Hemorrhaging And Large Dematofibrosarcoma Protuberans With Tumor Recurrence Of The Left Shoulder In An Adult Male: A Rare Complication

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Citation

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Abstract

Dematofibrosarcoma protruberans (DFSP) is a rare soft-tissue tumor. Massive hemorrhage is not one of its common features. It occurs slightly more often in males than in females and trauma to soft tissue has been implicated in its etiology. Mohs micrographic surgery (MMS) is the treatment of choice for DFSP. Our report reviews a case of DFSP with a rare complication at presentation and its management as well as published literature.

A 55-year-old business man and farmer presented with a large mass at the right shoulder with ulceration over the surface, which was bleeding profusely. He was pale and weak, morbidly obese with a BMI of 37.37kg/m2, and his pulse rate was 124bpm. Blood pressure was 130/80mmHg, respiratory rate was 30cpm and the chest was clinically clear. He was resuscitated with two units of blood before a wide excision was carried out. The histopathology report showed features consistent with DFSP. The results revealed a mesenchymal neoplastic tumor with homogenous spindle cells, arranged in radial whorls producing a storiform or Cartwheel pattern. They were infiltrating in-between the adnexa with extension into the subcutis, trapping fat. Mitotic figures and mild atypia was observed. Before discharge, the patient was noticed to have two newly-growing buds of the tumor on the right shoulder. He had had excisions in the past for tumors in the same shoulder.

Clinicians need to be aware of re-occurrences of DFSP. Furthermore, DFSP may present very tumorous, large, ulcerated and bleeding profusely, leading to severe anemia, which is not a common clinical presentation, as seen in the reported cases.

INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) was originally described in 1924 by Darier and Ferrand.1,2 It accounts for less than five percent of soft-tissue tumors and 0.1 percent of all malignancies with an annual incidence of 0.8 to 4.5 per million.3,4 It is a rare, slow-growing, fibrohistiocytic neoplasm commonly seen among those in their third or fourth decade of life and it is commoner in the trunk (40-60%), followed by the proximal extremities (20-30%) and the head and neck (10%-16%).4,5 DFSP frequently recurs locally after incomplete excision. The general immuno-staining pattern of DFSP is CD34-positive and factor-XIIIa-negative6. Mohs micrographic surgery (MMS) is the treatment of choice for DFSP. DFSP is one of the malignant variants of fibrous tissue tumors; others are malignant fibrous histiocytoma (MFH), low-grade fibromyxoid sarcoma, fibrosarcoma, desmoid fibromatosis, and nodular fasciitis. We report the rare presentation of a hemorrhaging large DFSP in a black African man.

CASE REPORT

A 55-year-old businessman and farmer presented this year with a mass of the right shoulder of 5 years duration. It progressively increased in size, was painless, and firm to hard in consistency. Four month before presentation, it became associated with pain as the surface began to ulcerate, and has been bleeding spontaneously and profusely till he was weak. Bleeding usually stopped after using hydrogen peroxide and cottonwood, and dressing with gauze and bandage by a chemist, for about 3 to 5 days before a new episode of bleeding. Prior to the bleeding, there was neither history of trauma, nor use of traditional scarifications, nor an attempt at excision by health personnel. He had had excision for recurrent tumor growth in the right shoulder thrice; the first excision was 22 years ago, the second and third were 17 and 3 years before the current excision at other health centres. He also had right inguinal herniorrhaphy 21 years ago.

Figure A

Superior view of the right shoulder; DFSP with distended vessels; Old incisional scars on the right shoulder

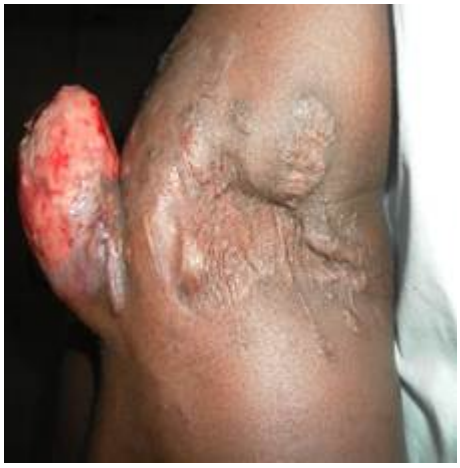


Figure B

Ulcerated DFSP

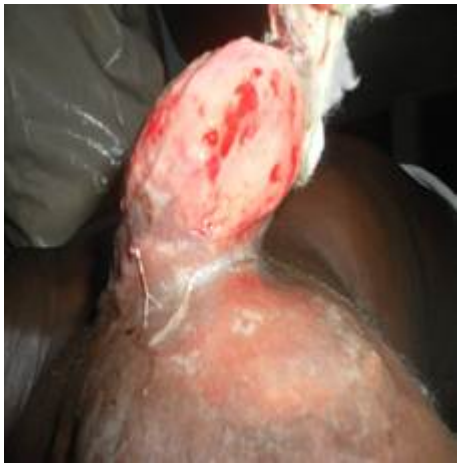
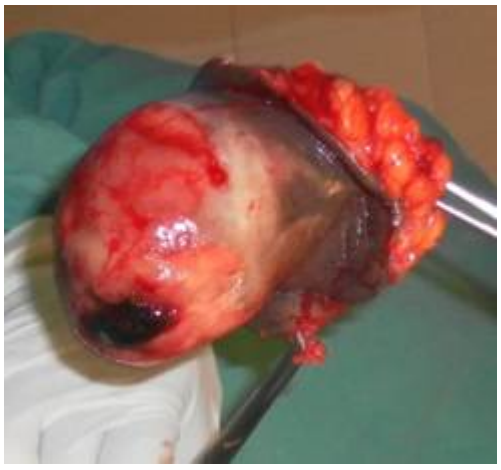


Figure C

Excised Tumor



He was not a known hypertensive or diabetic patient, not asthmatic, and there was no history of epistaxis. He does not take alcohol nor tobacco products.

On examination, he was severely pale, with a right shoulder mass oozing profusely, acyanotic, anicteric, with bilateral pedal pitting edema, and morbidly obese with a BMI of 37.37kg/m2. The pulse rate was 124bpm. Blood pressure was 130/80mmHg, respiratory rate was 30cpm, the chest was clinically clear and no abnormality was noticed per abdomen. The size of the shoulder mass was of an average fist. The surface was ulcerated with distended vessels (figures A and B), and dressed with soiled bandage to prevent bleeding. His pre-transfusion and preoperative packed cell volume was 18% and his hemoglobin level was 5.9g/dl. There was leucocytosis with a differential count of 40% and 48% for neutrophils and lymphocytes, respectively. His urinalysis, serum electrolyte, urea and creatinine levels were normal. He was stabilized with two units of screened blood under frusemide cover.

Figure D

Excision wound

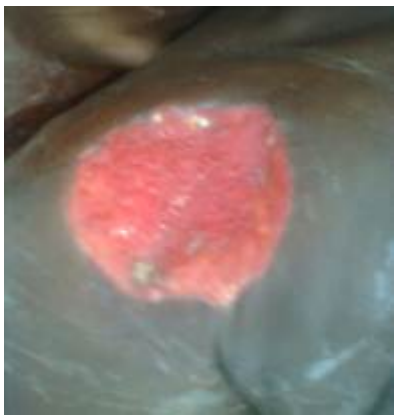


Figure E

Early un-excised tumor bud

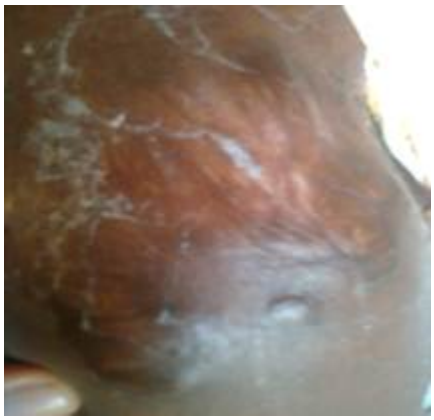
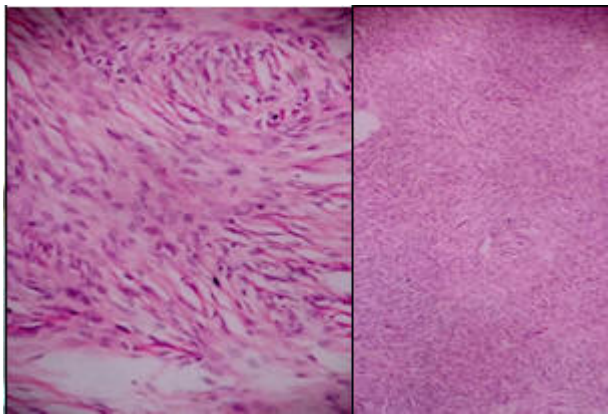


Figure F

Histocytologic patterns (x10 & x4, Hematoxylin & Eosin stain



He had a wide excision of the mass (Figure C) through the intra-fascial plane of the deltoid muscle, under general anesthesia with propofol and midazolam intravenously for induction and halothane for maintenance via laryngeal mask. Hemostasis was secured with artery forceps temporarily followed by stakes of chromic catgut sutures permanently. The surgical site was dressed with Sofra-Tulle® and covered. His post-operative vital signs were the following: a temperature of 35.5°C, a pulse rate of 95 beats per minute, a respiratory rate of 24 cpm, a blood pressure of 109/82 mmHg, and an SpO2 of 94%. First-day post-operative PCV was 25%. A few days thereafter, the wound was skin-grafted. The patient was managed with analgesics and antibiotics. After 21 days post-operatively, the donor and recipient sites were covered with gentian violet before discharge in stable clinical condition, to be followed-up in the clinic. During the post-operative care, the patient was noticed to have other sites of the lesion in old incision scars of the shoulder, one was 6cm proximal to the superior edge of the wound, mobile, firm, circumscribed and 3x3cm, while another lay at the contour of the shoulder, also of small size. The patient was informed. The histological report of the biopsy described an ulcerated penduculated mass, partly covered by negroid skin, partially embedded in 2 blocks. Microscopically, sections showed atrophic and ulcerated skin tissue containing a mesenchymal neoplastic tumor. The cells were homogenous spindle cells, arranged in radial whorls producing a storiform or Cartwheel pattern (Figure F). They were seen to infiltrate between the adnexa with extension into the subcutis, trapping fat. Mitotic figures were observed and atypia was mild (Figure F). On a follow-up visit, the patient's packed cell volume had become 31%.

DISCUSSION

Dermatofibrosarcoma protuberans (DFSP) is a relatively uncommon soft-tissue neoplasm with intermediate to low-grade malignancy. DFSP is a locally aggressive tumor with a high recurrence rate. The term came into existence in 19257. Dermatofibrosarcoma protuberans (DFSP) is a cutaneous malignancy that arises from the dermis and invades deeper subcutaneous tissue (e.g., fat, fascia, muscle, bone). This finding was similar to the histological report where some malignant cell was noticed to invade subcutis trapping fat. In our reported case, past history was supportive to the assertion made by the British Association of Dermatologists that injury to the skin is a predisposing factor8. Its pathogenesis has been clearly demonstrated in the following report that chromosomal aberrations such as reciprocal translocations of chromosomes 17 and 22, t(17;22), leads to expression of platelet-derived growth factor B, which binds to the PDGF receptor leading to intracellular reaction that promotes proliferation of the tumor (DFSP) 9, 10, 11, 12,13,14,15. Further investigations may be carried out which include immuno-staining for CD34 and factor XIIIa. Other investigation that may be required include chest X-ray to rule out lung metastasis16, computed topography scanning to exclude bone metastasis, MRI to delineate tumor depth and border16,17,18,19, ultrasonography to reveal lymph node involvement and fluorodeoxyglucose (FDG) positron emission tomography for monitoring metastasis20. The reported case had a history of recurrence which was coherent with the nature of DFSP. Chih-Shan and Dirk21 stated that most recurrences occur within 3 years of the primary excision. Patients should be seen every 6 months during this period and annually thereafter.16

A literature review of DFSP case series treated with Mohs surgery showed that 50% of recurrences appear within the first 3 years after operation and 25% of local recurrences are detected after 5 years. A large case review from a series of 159 patients treated at Memorial Sloan-Kettering Cancer Center (New York) showed that the medium time to the development of a local recurrence was 32 months. The indolent nature of DFSP requires lifelong surveillance for recurrence.22 Dermatofibrosarcoma protuberans (DFSP) is characterized by its aggressive local invasion; this is expressed by extending tentacle-like projections underneath healthy skin, rendering complete removal of the tumor very difficult. Incomplete removal of these neoplastic cells results in a high local recurrence rate. This could be a strong clue to repeated excisions which our patient has had; furthermore, during one of the reviews in the post-operative periods small lesions were noticed just few centimeters away from the operation site. Despite local invasiveness and recurrence, DFSP rarely metastasizes21. The German guideline stages16 the disease from 1 to 111. Clinical stage 1 stands for the tumor itself while 11 and 111 represent regional lymph node involvement and distant metastasis, respectively. The lungs are the most common site of distant metastasis that occurs via hematogenous spread21. Usually, metastatic disease is preceded by multiple local recurrences.16 The type of the surgical procedure has been reported to impact on the risk of recurrence. Mohs technique has been known to be better than wide excision. However, our patient had the latter. Better prognostic factors are low number of mitotic figures, reduced cellularity, DNA euploidy, TP53 gene de-expression, the absence of fibrosarcomatous changes within the tumor and age less than 50 years. Our patient was 55 years old and had mild atypia with mitotic figures. Dermatofibrosarcoma protuberans (DFSP) usually occurs in adults aged 20-50 years. Rarely, DFSP has been reported in newborns and elderly individuals (80 years)23. Fibrosarcomatous progression, a DFSP variant, is more aggressive in nature, and the clinical outcome usually is poor24. The loss of the t(17,22) cytogenetic marker in the fibrosarcomatous progression variant of DFSP may represent progression of malignancy7, 20,25. The Bednar tumor, a variant of DFSP, has been shown to occur 7.5 times higher in blacks than in white patients26. Our reported patient was a black native African. Moreover, several studies26,27,28 of dermatofibrosarcoma protuberans (DFSP) reveal an almost equal sexual distribution or a slight male predominance. Finally, profuse bleeding as seen in our patient may be explained by high vascularity that was associated with the tumor which could have ulcerated and bled. The tumor has been noted to be associated with telangiectasia21. Therefore, bleeding episodes can be managed by applying bandage, blood transfusion and excision of hemorrhaging DFSP from the site of occurrence as in our patient.

CONCLUSION

Conclusively, DFSP can be highly vascularised if it grows to a relatively large size. It may ulcerate and bleed profusely to such extent to require blood transfusion. Management of DFSP includes proper staging, prognostic evaluation, explanation of treatment options, and planning. All these depend on thorough history taking and physical examination. Imaging studies may facilitate the assessment of local invasion and distal metastasis. Multidisciplinary collaboration21 between a dermatologist, surgical oncologist, plastic surgeon, medical oncologist, radiation oncologist, and pathologist is necessary in locally advanced, recurrent, or metastatic cases of DFSP.

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