Dermatofibrosarcoma Protuberans: A case report and review of the literature

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ABSTRACT

Background: *Dermatofibrosarcoma Protuberans is categorized as a skin tumour by WHO recent classification. It is slow growing, nodular neoplasm of intermediate grade malignancy that is found almost exclusively in the dermis from which it often invades the subcutaneous tissue. Most cases are seen mainly in adulthood but it can also be seen in childhood. They are commoner in blacks, usually found on the trunk and as a group; they are usually much larger than Dermatofibromas.*

Case Presentation: *We present the case of a 52year old retired driver with a 29year history of recurrent facial tumour. He had excision of the lesion14 times in various hospitals prior to presentation to us. He sustained facial trauma from a fall while trying to collect water from a well some months prior to the growth of the first tumour. At presentation we found a middle aged man with multiple facial tumours, multiple facial scars, nasal dorsum deviation with reduced patency. He had various investigations done, which include computerized tomographic scan of the paranasal sinuses and brain that showed no brain, bone or sinuses affectation. Excision biopsy was done and features on histology were consistent with those of dermatofibrosarcoma protuberans.*

Conclusion: *In multiple recurrences with previous surgical scars, it may not be possible achieving a good margin during excision as was noticed in our case. For such cases adjuvant radiotherapy is of benefit in lowering the rate of recurrence.*

Keywords: *Fibroma, dermatofibroma, fibrosarcoma, soft tissue sarcoma, Fibrous Histiocytoma.*

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 and with a high recurrent rates.[1,2] For this reason, the tumour may be left unattended for many years as the patient may not see anything wrong with the lesion. It was first described by Ferrand and Darier in 1924 as a skin tumour[3] and was given its present name a year later by Hoffman.[4]The tumor can occur in any part of the body with a larger proportion of them involving the trunk. With only clinical findings, it may be difficult to differentiate this tumour from similar nodular tumours like fibromas, dermatofibromas and histiocytomas.[5] However, the tumour size after excision and the histological appearance will give enough evidence in making the diagnosis. Variable laboratory and radiological investigations may be useful in managing these patients

depending on the site, size and extent of the tumour. Magnetic resonant imaging has also been found useful especially in making the diagnosis of difficult cases with atypical clinical presentations.[6] Since the lesion has a high recurrent rate, wide local excision whenever possible is usually needed for a good prognostic outcome.[7] If this is not possible as occurred in our case, post operative adjuvant radiotherapy should be given to the patient to lower the rate of recurrence.[8]

CASE REPORT

We present the case of a 52year old man, a retired long distant driver who presented with a 29yearhistory of recurrent facial tumours for which he has had excision 14 times in various hospitals. He had facial trauma prior to the first episode when he fell and hit his face on a well while trying to collect water to perform his ablutions for prayers. He later developed a facial tumour which started as a firm patch of skin approximately 1-5cm in diameter, flesh coloured, painless and slowly growing over several months to years before becoming araised nodule. For cosmetic reasons he went to a hospital where it was first excised. Since then the tumour had reoccurred 13 times necessitating further surgeries. At presentation we found a middle aged man with multiple facial tumours, multiple facial scars, deviated nasal dorsum to the right side and partial closure of the nasal cavities with greatly reduced nasal patency (Fig. 1). He had various investigations done, which include computerized tomographic scan of the paranasal sinuses and brain that showed no brain, bone or sinus affectation. Excision biopsy was done (Fig.2) and the removed masses (Fig. 3) were sent for histology. Histology report showed features consistent with dermatofibrosarcoma protuberans (Fig.4). He was subsequently referred for post operative adjuvant radiotherapy.

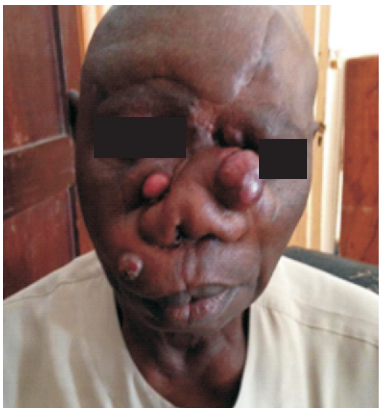

Fig.1 (before surgery)

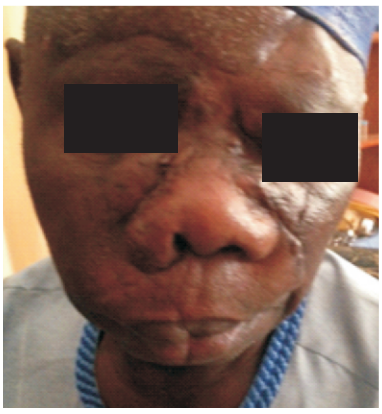

Fig. 2 (after surgery)

Fig. 3 (masses removed)

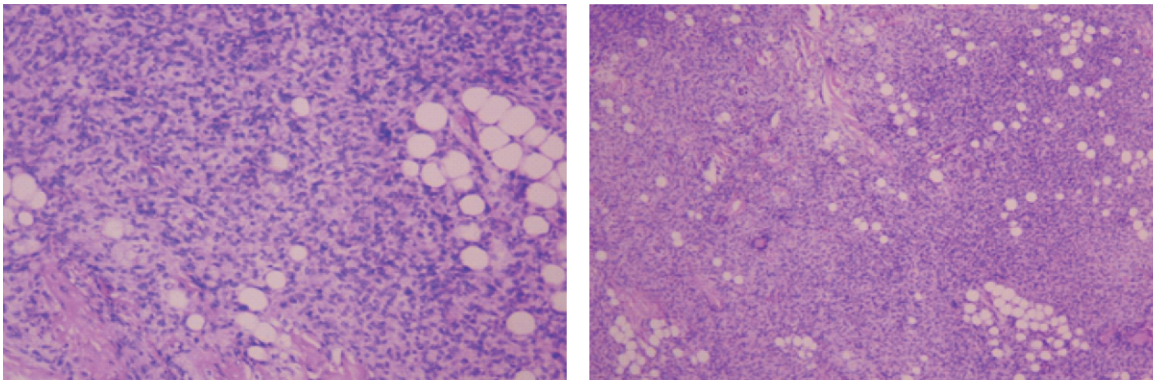

Figure 4: Histologic section of lesion shows non circumscribed, highly cellular, tight storiform pattern of growth that infiltrates deeply into subcutaneous tissue and entraps fat cells to form characteristic honeycomb pattern. Areas of fascicular growth also noticed.

DISCUSSION

Dermatofibrosarcoma Protuberans (DFSP) has varied prevalent rates as a skin malignancy. In Benin city it was found to be 3.7%, while in Oshogbo, Zaria, and Kano it constitutes 7.1%, 12% and 12.7% respectivelyof allskin malignancies.[9,10] It is slow growing, nodular neoplasm of intermediate grade malignancy that is found in the dermis from where it invades the subcutaneous tissue.[11]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.[12] Other variant includes Myxoid DFSP and Giant cell fibroblastoma. DFSP is commoner in blacks than white and usually found in various sites especially on the trunk.[13] As a group they are usually much larger than Dermatofibroma. The histiogenesis of the tumour is not clear, it shows strong resemblance to pericytes and perineural cells as well as marked ultrastructural similarity to neurofibroma.[14] Molecular changes include translocation (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.[15]

Microscopically, the 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.[16] 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.[17]Usually painless and slow growing that it may be ignored for years without treatment. History of trauma may exist 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 a low grade malignancy, some have been found to

have metastasized to the lungs in about 6% of cases.[18]

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 to have good prognosis.[19] In case atumour free margin is not possible, adjuvant chemotherapy or radiotherapy should be used to minimize the rate of recurrence as reported in our case.[20] However, studies have shown that recurrence of the disease can still occur despite adjuvantradiotherapy.[21] Chemotherapy has little or no effect in the treatment of DFSP. However, Imatinib may be use in unresectable tumours. Mohr's micrographic surgery is increasingly gaining ground in the treatment of DFSP due to its high oncologic effectiveness and maximal tissue saving effect.[22]

CONCLUSION

In multiple recurrences with previous surgical scars, it may not be possible to achieve a tumour-free margin during excision as reported in our case. In view of this, adjuvant radiotherapy will be of benefit in lowering the rate of recurrence. The 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

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Competing interests: None.

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, did literature review and assisted in proofreading the manuscript for error in language and medical terms. G. Joseph contributed to the preparation and revision of the manuscript.

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