue.[4,9,10] Perineural cells have also been suspected as likely precursors and genetic abnormalities have been described where in 90% of cases a genetic abnormality

resulting in reciprocal translocations of chromosomes 17 and 22: t(17;22) (q22;q13) is present.[11,12]

While there are no known risk factors, trauma is associated with about 10%– 20% of DFSP cases, especially in older patients.[1] The tumour often begins as a painless, firm, slow-growing mass.[13,14] The lesion may progress to mimic other skin lesions or become locally invasive, metastasis is late and uncommon. Due to its rarity, the diagnosis is often missed.[4,15] DFSP relies on clinical and histopathological features. Key characteristics include spindle cells arranged in a storiform or cartwheel-like pattern, minimal cytological atypia, uniform nuclei, low mitotic activity and intense staining for CD34 and vimentin.[4,9] In addition, Polymerase Chain Reaction (PCR) detection of gene fusion events may aid in diagnosis.

Like other cancers, a multidisciplinary evaluation and a combination of treatment approaches are essential for achieving better outcomes. The first line of treatment for dermatofibrosarcoma protuberans (DFSP) is surgical excision. This is often combined with radiotherapy (RT) and/or systemic treatment, such as imatinib mesylate, which can be used in both neoadjuvant and adjuvant settings.[3,16]

Modern surgical options comprise wide local excision with a recommended safety margin of at least 3 cm using standard 2D surgical excision; where available, Mohs Micrographic Surgery (MMS) is preferable; and surgery followed by three-dimensional complete circumferential and peripheral deep margin assessment (or CCPDMA).[4,17-19]

Radiotherapy is recommended for DFSP as adjuvant therapy in the presence of risk factors: close margins, recurrent tumors, aggressive histological subtypes or as salvage treatment.[3] A report of this rare tumour will increase awareness for improved patient care. This case series aimed to document the occurrence of rare dermatofibrosarcoma protuberans as seen at the Rivers State University Teaching Hospital and neighboring clinics over five years (2019-2024).

CASE SERIES

Case 1

36-years-old female, who presented with three-month history of a recurrent mass in the right gluteal region of three months duration. The previous mass was excised in a peripheral hospital and the histology revealed that it was a fibroma. Physical examination revealed presence of a right gluteal mass measuring 14 cm by 10 cm, with nodularity and areas of ulceration and contact bleeding. There was no regional lymph node enlargement and the histopathology showed dermatofibrosarcoma. She had wide local excision with excision of deep fascia and part of gluteal muscle.

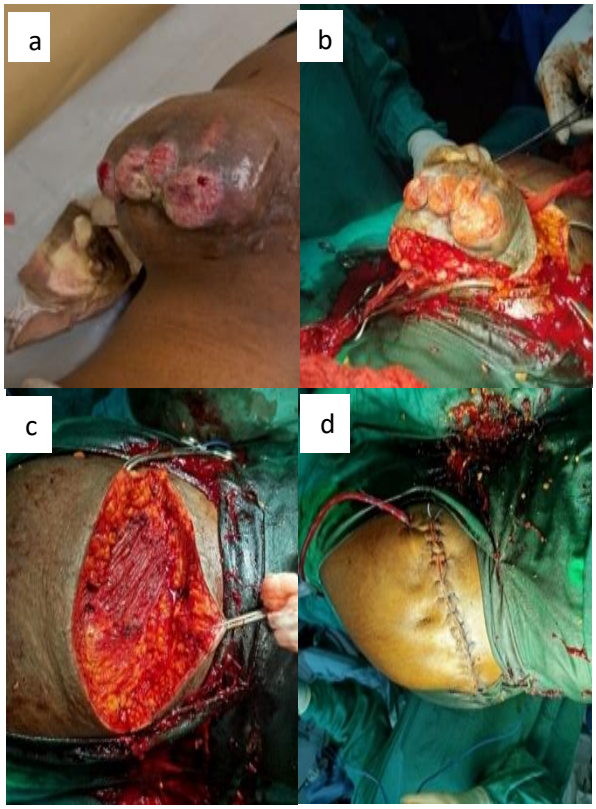

Figure 1 (a-d): Pre-operative and intra-operative clinical photograph of case 1 mass.

Case 2

A 39-years-old male who presented with a year history of a growth on the right lower part of the abdomen measuring 16×18 cm. The patient was unable to do requested preoperative imaging studies (Abdominal magnetic resonance imaging/computerized tomography). He was offered wide local excision with 3 cm resection margin. Intraoperative findings revealed involvement of anterior abdominal wall deep fascia and the external oblique aponeurosis, which were excised along with the tumour and a mesh was used to cover the defect.

The histopathology of the tumour revealed dermatofibrosarcoma. Postoperatively the patient received only a single cycle of chemotherapy and declined/absconded from further treatment on the ground that he was informed that it was “poison” which has to be treated in a “native way”.

Case 3

A 31-years-old married woman and mother of 5, who was seen in a medical outreached organized by the Rivers State University Teaching Hospital, presented with a huge ulcerated anterior abdominal wall mass of nine months duration. It was said to have started on the skin like a boil and ulcerated over time requiring dressing. It was hard and nodular, bilobed, measuring 18 cm by 14 cm.

There was no regional lymph node enlargement and no other significant findings. The mass was excised with a wide margin 3 cm and there was no involvement of the anterior abdominal wall fascia. The wound healed primarily. However, the patient had not been seen for follow up of tissue histopathology findings despite several efforts.

Figure 2 (a-c): Pre-operative and intra-operative clinical photograph of case 2 mass.

Case 4

A 72-years-old man had presented with huge fungating mass at the right posterior trunk with a histology report showing dermatofibrosarcoma protuberance, indicating that there had been a recurrence after previous resection elsewhere (at a peripheral center).

Further probing revealed that he did not receive complete chemotherapy and did not have radiotherapy during the previous therapy.

He was commenced on imatinib to which there was an initial response, but patient absconded and represented after six months in coma with cerebral metastasis as confirmed by brain computed tomography scan.

Figure 3 (a-c): Pre-operative and intra-operative clinical photograph of case 3 mass.

Case 5

A 68 years old man who presented with a left breast mass of a year duration. Examination revealed a hard nodular measuring 12×13 cm. He had wide local excision with 3 cm resection margin, with flap closure. He has been free of recurrence for 4 years.

Case 6

A 43-years-old woman who presented with an ulcerated left chest wall mass of 8 months duration.

The mass measured 10×8 cm and had overlying ulcer 4×3 cm. She was offered wide local excision with 3 cm resection margin and has been free of recurrence for 7 years.

Case 7

26 years of female with a mass at the left thigh of 9 months duration. The dimensions of the mass 14×12 cm. She had wide local excision with 3cm resection margin and has been free of recurrence for 4 years.

Table 1: First five cases with the clinical features and outcome.

Cases	Age	Sex	Presentation and location of mass	Duration of symptoms (months) and size of mass	Treatment	Outcome / follow up
1	36	F	Ulcerated recurrent right gluteal	3 months / 14×10 cm	Wide Local Excision with Excision of deep fascia and part of gluteal muscle Awaiting adjuvant therapy	Two months
2	39	M	Ulcerated right lower part of the abdomen	10 months / 16×18 cm	Wide local excision with 3cm resection margin, anterior abdominal wall deep fascia / the external oblique aponeurosis+Mesh placement	Declined adjuvant treatment
3	31	F	ulcerated) anterior abdominal wall	9 months / 18×14 cm	Wide local excision with 3cm resection margin	Primary wound healing, lost to follow up
4	72	M	Recurrent fungating mass at the right posterior trunk	6 months 18×20 cm	Wide local excision with 3cm resection margin+Imatinib	Absconded after initial response. Re-presented in coma after six months with cerebral metastasis
5	68	M	Hard nodular left breast	1 year / 12×13 cm	Wide local excision with 3cm resection margin, Flap Closure	Free of recurrence after 4 years

Table 2: Last three cases with the clinical features and outcome.

Case	Age	Sex	Presentation and location of mass	Duration of symptoms (months) and size of mass	Treatment	Outcome / follow up
6	43	F	Ulcerated left chest wall	8 months /10×8 cm	Wide local excision with 3 cm resection margin	Free of recurrence for 7 years
7	26	F	Left thigh	9 months / 14×12 cm	Wide local excision with 3 cm resection margin	Free of recurrence for 4 years
8	36	F	Anterior abdominal wall mass	24 months / 8×6 cm	Wide local excision with 3 cm resection margin	Free of recurrence for 6 years

Case 8

A 36-years-old woman who presented with anterior abdominal wall mass of 2 years duration. The mass measured 8×6 cm. She had wide local excision with 3 cm resection margin and has been free of recurrence for 6 years.

DISCUSSION

The eight patients described in this series had age range of 26–72 years, with a mean of 43.9 years. This finding is higher than the mean age of s 35.05±18.9 years reported in a histological spectrum of soft-tissue tumors in Makurdi Nigeria and the mean age of 39.0 years in a Kano, Nigerian study.[20,21]

In this case series, there were five females and three males, which contrasts with the commonly reported predominance of malignant soft tissue sarcomas in males. Most patients were from low socio-economic backgrounds, except for one middle-class patient with a gluteal mass. The duration of symptoms of the patients described were varied from 3 months to 24 months, with a mean of 10.1 months. The tumors were relatively large in size varying from 8×6 cm to 18×20 cm. Rapidity in growth associated with ulceration and no clinical evidence of distant spread suggests locally aggressive tumors of intermediate malignancy. The histologic findings of dermatofibrosarcoma in all the cases are therefore not surprising, aligning with the WHO 2020 classification of tumors of intermediate malignancy. However, this is not full proof, as it is not backed up with confirmatory tests for occult distant spread. The thigh/gluteal region (2), anterior abdominal wall (2) and posterior trunk (1) were the sites of occurrence of 75% of these tumors as reported in this study. Our finding is relatively similar to most of the cases reported in the trunk and limbs in a study in Calabar Nigeria.[22]

The histology of the tumor was dermatofibrosarcoma in all cases, each measuring more than 3-5 cm, indicating a high risk of metastasis.[23] However, there was no clinical

evidence of distant spread but for one patient who succumbed to brain metastases. The size of tumor reported in our study is larger than that reported from in China in 2010.[24] The size of tumors in this report is larger than some earlier reported studies, but comparable to some Nigerian studies.[22,23,25-27] In Nigeria, cancer treatment is challenging due to the required "out-of-pocket" payments for imaging and therapies like surgery and chemotherapy.[28-30] The treatment provided to these patients included wide local excision with a 3 cm resection margin, excision of the anterior abdominal wall deep fascia and the external oblique aponeurosis, followed by mesh placement, when necessary. None of the patients underwent radiotherapy and only two patients received only a few cycles of imatinib. The follow-up period for the patients ranged from two months to seven years and the outcomes during this time were also inconsistent.

CONCLUSION

Eight cases of large dermatofibrosarcoma protuberans were reported. The mean age of patients was 43.9 years, with a mean duration of symptoms was 10.1 months. There were relatively large tumor sizes in this series and a history of recurrence following previous treatment in two of the cases. All but one had wide local excision. Access to radiotherapy services was limited by patients' inability to afford them. Providing functional radiotherapy services will improve the care of these diseases.

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