Original Article

Dermatofibrosarcoma Protruberans A Locally Aggressive And Recurent Malignant Tumour Of The Skin: Review And Case Report

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ABSTRACT

Dermatofibrosarcoma protuberans (DFSP) is uncommon but represents one of the most common dermal sarcomas, which is said to be a locally aggressive, low grade, and relatively uncommon cutaneous tumour, which has a high propensity for local relapse with low metastatic potential.

Over 90% DFSPs are genetically characterized by chromosomal rearrangements (translocation t (17;22) (q22; q13), resulting in the collagen type-1 alpha 1 (COL1A1)-platelet-derived growth factor β (PDGFB) fusion gene with well-known risk factors for developing DFSP, some cases develop at the site of previous trauma and reports have included a burn scar and the site of vaccination.

The mainstay mode of treatment of DFSPs is wide local excision with traditionally 3-cm gross margins excision surgery. Mohs Micrographic Surgery (MMS) is highly recommended surgery for DFSP and is very useful treatment choice for recurrent DFSP.

This case report discusses two patients with confirmed DFSP and have received adequate therapy. It also reviews salient features of evaluation and prompt management of DFSP and much emphasis on long-term follow-up of patients.

Key Messages:

Early diagnosis of pediatric DFSP is critical to minimize surgical disfiguration.

We emphasize early evaluation of any suspicious skin lesion should not be ignored and should be excised and sent for histopathology.

Undertreating a DFSP is even more problematic, as the tumour is locally destructive, can metastasize, and can prove fatal without treatment.

A full metastatic workup should be done and patient should be regularly followed up, if it turns out to be malignant.

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INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) is a locally invasive tumour arising in the dermis and showing fibroblastic differentiation.[1] It accounts for only 1% of all soft tissue sarcomas and <0.1% of all malignancies.[2] It is said to be a locally aggressive, low grade, and relatively uncommon cutaneous tumour which has a high propensity for local relapse with low metastatic potential.[3,4,5]

Dermatofibrosarcoma protuberans is uncommon but represents one of the most common dermal sarcomas. The incidence in the USA has been estimated as 4.1 cases per million. According to Kreicher KL et al, the trunk was the most common anatomic site except in older men. Incidence among women was 1.14 times higher than in men (95% confidence interval [CI] of rate ratio: 1.07-1.22). Incidence among blacks was almost 2 times the rate among whites (95% CI of rate ratio: 1.8-2.1).[6]

DFSP tumours more commonly develop during the third and fifth decades of life. It usually occurs in young adults (aged between 20 and 40 years), and there is no definite predominance considering the sex of the patient.

However, presentation during childhood and late life is not particularly rare. Congenital cases have been described as evidenced by Han HH et al.[7]

Key words: dermatofibroma, dermatofibrosarcoma protruberans, Lipoma, fibrosarcomatous, recurrence, aggressive cutaneous tumour

There are some well-known risk factors for developing DFSP, some cases develop at the site of previous trauma and reports have included a burn scar and the site of vaccination. Exceptional cases have been associated with previous radiotherapy to the area. There is an association between DFSP and children with adenosine deaminase deficient severe combined immunodeficiency. Patients affected by the latter have a higher incidence of tumours presenting at early age and often multicentric.[8]

The tumour is more often commonly affects the trunk in about 40%-50% of cases, the proximal extremities (30%-40%), and the head and neck represents 10%-15% of cases[9,10]. Involvement of the limbs is usually proximal. Presentation on the hands and feet, particularly on the digits, is very rare. It may begin in early adult life with one or more small, firm, painless, flesh-colored or red dermal nodules.

Clinically DFSP tumours start as a plaque, which may occasionally be atrophic in nature. Their progression is usually very slow, and may occur over many years; a significant proportion of these tumours only become protuberant after a long period. Jonathan N at al classified DFSP into three different forms: 1. a morphea-like form mimicking a scar, morphea, morphea-form basal cell carcinoma, 2. a dermatofibroma plaque; an atrophoderma-like form similar to atrophoderma or anetoderma; and finally, 3. an angioma-like form resembling vascular malformations.[11] Eventually, one or multiple nodules may appear in the protuberant phase. Eventually, nodules develop, coalesce and extend, becoming redder or bluish as they enlarge to form irregular protuberant swellings. At this stage, the base of the lesion is a hard-indurated plaque of irregular outline. In the later stages, a proportion of lesions become painful and there may be rapid growth, ulceration and discharge.[12]

In this case report we discuss two cases of seemingly innocent painless occipital-parietal tumour in a 24-year-old African man and left iliac tumour in a 42-year-old Chinese woman. We hope to review, discuss the evaluation, management and follow up of DSFP patients especially in resource depleted environments and to emphasis that most innocent lumps (tumours) should be evaluated and managed appropriately and promptly.

Case Presentations

CASE 1

A 24years old African man; presents to the outpatient department of Livingstone Central Hospital with a history of a painless lump on the back of head with an increased growth rate for the past 3months.He had become aware of his lump for the past 11months, informed by his bar-barman. However, he gives a positive history of having a scar at the exact position for the past 13years.

He had no any other complaints related to the lump apart from psycho-social-cosmetic awareness of the lump on head. He is worried that despite it being painless, it is growing fast in past 3months.

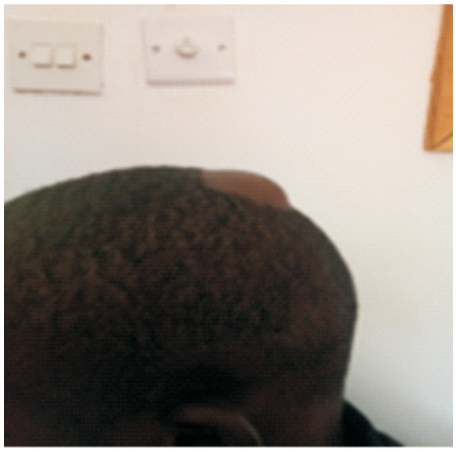
He has no relevant medical and surgical history. He is HIV/AIDS negative. He has no known food or drug allergies. He does not consume alcohol nor smoke cigarettes.

On physical exam, he is a well-built man with normal vitals. Oriented in time, place and person. The other exam was essentially non-reveling.

Local exam: He has a dome shaped mass on the left side of the parietal-occipital region with normal overlaying skin and hair stumps. An old scar on the left aspect of the lump about a centimeter. The mass is soft to firm soft in consistency, non-tender and mobile. It was free from the base of the skull and there was no skin tethering on traction of the lump. It was approximately 5 to 6 centimeters.

Figure 1: Arial view of the dome shaped tumour with hypopigmented scar on the left posterior lateral.

Figure 2: lateral view of the protuberant tumour.



Baseline Biochemistry and Haemogram results were normal. No radiological exam was done.

A clinical diagnosis of Lipoma was entertained with differentials of dermatofibroma, Liposarcoma, dermatofibrosarcoma protuberans.

An excision biopsy was done under local anesthesia using 2% lignocaine mixed with 1mil of adrenaline. The Lump was about 4*2 centimeters. Patient was covered on adequate analgesia and antibiotics (Para-codeine and cloxacillin)

Results and Management

Histopathology report: reviewed a grayish piece of tissue measuring 43*25*12mm with a whitish cut surface macroscopically. Microscopic examination showed a non-circumscribed, highly cellular, tight storiform pattern that infiltrates deeply into the subcutaneous tissue and entraps fat cells to form characteristic honeycomb pattern. A fascicular growth pattern was also seen the cells are monomorphic, thin and spindly with scant eosinophilic cytoplasm and hyperchromatic nuclei.

Immunohistochemical report: sections showed fibrofatty connective tissue containing a tumor with a spindled morphology. The tumor shows vague storiform pattern of growth with round to oval plump nuclei showing inconspicuous nucleoli. There is brisk mitotic activity with average 6 per 10 High Power fields. Infiltration of the fatty tissue is noted producing a "honey-comb" effect. No fibrosarcomatous transformation is observed.

In the presence of appropriate and adequate controls, the tumor cells show the following immunophenotype: **Hematopoietic progenitor cell antigen CD34**: diffusely positive while S-100, AE1/AE3, EMA were all negative.

Conclusion: Soft tissue mass with spindle cell tumor with histopathological and Immunohistochemical features compatible with dermatofibrosarcoma protruberans (DFSP) with no fibrosarcomatous transformation.

Definitive diagnosis: Dermatofibrosarcoma Protruberans in a 24-year-old man on Parietal-occipital region of the scalp with no obvious local infiltrates.

A Multidisplinary approach in the management of patient was used. The lump was completely resected in a sterile procedure room under local anesthesia. After the histopathological and Immunohistochemical reports the patient was referred to the oncologists (Cancer Disease Hospital) for evaluation and patient indexing. They advised for possible radiotherapy if recurrence of tumor occurs. The patient was reassured and will be followed up at 3,6,18,36 and 72months respectively via the dermatologist (primary clinician).

CASE 2

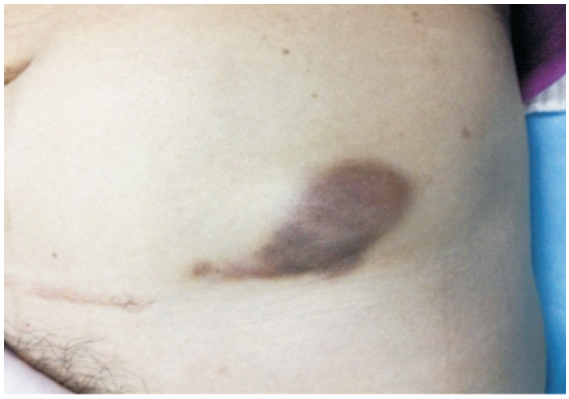
A 42-year-old Chinese woman presents to the dermatology outpatient wing of Zhongnan hospital of Wuhan University with a 20year history of a progressive growing left abdominal mass. The mass was noticed initially as a clearly well-defined plaque. It started as a painless growing hypertrophic scar but in the past one year the mass had reached an enormous size of protrusion. It was associated with change in overlying skin colour, itchiness and discomfort. There was no disturbance of the bowel habitus. Patient had no any other systemic symptoms.

She is HIV/AIDS, Hepatitis B Virus seronegative. Her past medical history is unremarkable. She has a history of caesarean section. She also has no known food or drug allergies. She does not indulge in alcohol nor cigarette smoking.

She was oriented in in time, place and persons. All her vitals were normal. She had no hepatosplenomegaly or any abdominal mass.

On local exam, she had a brownish tan lump on the left lower parts of the iliac region with features of mild excoriation (suggestive of pruritus). the mass was about 15*5centimeters, non-tender firm to hard in consistency and fairly mobile.

Figure 3: Hyperpigmented protuberant tumor on the left iliac region, adjacent is an old caesarean section scar.



Histopathology report reviewed classic features of DFSP with fibrosarcomatous change. Immunohistochemical tests showed CD34-positive, K1-67 (positive rate of about 10%). For Desmin, S-100, SMA, SOX-10, STAT6 and MB45 were negative.

Figure 4: DFSP specimen with typical histopathological features at low power showing spindle cells in storiform arrangement. The cells stain positive for CD34.

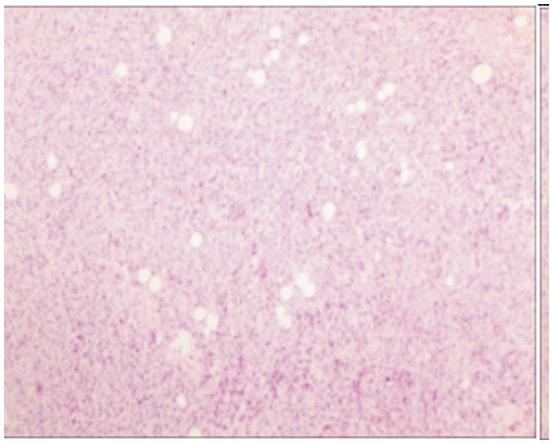
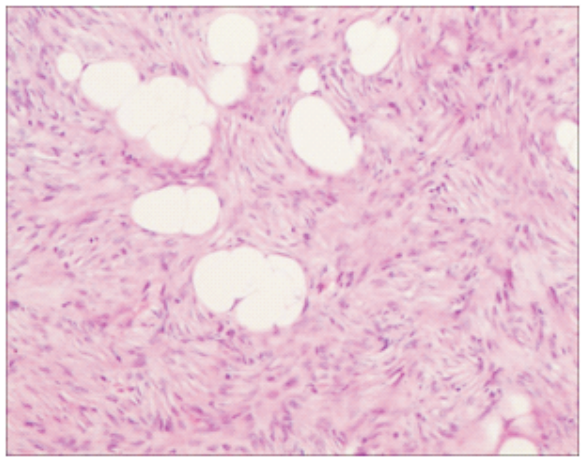


Figure 5: DFSP specimen with typical histopathological features at High power showing spindle cells in storiform arrangement and fibrosarcomatous change.



She is being managed as a case of Dermatofibrosarcoma Protuberans with fibrosarcomatous change.

The tumour was excised completely taking care of the 3cm clear zone margin from the lump. She was covered on adequate analgesia and prophylaxis antibiotics. Care of the wound was done. Patient will have serial follow-ups to 5years.

DISCUSSION

The pathogenesis of DFSP still remains elusive, however, over 90% DFSPs are genetically characterized by chromosomal rearrangements (translocation t(17;22) (q22;q13), resulting in the collagen type-1 alpha 1 (COL1A1)-platelet-derived growth factor β (PDGFB) fusion gene[13,14]. DFSP commonly occurs in patients in who are in the age groups between 20 and 40 as was with the case with our two patients from different ethnic groups and social backgrounds. However, both congenital and pediatric cases of DFSP have been reported.[7, 13, 15, 16, 17]

Clinical presentation of DFSPs varies from patient to patient from simple deceptively innocuous indolent plaque to a protuberant tumor with or without multiple nodules. Wrotnowski et al reports that one or multiple nodules may appear in the protuberant phase of DFSPs. In late clinical presentation, multiple nodules develop, coalesce

and extend, becoming redder or bluish as they enlarge to form irregular protuberant swellings (most appreciated in the light skin patients, type 1-3/4). In later stages, the base of the lesion, is a hard-indurated plaque of irregular outline with a proportion of lesions become painful and there may be rapid growth, ulceration and discharge. Complications such as bleeding, ulceration, and pain may arise at this stage. The tumour often invades deep structures such as the fascia, muscle, or bone. Most patients tend to present at this stage, the rapid growth stage. Thus, period of presentation is crucial in the management of DFSPs. The decision to biopsy will be driven by the evolution of the tumour and its location. Even more challenging is reconciling clinical presentation with a potentially ambiguous histologic picture to determine the best treatment. Thus, it is always important to follow a Multidisplinary approach in the management of these patients.

The mainstay mode of treatment of DFSPs is wide local excision with traditionally 3-cm gross margins excision surgery.[4, 18-20] Some literature advises 2-4cm gross margin. Post-excision reconstruction included direct closure, skin grafting, and local or free flap reconstruction surgery. Thus, the size and location of the tumour plays a major role in deciding what mode of post-excision reconstruction. Mohs Micrographic Surgery (MMS) which is practiced in highly advanced centers has showed much better results.[21] Fontecilla et al denotes that wide local excision for DFSP yields a recurrence rate of 30.8% compared with 3% in patients who underwent Mohs micrographic surgery. Li Y et al had recorded zero recurrence in his cohort study of 57 patients who underwent Mohs micrographic surgery and suggests that MMS as a very useful treatment choice for recurrent DFSP.[22] It is also recommended for areas where reconstruction may pose challenges like the head and neck.[10, 23-25]

Our two patients underwent wide local excision biopsy with respect to the 3cm margin thus, they will be followed up serially up to 5years.In case of recurrence, depending on other clinical factors, either re-excision surgery or radiotherapy would be offered. Hamid et al observed that radiotherapy is effective, and it decreases the recurrence rate in the treatment of DFSP. It is especially helpful in margin-positive disease.[26] Preferably the excision should be followed by adjuvant radiotherapy, when margins are found close or persistently positive and repeat wider resection is not feasible due to anatomic limitations.[27, 28] The use of adjuvant radiotherapy after wide resection in the fibrosarcomatous subtype is highly recommended, even with negative margins.[29]

Neoadjuvant therapy with imatinib mesylate aims to reduce the size or extent of the tumour before using radical treatment intervention, surgery. This drug is given for at least 6 weeks prior excision of the tumour.[24, 30, 31] It can also be use post excision especially if tumour is located in areas where adjuvant radiotherapy may be impossible to administer.[32]

The definitive diagnosis of DFSP is made via careful evaluation of the patient: from history, histopathology and Immunohistochemical analysis of the sample. Today, most in situ hybridization procedures use fluorescent probes to detect **DNA** sequences, and the process is commonly referred to as **FISH** (fluorescence in situ hybridization).[33] This could be used to characterized by chromosomal rearrangements (translocation t (17; 22) (q22; q13), resulting in the COL1A1-PDGFB fusion gene as it has been found to be positive in over 90% of DFSPs.

In both our patients, only histopathology and Immunohistochemical analysis where of note. The classical picture on histopathology, shows spindle-shaped neoplastic cells with elongated nuclei and scanty pale cytoplasm primarily in the dermis arranged in storiform, cartwheel or whorled pattern with irregular infiltration of the fibrous septae of subcutaneous fat forming pseudoseptate or lace-like pattern, also known as honeycomb appearance[8, 34, 35] as evidenced in figure 5 and 6.

Other histological variants of DFSP include Bednar tumours (pigmented DFSP)[36], myxoid tumours[37], giant tumour), neuroid, fibrosarcomatous[38], myoid, and granular cell types depending upon the tissue admixed in DFSP. Our Chinese patient had the fibrosarcomatous type.

The immunostaining pattern of DFSP is Hematopoietic progenitor cell antigen CD34 positive and factor XIIIa negative, which helps in differentiating DFSP from other conditions. Dermatofibromas are CD34 negative and factor XIIIa positive.[33]

Immunohistochemical tests showed positive CD34- in both patients and K1-67 was at a positive rate of about 10% for the 42year old Chinese woman which indicates a much higher proliferation index in DFSP.[18] The other Immunohistochemical tests done for her were Desmin, S-100, SMA, SOX-10, STAT6 and MB45, which were negative. For the 24-year African man the S-100, AE1/AE3, and epithelial membrane antigen (EMA) proteins where negative. However, proteins vimentin, coagulation factor XIIIa and FISH analysis for PDGFB rearrangement were not done on both patients (different centers). Studies have shown that CD-34 and vimentin are usually expressed in many cases of DFSP. Coagulation factor XIIIa is generally not expressed in DFSP, but is strongly expressed in dermatofibroma.[39] Must emphasis that benign dermatofibromas will stain strongly positive for factor XIIIa but negative for the marker CD-34.

Other laboratory and radiological investigation were unremarkable for both patients. Most of the patients with DFSPs do not have any other co-morbid conditions and the evaluation of the system is usually normal. These are used for monitoring purposes of the patient in case of chemotherapy or radiotherapy side effects.

CONCLUSION

Despite DFSP not being known to have distant metastatic lesions, its deceptively innocuous nature has lead to most patients presenting to the clinicians during its protuberant stage. At this stage, DFSP is even more problematic, locally destructive thus posing a difficult challenge in its management, as undertreating it is associated with high rates of recurrence. This condition has both cosmetic and social-economic problems, as patient may need adjuvant therapy in difficult cases of DFSP to resect. Reconstructive options, such as skin grafting, and modalities, such as adjuvant postoperative radiotherapy, are suggested to best complement each other. The former minimizes disfigurement while the latter minimizes recurrences. We highly recommend early evaluation of any deceptively innocuous tumour and prompt management to be offered. In addition, all tumours to be sent for histopathology and much emphasis on long term follow-up of patients.

Conflict of interest: to the best of my knowledge, there is no conflict of interest.

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