

## Giant Dermatofibrosarcoma: A case report

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### SUMMARY

Dermatofibrosarcoma Protuberans (DFSP) is a rare skin tumour with high recurrences. World Health Organization report has shown it to be a slow growing, nodular neoplasm of intermediate grade malignancy that is found almost exclusively in the dermis from where it often invades the subcutaneous tissue. It is more common in adults than children, in blacks than whites and on the trunk than other parts of the body. Management is challenging due to its propensity to recur after local wide excision and for this reason, it is believed that combining either wide local excision with Mohs micrographic surgery or adjuvant chemotherapy or radiation therapy may reduce its recurrence. In this follow up case report, we present a 53-year-old man who had wide local excision of his recurrent facial DFSP 14 months post-surgery. His past medical history has shown that the man has had this tumour excised 13 times in other hospitals prior to his first presentation to us 14 months ago. In multiple recurrences with previous surgical scars in which free margin is doubtful as noticed in our case, adjuvant radiotherapy will be of benefit. This was recommended to the patient during his first surgery but could not have it done due to some logistics reasons.

**Keywords:** Recurrent, dermatofibrosarcoma, protuberance, adjuvant chemoradiation.

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### Introduction

DFSP is a slow growing, low-grade sarcoma of skin and the subcutaneous tissue with an incidence rate of 0.8 cases per million per year.<sup>1</sup> Sometimes this tumour may be left unattended for many years due to poor health seeking

behaviour. It was first described by Ferrand and Darier in 1924 as a skin tumour and was given its present name a year later by Hoffman.<sup>2</sup> Any part of the body can be affected with a larger proportion of them involving the trunk.<sup>3</sup> Some laboratory tests and radiological investigations are useful in managing these patients pre-operatively while Magnetic resonant imagery and Computerized Tomographic Scan may be useful especially in making the diagnosis of difficult cases with atypical clinical presentations.<sup>4</sup> Since the lesion has a high recurrence rate, wide local excision with or without Mohs' microsurgical operation is usually needed for a good prognostic outcome.<sup>5</sup> If this is not possible as reported in our case, postoperative adjuvant chemotherapy with imatinibmesylate or radiotherapy can be given to the patient to reduce the chances of recurrence.<sup>6</sup>

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### Case Summary

We present the case of a 53years old man, a retired long distant driver who presented with a recurrent DFSP 14months post excision. He first presented 14months ago with 29-year history of recurrent facial tumour for which he has had excision 14 times in various hospitals with no record of histological diagnosis. He was said to have had facial trauma from a fall with his face against a well brick while trying to fetch water in one of his driving trips. He later developed a facial tumour that was painless and slow growing and the size of a pea nut. For cosmetic reasons he went to a hospital where it was first excised. Since then the tumour had reoccurred 13 times in various sites of his face necessitating further surgeries. At first presentation we found a middle aged man with multiple facial tumours, multiple

facial scars, deviated nasal dorsum to the right side and partial obstruction of the nasal cavities with greatly reduced nasal patency. Of note was a rectangular scalp scar that suggested previous skin flap mobilization for possible wound repair and a skin donor site on the left thigh for possible skin cover in one of his surgeries as shown in Fig.1a below. He had various investigations done, which included computerized tomographic scan of the paranasal sinuses and brain that showed no brain, bone or sinuses involvement. Post excision biopsy as shown in fig. 1b and the histology findings had features consistent with that of dermatofibrosarcoma protuberans (Fig.1c). Patient was referred to another Healthcare facility for postoperative adjuvant radiotherapy.



Fig.1a (Pre-surgery 14months ago)



Fig. 1b (Post-surgery 14months ago)

Fourteen months later, he again presented with a huge ulcerating tumour that was foul smelling. About 50% of his mouth was covered by this tumour preventing him from feeding well per os (Figures 2a and 2b). His medical history has shown that he couldn't have the recommended adjuvant radiotherapy owing to financial constraints.

After a chest x-ray and haematological tests that were found to be within normal limits, he again had local excision and his histology findings were still in keeping with DFSP. His present state is stable (Fig.2c) and patient has again been referred for adjuvant radiotherapy.



Fig. 2a (Pre-surgery)



Fig.2b (Pre-surgery)



Fig.2 c (Post-surgery)

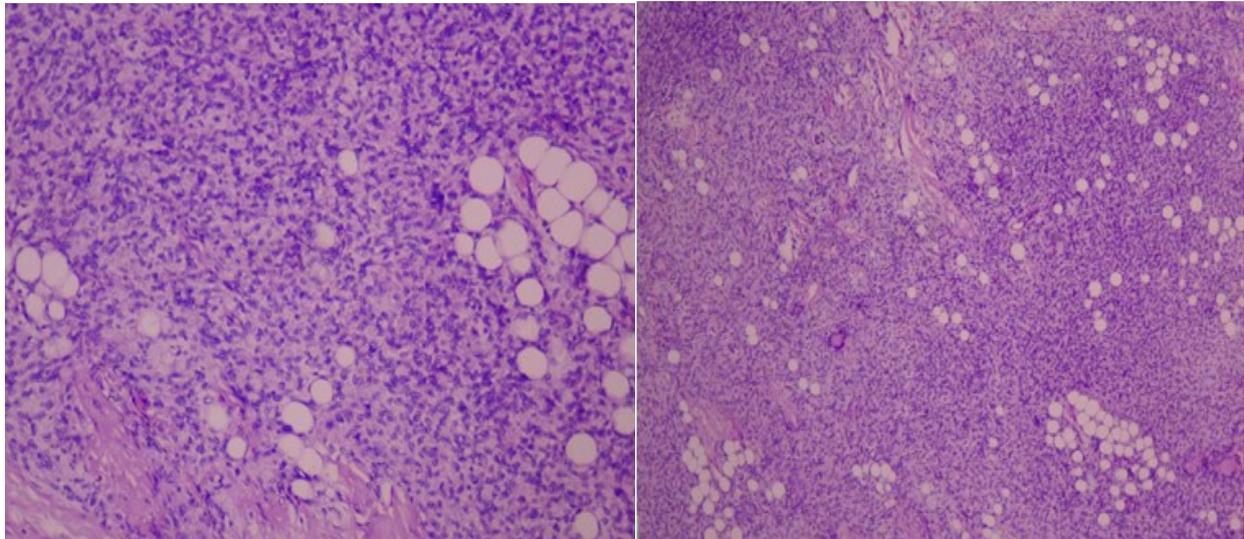


Fig. 1c: Histologic section of the lesion shows non circumscribed, highly cellular, tight storiform pattern of growth that infiltrates deeply into subcutaneous tissue and entraps fat cells to form characteristic honeycomb pattern. Areas of fascicular growth also noted

## Discussion

Dermatofibrosarcoma Protuberans (DFSP) is a skin tumour that is slow growing, nodular neoplasm of intermediate grade malignancy that is found in the dermis from where it invades the subcutaneous tissue.<sup>7</sup> It is also called borderline/ intermediate Fibrous Histiocytoma, most cases are seen mainly in adulthood but it can also be seen in infancy and childhood.<sup>8</sup> They are commoner in blacks and usually found on the trunk and various sites but rarely hand and feet.<sup>9</sup> As a group they are usually much larger than Dermatofibroma. The histogenesis of the tumour is not clear, it shows strong resemblance to pericytes and perineural cells as well as marked ultrastructural similarity to neurofibroma.<sup>10</sup>

Molecular changes include translocation t(17;22)(q21;q13) and platelet derived growth factor beta chain gene and both are seen in almost all cases using multiplex RT-PCR. Differential diagnosis include Dermatofibroma which is also storiform but non-infiltrative, less cellular than DFSP, factor XIIIa positive and CD34 negative; Malignant fibrous Histiocytoma with storiform pattern but shows moderate to severe pleomorphism with nuclear atypia.<sup>11</sup> Microscopically tumour shows non-circumscribed highly cellular cat wheel pattern permeating deeply into subcutaneous tissue and entraps fat to form distinctive honeycomb arrangement with mild nuclear pleomorphism and atypia, it may have numerous mitotic figures but not atypical ones.<sup>12</sup>

The other variant is the Bednarstumour/ Pigmented DFSP which is similar to DFSP but contain variable population of cells containing large amount of melanin pigmented.<sup>13-15</sup> Usually painless and slow growing that it may be ignored for years

without treatment.<sup>16</sup> History of trauma may be found prior to the development of this lesion in most cases and for recurrent ones, surgical scars, tumor ulceration, and pain may be found as reported in our case. Though low grade malignancy, some have been found to have metastasized to lungs in about 6% of cases.<sup>17</sup>

The treatment of choice is wide local excision with adequate tumor-free margins and this can be carried out by a dermatologist in the form of micrographic surgery or a Plastic Surgeon if skin grafting and reconstruction is needed or by an Otolaryngologist/Head and Neck Surgeon. In recurrent lesions, the use of paraffin sectioning and three-dimensional histological evaluation has been found useful in the management of the disease.<sup>18</sup> In case tumour free margin is not possible, adjuvant chemotherapy or radiotherapy or both should be used to minimize the rate of recurrence as reported in our case.<sup>19</sup> However, researches had shown that recurrence of the disease can still occur despite adjuvant radiotherapy or chemotherapy.

## Conclusion

In multiple recurrences with previous surgical scars, it may not be possible achieving a tumour-free margin during excision as reported in our case. However, patient must be educated on the need for a long term follow up to detect recurrence.

## Consent

An informed consent was taken from the patient to allow us publish this case report with the accompanying images. A copy of this has been submitted along with the manuscript for the Editor's perusal.



**Declarations****Acknowledgement**

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**Competing interests**

None to be declared.

**Authors' contributions**

SA Ogah performed the surgery, assisted in the manuscript preparation and initiated the idea to report the case. D. Awelimobor and O.O. Fadahunsi prepared the pathology slides, report them, did literature review and assisted in proofreading the manuscript for error in language and medical terms. OW. Adeyemi, EE. Agu and OO Ajiboye were part of the anaesthetist team that assisted in managing the patient pre, intra and postoperatively and also contributed to the literature search and review of the case.

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