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DFSP- Study of Cases in Our Hospital

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Abstarct

Dermatofibrosarcomaprotuberans (DFSP) is a rare, slow-growing, fibrohistiocytic neoplasm with intermediate-to low-grade malignancy that commonly favors young to middle-aged adults. It is most commonly seen on the trunk followed by the proximal extremities and the head and neck [1,2]. The potential for distant metastasis is low, but DFSP frequently recurs locally after incomplete excision. The general immunostaining pattern of DFSP is CD34 positive and factor XIIIa negative. Mohs micrographic surgery (MMS) is the treatment of choice for DFSP.

Presenting a study of five cases of Dermatofibrosarcoma protruberans which were admitted to our hospital during the period of two years from Jan 2015 to December 2016. All five patients presented with large irregular growths over the body, slow-growing and accompanied with itching and bloody discharge. Four out of five were males and one patient was female. All five patients were thoroughly investigated, biopsy proved and then subjected for surgery. Immunological assessment was also done. Wide local excision with skin grafting was done. All had positive histopathology for DFSP. Four out of five patients came back for follow-up on a regular basis and have shown no recurrence. One patient was lost on follow-up.

Keywords: Dermatofibrosarcoma protruberans; Ulcerated growths; Immunostaining; Wide excision; biopsy.

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Introduction

DFSP is a low-grade, relatively uncommon, soft-tissue sarcoma that was originally described in 1924 by Darier and Ferrand, [1,2,3].

The term dermatofibrosarcomaprotuberans was coined by Hoffman in 1925 [3]. 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 [4,5]. It is most commonly seen among those in their third or fourth decade and favors the trunk (40–60%), followed by the proximal extremities (20–30%) and the head and neck (10%–16%) [1,2,5].

A slight male predominance has been reported among patients with DFSP [6]. The tumor has a low chance of metastasis, either to regional lymph nodes or distantly, but is aggressive locally. A local recurrence rate of DFSP of up to 60 percent has been reported [7].

The tumor most commonly presents as a slow-growing, asymptomatic, skin-colored, indurated, firm plaque that eventually develops into reddish-brown nodules that vary in size. A genetic link has been found in some patients with DFSP, which frequently (more than 90 percent) exhibits translocation of chromosomes 17 and 22 [18,19].

The general immunostaining pattern of DFSP is thus CD34 positive and factor XIIIa negative. Dermatofibromas are CD34 negative and factor XIIIa positive.

MMS is the treatment of choice for DFSP. Prior to MMS, surgical excision with 3- to 5-cm-wide margins was the recommended treatment, but was associated with high rates of local recurrence.

A recurrence rate of up to 20 percent with 3-cm surgical margins has been described [14].

Materials and Methods

AB, 35 year old male came to our hospital with history of swelling on the upper arm since one month. No history of trauma, fever or pain with rapid increase in size. On examination, there was a large, ovoid, hard, non-tender lump on the anterior aspect of left upper arm with multiple ulcerations and sero-sanguinous discharge. Patient was thoroughly investigated. CD34 was strongly positive. Wide excision with skin grafting was done.

EF, 28 year old female presented to our hospital with a slow growing hard lump on the posterior axillary fold on the right side. Her investigations confirmed DFSP. Wide excision with primary skin grafting was done. DFSP was confirmed by histopathology. On follow-up patient is doing well.

YZ, 45 year old male presented with hard, non-tender swelling over the left forearm which was gradually growing. His investigations confirmed DFSP. Wide excision with primary skin grafting was done. DFSP was confirmed by histopathology. Lost on follow-up.

XY, 39 year old male presented with ulcerated lesion on the back that was gradually increasing in size. Biopsy and immunological studies confirmed the diagnosis. Wide excision with primary skin grafting was done. Patient regularly visits the hospital. At present free of disease.

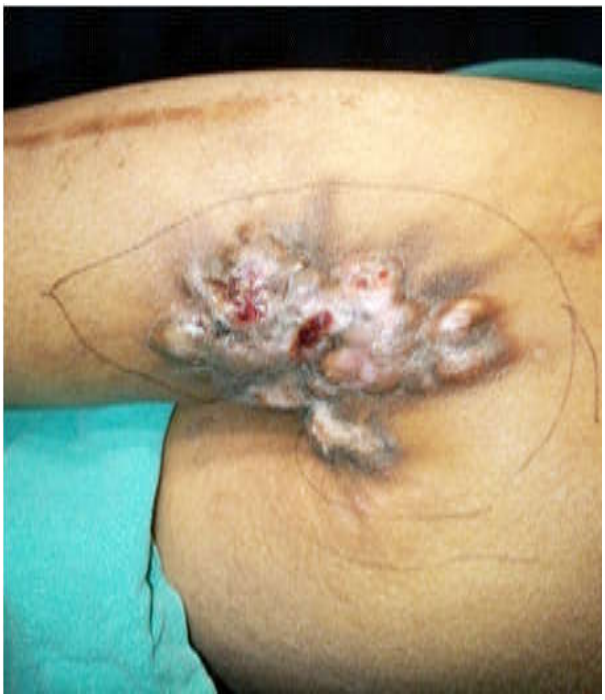


Fig. 1:

Fig. 2:

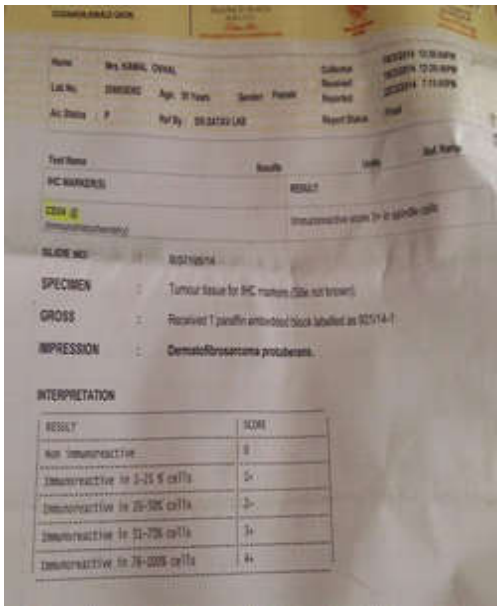
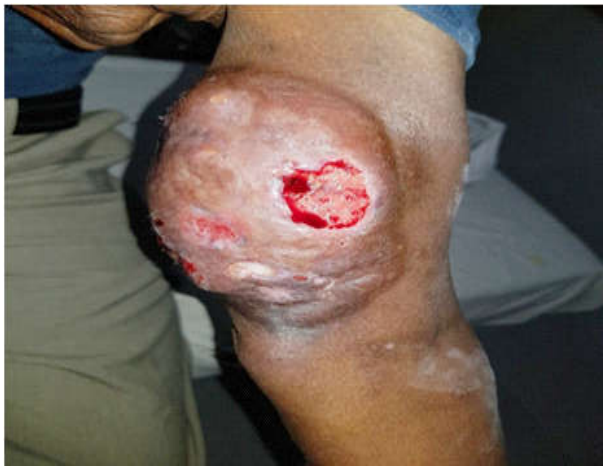


Fig. 3:



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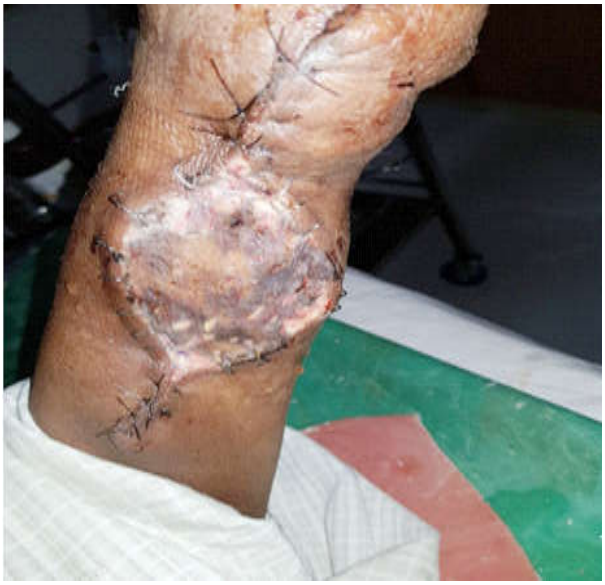

Fig. 5:

CD, 49 year old male came to out-patient department with a small nodular lesion, hard in consistency and non-tender. Excisional biopsy revealed DFSP. Confirmed by immunology. Follow-up continues with no evidence of recurrence at present.

Discussion

DFSP is a low-grade, relatively uncommon, soft-tissue sarcoma that was originally described in 1924 by Darier and Ferrand [1,2,3]. The term dermatofibrosarcomaprotuberans was coined by Hoffman in 1925 [1,3]. 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 [4,5]. It is most commonly seen among those in their third or fourth decade and favors the trunk (40–60%), followed by the proximal extremities (20–30%) and the head and neck (10%–16%) [2,5]. A slight male predominance has been reported among patients with DFSP [6]. It occurs less frequently in children, and congenital forms of DFSP have been reported in the literature [7]. The tumor has a low chance of metastasis, either to regional lymph nodes or distantly, but is aggressive locally. A local recurrence rate of DFSP of up to 60 percent has been reported [8]. The tumor most commonly presents as a slow-growing, asymptomatic, skin-colored, indurated, firm plaque that eventually develops violaceous to red-brown nodules that vary in size from one to several centimeters in diameter. DFSP may present as an atrophic plaque, which may resemble morphea and can be misdiagnosed as such [9].

A genetic link has been found in some patients with DFSP, which frequently (more than 90 percent) exhibits translocation of chromosomes 17 and 22, t [12,18]. This rearrangement fuses the collagen, type I, alpha 1 (COL1A1) gene to the platelet-derived growth factor B-chain (PDGFB) gene. The resultant rearrangement causes unregulated expression of platelet-derived growth factor leading to constitutive activation of the platelet-derived growth factor receptor (PDGFR). This step is believed to be a critical event in DFSP tumorogenesis. Germ-line mutations of p53 have been described in patients with DFSP and breast adenocarcinoma.

The biopsy typically shows the lesion to be located primarily in the dermis with irregular infiltration of the subcutaneous fat in a lace-like pattern. The epidermis is usually spared, but can be hyperplastic. The lesion usually comprises fairly uniform spindle cells with elongated nuclei and scanty pale cytoplasm. Pleomorphism is minimal or absent. The cells are typically arranged in a storiform or a mat-like pattern. Immunohistochemically, in the plaque stage, the spindle-shaped cells are strongly positive for CD34 immunostaining [18,19,20].

MMS is the treatment of choice for DFSP. Prior to MMS, surgical excision with 3- to 5-cm-wide margins was the recommended treatment, but was associated with high rates of local recurrence. The extent of invasion is difficult to ascertain because of its ability to penetrate not only cutaneous and subcutaneous tissue, but also underlying fascia and muscle. In a three-dimensional view, the tumor can be visualized as sending projections in different directions so even a wide excision may leave residual tumor in a single or multiple foci [12,13].

A recurrence rate of up to 20 percent with 3-cm surgical margins has been described [14]. Ratner, et al., reported that a standard wide excision with a width of 1cm would leave residual tumor in 70.7 percent of tumors, a width of 2cm would leave residual tumor in 39.7 percent of tumors, a width of 3cm would leave residual tumor in 15.5 percent of tumors, and a width of 5cm would leave residual tumor in 5.2 percent of tumors. The authors concluded that MMS is the treatment of choice for DFSB [10].

Nouri, et al., reported no recurrences in a series of 20 patients treated with MMS [8]. The mean number of MMS stages done to achieve clear margins was 2.5. Snow, et al., reported no local recurrences and a five-year local cure rate of 100 percent in 29 patients who were treated with MMS. In the literature review conducted by Snow, et al., nine out of 136 patients

treated with MMS developed local recurrences [21]. Five patients developed recurrences even after undergoing the Mohs procedure twice. The local cure rates reported in that paper were 93.4 percent and 98.5 percent after first and second Mohs surgeries, respectively [8,14]. In the Geisinger experience, no local recurrences were seen in 35 patients at a mean follow-up of 39 months.

Radiotherapy has been used as an adjuvant therapy after wide surgical excision or in those patients who have inoperable macroscopic disease. Postoperative radiotherapy has been associated with a cure rate of ≥ 85 percent [17]. Ballo, et al., evaluated a combination of conservative resection and adjuvant radiation therapy and noted a local recurrence rate of 5 percent [17]. Risks of adjuvant radiotherapy include acute and chronic radiodermatitis and further development of new skin cancers.

Imatinib, a tyrosine kinase inhibitor, has been approved to treat adult patients with unresectable, recurrent, and/or metastatic disease. Imatinib inhibits the platelet-derived growth factor receptor tyrosine kinase and has been effective in treating DFSP in some patients despite CD117 negativity [19].

The majority of local recurrences of DFSP occur within the first three years, with about half presenting within one year of surgery, but recurrences after five years have also been reported [18,20]. Thus, it is important to follow these patients over a long period after treatment.

Conclusion

DFSP is a rare disorder of low grade malignancy but must be cosidered in the differential diagnosis of non-tender hard soft tissue swellings presenting as ulcerated growths or nodular growths. Diagnosis is by biopsy and immunological tests. Immunohistochemically, in the plaque stage, the spindle-shaped cells are strongly positive for CD34 immunostaining. Wide local excision with primary skin grafting is the surgery of choice. As we did not encounter advanced cases or recurrences, other modalities of treatment were not given.

Conflict of Interest: None

Referrences

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