Dermatofibrosarcoma Protuberans of the Hand*

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ABSTRACT

Dermatofibrosarcoma protuberans (DFSP) is a rare, locally aggressive dermal and subcutaneous mesenchymal neoplasm that often presents as a slow-growing lesion on the trunk, proximal extremities, head and neck. We report a rare case of DFSP arising on the dorsum of the left hand. A 35-year-old male, who presented with a solitary verrucous nodule on the dorsum of the left hand and had experienced two recurrences of the lesion following local excision and electrocautery. Histopathologic examination was consistent with DFSP. CD34 immunostain was positive. Wide surgical excision with a three centimeter margin and direct skin closure was done. Although metastasis is rare, DFSP is associated with marked tendency towards local recurrence after surgery hence patient is for follow-up every six to twelve months post operatively.

Keywords: dermatofibrosarcoma protuberans, CD34 immunostain, wide surgical excision

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INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) is a locally aggressive sarcoma arising from the skin. It is relatively rare and accounts for approximately 4% of all soft tissue sarcomas. [1] It typically arises on the chest, back, head and neck and proximal parts of the extremities. DFSP arising on the hand is rarely reported in literature.[2] Worldwide, there are only 8 reported cases of DFSP of the hand from 1977 to the present.[2,8-14] Locally, there have been no published reports of DFSP of the hand. We report a case of a 35-year-old male with recurrent DFSP occurring on the hand treated with wide excision.

CASE

A 35-year-old male policeman from Tondo, Manila, Philippines presented with a nodule on the dorsum of the left hand, in between the proximal portion of the 4th and 5th digit.

Approximately fifteen years prior to consultation, he noted a slightly pruritic erythematous papule on the dorsum of the left hand, in between the proximal portion of the 4th and 5th digit. The lesion was noted to gradually increase in size from pinpoint to about 1 x 1 cm. No consultation was done and no medications were applied or taken.

Ten years prior to consultation, the lesion then measured approximately 1.5 x 1.5 cm with no accompanying symptoms. Excision was done during a medical mission. Post-operative course was unremarkable. Five months later, he noted recurrence of the lesion on the previous site. The lesion gradually increased in size evolving into a nodule. There was also noted pain with trauma.

Six years prior to consultation, still with increasing size of the lesion, he consulted with a surgeon at a private hospital where electrocautery of the lesion was done. Post-operative course was unremarkable.

5 years prior to consultation, recurrence of the lesion was noted at the same area. Lesion was brownish with a verrucous surface. There was gradual increase in the size of the lesion to more than 1.5 cm in diameter. No pain was noted however, there was minimal bleeding with trauma. No change in color as well as ulceration observed.

4 months prior to consultation, there was noted rapid growth of the lesion to about 3 x 2.5 cm. There was no fever, no weight loss at that time.

Due to persistence of the lesion, patient sought consult at our institution.

The systems review was unremarkable. The patient was non-hypertensive and non-diabetic. He was a non-smoker and an occasional alcoholic beverage drinker. Family history revealed no similar illness and no known malignancies.

The patient came in conscious, coherent , not in cardiorespiratory distress with the following vital signs: blood pressure of 130/80, heart rate of 88 beats/min, respiratory rate of 16 cycles/min, and a temperature of 36.5 ^{0}C. No axillary lymphadenopathy noted. On dermatological examination, he had a solitary, firm, nontender, brownish, verrucous nodule with an eroded ulcerated surface measuring approximately 3 x 2.5 cm in size **(Figure 1)** .

An incision biopsy was done on the nodule of the left hand. The epidermis showed acanthosis. **(Figure 2)**. The dermis showed closely spaced spindle cells with pleomorphic nuclei and few mitotic figures in a storiform pattern extending to the subcutis **(Figure 3)**. CD34 immunostain done was positive **(Figure 4)**. With clinicopathologic correlation, the case was signed out as dermatofibrosarcoma protuberans.

The patient was referred to orthopedic surgery for management of the lesion. They planned to do wide excision of the lesion with possible grafting. However, he opted to have surgery done with a general surgeon at a Chinese General Hospital and Medical Center. Wide excision with a 3 cm margin and direct skin closure was done **(Figure 5)**. Histopathology of the lesion confirmed the diagnosis of dermatofibrosarcoma protuberans. No adjuvant treatment was given. Patient was advised regular self -examination and follow up every six months to check for possible recurrence and metastasis of the lesion.

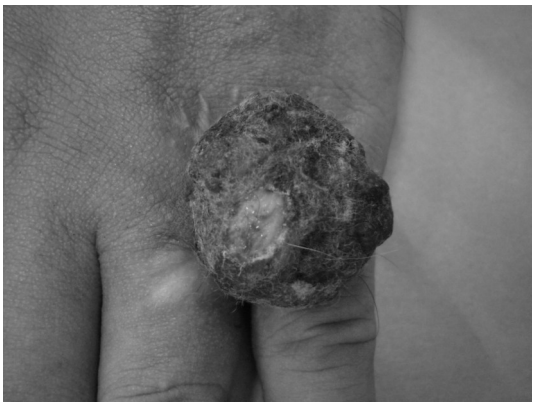

Figure 1. *Solitary, firm, nontender, brownish, verrucous nodule with an eroded ulcerated surface measuring approximately 3.5 x 2 cm in size located on the dorsum of the left hand.*

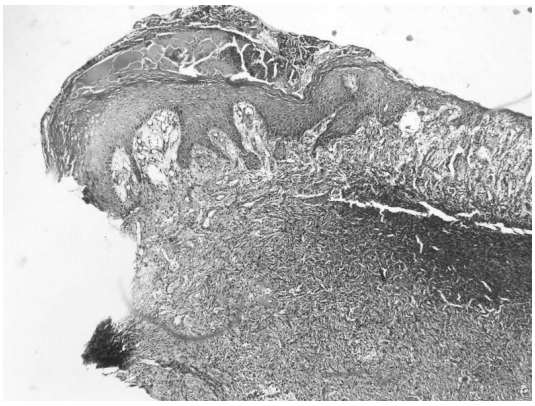

Figure 2. *Incision biopsy done on the nodule at the left hand showed acanthotic epidermis on low power (10x) magnification view (hematoxylin and eosin stain)*

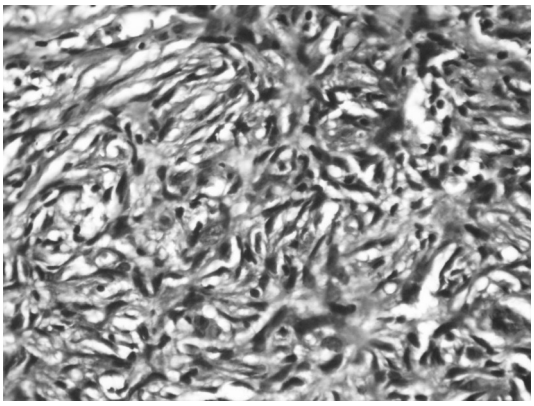

Figure 3. *Oil immersion field (400x) showed presence of closely spaced spindle cells with pleomorphic nuclei and mitotic figures in a storiform pattern in the dermis (hematoxylin and eosin stain)*

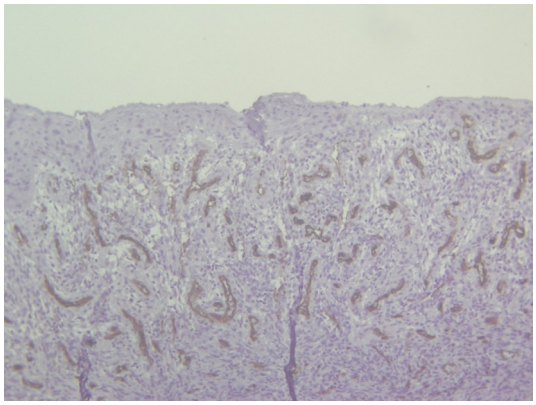

Figure 4. *CD34 Immunostaining is positive on low power (10x) magnification view*

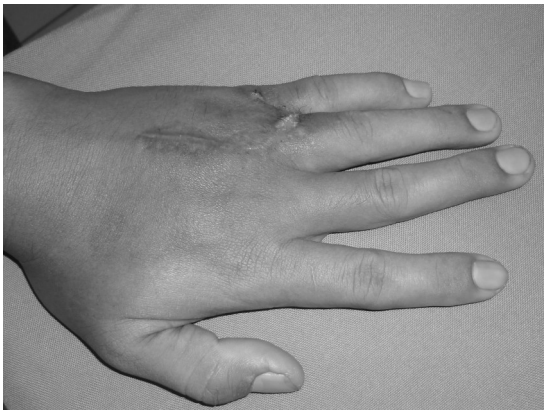

Figure 5. *Post-operative photo after wide surgical excision was done.*

Table I. Summary of Immunohistochemical staining of DFSP and other spindle cell dermal tumors

	CD34	Factor XIIIa	S-100	Actin	Desmin	Vimentin
DFSP	+	-	-	-	-	+
Dermatofibroma	-	+	-	+	-	+
Nerve Sheath tumors	+/-	-	+	-	-	-
Fibrosarcoma	-	-	-	+	-	+
Malignant Fibrous histiocytoma	+/-	-	-	-	+/-	+

+, More than 80% of cases staining; -, less than 20% of cases staining; +/-, variable staining.

DISCUSSION

Before 1924, several authors independently described DFSP, but it was the description of "progressive and recurrent dermatofibroma" by Darier and Ferrand that established it as a distinct clinicopathologic entity. In 1925, Hoffmann proposed the descriptive term dermatofibrosarcoma protuberans. Taylor and Helwig, later in 1962, emphasized the microscopic findings of a spindle-cell proliferation arranged in a cartwheel or whirligig pattern.[3]

DFSP is a rare tumor that constitutes less than 0.1% of all malignancies. The incidence in the United States has been estimated at 4.2 million cases per million population per year.[4] Data from the Jose R. Reyes Memorial Medical Center Dermatology Department showed 6 reported cases of DFSP from year 2000 up to present time. DFSP occurs in early to middle adult life between 20 and 50 years of age, although its range of occurrence is from birth through the 80s. An almost equal sex distribution with a slight male predominance was seen in literature.[5]

DFSP arises from the rearrangement of chromosomes 17 and 22 such that the collagen Type Iα1 gene (COLIA1) becomes fused to the platelet-derived growth factor (PDGF) β-chain gene (PDGFB). This rearrangement results in the deregulation of PDGF β-chain expression and leads to continuous activation of the PDGF receptor β (PDGFRβ) protein tyrosine kinase, which promotes DFSP cell growth.[6]

DFSP presents most commonly on the trunk (47%), followed by the lower extremity (20%), upper extremity (18%) and finally head and neck (14%).[7] It initially appears as an asymptomatic, indurated plaque that may be violaceous, red-brown or flesh-colored. It slowly and relentlessly enlarges and develops protuberant nodules within the plaque.[5]

On routine hematotoxylin-eosin-stained sections DFSP is characterized by a distinct storiform pattern, created by spindle-shaped cells arranged in an irregularly whorled pattern. The cells have been described as arranged in cartwheel-like arrays around collagenous centers. There may be only slight to moderate nuclear atypia, although mitoses can be present.[16] However, DFSP may sometimes be difficult to differentiate histologically from other fibrohistiocytic tumors such as dermatofibroma, fibromatosis and malignant fibrous histiocytoma. It is important to differentiate DFSP from dermatofibroma because their biological behaviour differs greatly.[17] Immunohistochemical stains can be used to augment routine histologic examination to aid in differentiation DFSP from other dermal spindle cell tumors. The immunohistochemical profile of DFSP as well as tumors often considered in the differential diagnosis is summarized in Table I. Many recent studies have shown the usefulness of CD34 in distinguishing DFSP from other dermal tumors, particularly dermatofibroma.[16] The human progenitor cell antigen CD34 is a surface antigen expressed on normal hematopoietic stem cells and in a significant proportion of acute leukemias. It is a single-chain transmembrane glycoprotein which seems to be involved in the process of leukocyte adhesion to vascular endothelium and may participate in acute and chronic inflammatory responses.[16] CD34 reactivity in DFSP was first reported in 1990 by Ramani, Bradley and Fletcher.[18] Thereafter, other studies have surfaced and most of them documented the sensitivity of CD34 staining in DFSP as ranging from 84% to 100%. CD34 was found to be strongly positive in the plaque areas where the neoplastic cells were slender and arranged in fascicles parallel to the skin surface. However, in nodular areas with a typical storiform pattern, the staining was uneven with strong to moderate staining in early nodular areas but weaker staining in fully developed nodules as seen in our case.[16]

DFSP of the hand is a rare condition with approximately only 8 cases reported in world literature since 1977 to 2009.[2,8-14] In the Philippines, no case reports of DFSP of the hand have been published from local journals, specifically from the Journal of the Philippine Dermatologic Society from 1990-2009.

Schvarcz LW presented a case of congenital DFSP on the dorsum of the hand treated by local radical surgical excision and skin closure. The function of the hand remained normal and no signs of recurrence were noted six months post-operatively.[8] Wirman et al. presented a case of a DFSP arising on the back of the hand, that was widely excised and the defect closed with a split thickness skin graft. There were no details regarding the follow-up of the patient.[9] Coles et al. reported a case of DFSP involving the thumb that was treated by excision biopsy. No evidence of tumour or lymphadenopathy is reported at six-month follow-up.[11] A case similar to the patient was reported by Demiri et al in 2006. They presented a 51-year-old woman with DFSP on the dorsum of the left hand and had experienced four recurrences of the

tumor following local excisions. Radical surgical excision with distal ulnar artery skin island flap was done as well as adjuvant radiation therapy following surgery. Forty months post-operatively, the patient had a functional hand without signs of recurrence and no evidence of disease.[2] Although DFSP is locally aggressive, no evidence of nerve involvement was noted in similar case reports of DFSP of the hand.

As with other sites of predilection of DFSP, DFSP of the hand is considered to be an intermediate malignant tumor with a high tendency to recur repeatedly if precedent management is not adequate.[15] The optimal treatment option is resection with wide margins, typically 2-4 cm. The likelihood of local recurrence after the procedure is less than 10%.[6] Relative 5-year survival rate of DFSP is 99.2%. It very rarely metastasizes to regional lymph nodes or distant sites, mainly the lungs.[2] Although our patient presented with recurrent DFSP, does not show findings in the review of systems and physical examination that would lead us to suspect metastatic disease. Hence, extensive evaluations with chest x-ray, computed tomographic scans, liver function tests, complete blood counts are not indicated at this time. Local recurrences are noted within 3 years of excision, but late recurrence of greater than 10 years has been reported.[5] Hence, patient is to be followed up every 6 months during the first 3 postoperative years and annually thereafter. Particular attention must be paid to careful inspection and palpation of the original excision site and regional lymph nodes on subsequent follow-ups.

CONCLUSION

Dermatofibrosarcoma protuberans is a rare soft tissue neoplasm with intermediate to low-grade malignancy. It presents as a large indurated plaque but may also exhibit a nodular pattern late in its course, similar to our case. Sites of predilection are the trunk, extremities, head and neck. DFSP of the hand can occur rarely. Although it is locally aggressive, there have been no reports of nerve involvement that would impede function of the hand in similar studies. Wide excision with 2-4 cm margin is warranted and recurrences after surgery have been minimal. Metastasis in DFSP is rare and it has a very high survival rate.

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