Dermatofibrosarcoma Protuberans of the Scalp and Lid - A Huge Challenge: Case Report

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ABSTRACT

Dermatofibrosarcoma protuberans (DFSP) is a low grade locally invasive soft tissue sarcoma that rarely metastasizes; it mostly affects the trunk and extremities, and less often the head and neck. The non-specificity of early presentation in terms of symptoms and signs makes it difficult to differentiate from other benign soft tissue tumours. The main modality of treatment is total surgical resection with or without adjuvant radiotherapy and oral medication-, imatinib mesylate. We report a case of aggressive DFSP involving the right part of head and right upper eye lid in a 35year old house wife. She subsequently had excisional biopsy and Culter-Beard procedure to construct the right upper eyelid. This study was done to create awareness, challenges and the role of Oculoplasty in the management of this rare condition. Dermatofibrosarcoma protuberans is a rare and difficult tumour to treat. The role of the Oculoplastic Surgeon in managing this tumor is underscored.

Keywords: Dermatofibrosarcoma, eyelid, management, role of oculoplasty

INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) is a low grade soft tissue sarcoma that starts in the dermis, then later involves the surrounding structures.[1] This condition was first recognized by Taylor in 1890 but Hoffman gave the name DFSP in 1925.[1-2] It accounts for less than 0.1% of all malignancies, and 1% of soft tissue sarcomas.[3] The tumour mostly affects the trunk followed by the extremities with less involvement of the head and neck region 13% [2, 4] of which 3.5% of the head and neck involve the periorbital region.[5-6] The DFSP usually occur in young and middle age[7] slightly more in male.[2] However, some studies show no gender differences.[8] The cause of DFSP is not clear but it has been proposed to follow an unknown parasitic infection and trauma.[9] Environmental and genetic predispositions have been suggested as factors in the occurrence of this disease.[10] An association between DFSP and spindle cells lipomas has also been reported.[11] The DFSP is locally invasive tumours but rarely metastasizes.[2-3] However the 5-year risks of recurrence of the tumour are 25%.[12]

The early presentation of the tumour is not specific, therefore making it difficult to differentiate it from other benign soft tissue tumours.

The main treatment of DFSP is total surgical resection with or without adjuvant radiotherapy and oral medication of imatinib mesylate[13] - a tyrosine kinase receptor inhibitor or a new promising vascular endothelial growth factor (VEGF) receptor inhibitor, sorafenib.[14]

We report a case of aggressive DFSP involving the right part of the head and right upper lid, managed at our hospital. This is to create awareness, challenges and the role of Oculoplasty in the management of this rare condition.

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CASE REPORT

A 35 -year old full time house wife, referred to oculoplastic unit from Plastic surgeon unit of our hospital. She came from a remote part of Kebbi State presented with swelling on the right side of the head and face of 18years duration. The swelling was initially small as a tiny nodule and later progressively increased in size and spread to involve the upper right part of the forehead and upper lid. This was occasionally associated with mild pain, bleeding on pressure and inability to open the right eye (RE) properly. No other ocular symptoms and no similar swelling in any other part of the body. There was no preceding history of trauma to the head and face. She is not a known hypertensive or diabetic. She previously had 2 excisional surgeries in the past 10 years at a health facility (general hospital), but this was not followed with histopathological examination of the samples. She had 4 children, 3 alive, and the last child birth was 8 years ago. There is no family history of similar problem.

Examination revealed a middle age woman, with multiple swellings measuring about 8 x 10 x 12cm involving the right parieto-frontal area and extending to the right upper lid. It was firm on palpation and fixed to the skin and underlying structures (Figures 1).

Ocular examination revealed unaided visual acuity of 6/24 and 6/6 in RE and LE respectively. Right eye examination revealed mechanical ptosis, with normal conjunctiva, cornea, pupil, and lens with a glimpse of flat retina. The left eye was normal.

The differential diagnoses were diffuse neurofibroma, dermatofibrosarcoma, fungal infection, nodular fasciitis, keloid, malignant fibrous histocytoma, atypical fibroxanthoma and giant cell fibroblastoma. Blood samples were taken and the results of investigations are as follows: normal FBS, FBC, E/U/Cr and negative retroviral screening. Computer Tomography scan report showed soft tissue involvement without bony erosion. The patient was co-managed with plastic unit of our hospital. She subsequently had incisional biopsy 2 weeks after presentation and histopathology report confirmed DFSP (spindle cells proliferation disposed in storiform pattern). A week after presentation, she had local resection of the tumour involving 3cm of normal margin and Split Thickness Skin Grafting (STSG) from the patient's right thigh was done to cover the parieto-frontal wound. The patient was discharged 26days postoperative, with 99% STSG taken. Histopathology report of the resection mass confirmed DFSP. The patient came back for follow up 6 months post-operatively with unaided VA of 6/18 and 6/6 RE and LE respectively. There was total loss of right upper lid, keratinisation of the right upper conjunctiva, and a superior limbal-cornea scar. Other eyes examinations were normal (RE and LE). Figure 2. She was planned for Cutler Beard procedure of the right eye lid. However, the patient declined the surgery. She came back to our center 14 months post-operative, with small recurrent growth in the scalp and right lower lid closer to the right medial canthus with unaided VA of 6/18 and 6/6 of RE and LE respectively (figure 3). She had emergency right Cutler Beard procedure and excisional biopsy with primary wound closure of the right lower lid growth and graft of the scalp wound (figure 4).

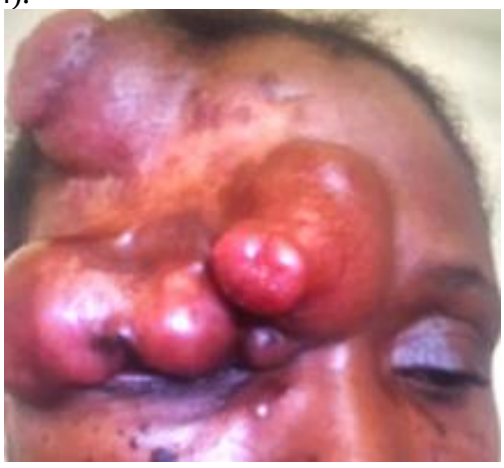

Figure 1: Patient at presentation

Figure 2: A 6months post-operative

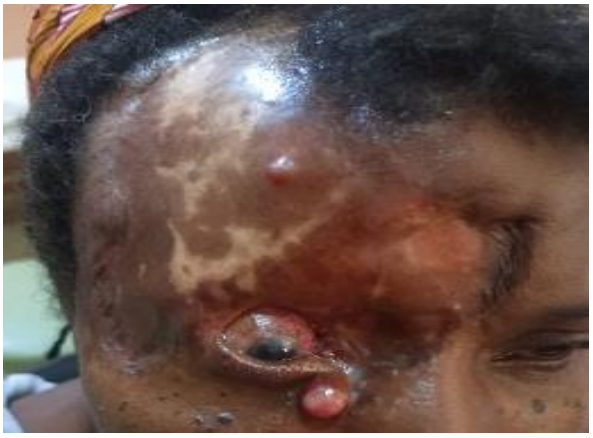

Figure 3: patient at 14 months postoperatively

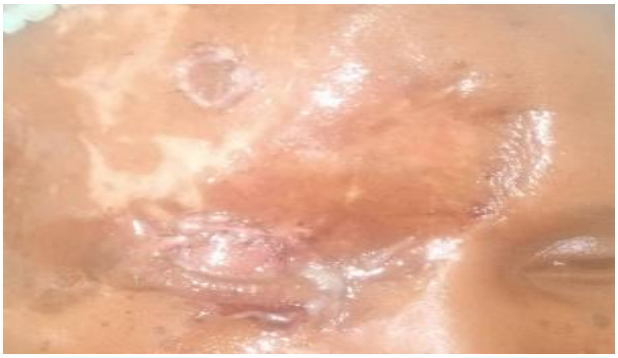

Figure 4: Patient at 1st day post-operative- Cutler- Beard procedure + FTSG (by Oculoplastic surgeon alone).

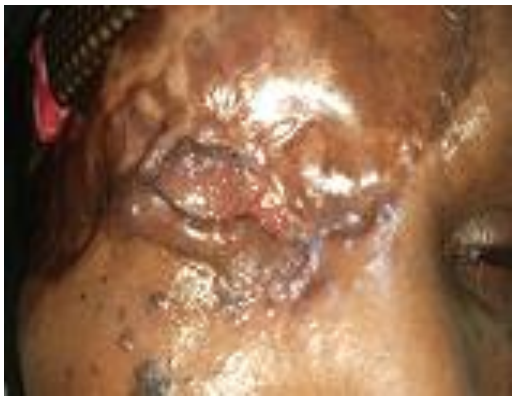

Figure 5: Patient 2/52 post Cutler- Beard procedure

Post-operatively, the patient did well and she was planned for secondary release of the Cutler Beard procedure at 2 months (Figure 5).

The patient is yet to come for second stage of Cutler-Beard procedure and all effort to locate her proved abortive.

DISCUSSION

The age of our patient is 35years is in tandem with previously reported studies[2, 7, 8,9] where tumour was reported to occur in young age. The presentation of the lesion follows the pattern presented in previous documented reports[2, 9, 15] of a tiny skin nodule which later progressed to form a confluence, of an irregularly circumscribed shape. The tumour in our patient was attached to both the skin and underlying tissue which is at variance from a previous study.[2] The difference may be because of late presentation and the 2 previous excisional surgeries might have distorted the natural pattern in our patient. In this case there was occasional mild pain in concordance with a previous study[9] but different from another study[2] that reported no pain. The pain in our patient may be the outcome of inflammatory reactions or effects from the 2 previous excisional procedures. There is no weight loss in our patient, and this is in agreement with previous documented reports on DFSP.[2, 9] Histology report of our patient confirmed the diagnosis of DFSP in agreement with previous documentation.[2-5, 9] It is worthy of

note that, because of non-availability of immunohistochemical staining (CD34 and Vimentin) in our centre, it is difficult to make early differentiation from other tumours such as giant cell fibroblastoma, myxomia liposarcoma, and fibrosarcomatous dermatofibrosarcoma .

Using American Musculoskeletal Tumour Society (MSTS) staging system[2] and European consensus–based interdisciplinary guidelines[15], our patient lesion was stage IB and I respectively because the lesion was attached to the underlying tissue, with no lymph node involvement and distant metastasis. We offered our patient a wide surgical excision extending to 3cm normal margin as recommended in literatures.[2, 9, 13]It's generally reported that the DFSP of head and neck usually have high rate of local recurrence of up to 56%. [13] The period of recurrence after our excision surgery was 14 months and this was far earlier than found in previous studies.[16-18] The difference might be because our patient has no opportunity and access to adjuvant therapies like radiotherapy and chemotherapy and also the innate propensity to have recurrence as she has had 2 previous excisional surgeries. The extensive involvement of the upper right eye lid with the tumour resulted in sacrificing the right upper lid, which subsequently necessitated the Cutler-Beard procedure. To our knowledge, this is the first case of DFSP involving the upper eye lid in which Cutler- Beard procedure was used in Nigeria.

Challenges in managing this patient included late presentation (a common problem in African patients including Nigeria), intraoperative bleeding (which necessitated postoperative transfusion), non-availability of immunohistochemistry (CD34 and vimentin), improper keeping of follow-up appointment (a common problem among Nigerian patients), and treatment challenges such as non-availability of radiotherapy and adjuvant drugs (imatinib mesylate is a tyrosine kinase inhibitor, which might help in preventing local recurrence in this patient) in our centre and financial constraints. Financial constraint was responsible for the inability of the patient to be regular for follow-up clinic appointments and the utilization of radiotherapy (patient is from a remote rural part of the state and of a lower socioeconomic status).

CONCLUSION

Dermatofibrosarcoma protuberans is a rare and difficult tumour to treat. The role of the Oculoplastic Surgeon is underscored. Poor follow- up appointments, financial constraint and non-availability of adjuvants (chemotherapy), and immunohistochemistry (CD34 and Vimentin) still remain a set back in managing this condition in this part of the world. Hence, it is advocated that necessary facilities for investigation and treatment of this type of cases, should be provided by the relevant agencies, as part of the cancer-free policy of the Government.

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