

A Fibrosarcomatous ("High-Grade") Variant of Dermatofibrosarcoma Protuberans (DFSP)

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Received date: May 15, 2014, **Accepted date:** Jun 21, 2014, **Published date:** Jun 25, 2014

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Abstract

Dermatofibrosarcoma protuberans (DFSP) is a relatively uncommon soft tissue tumor with intermediate-to-low grade malignancy. There is usually little nuclear pleomorphism and only low to moderate mitotic activity. Conventional DFSP, usually have fewer than 5 mitotic figures/10 high-power fields. We describe a case of a 74- year old male patient who presented to the surgical clinic with active bleeding from a painless mass in his right arm which has grown progressively over a period of two years. Local excision of the mass was performed. Histopathological evaluation showed dermatofibrosarcoma protuberans with high mitotic activity (15 mitotic figures/10 high-power fields and high nuclear pleomorphism. Therefore, it was diagnosed as a fibrosarcomatous (high grade) variant of dermatofibrosarcoma protuberans (FS-DFSP). Only 225 cases of DFSP-FS being described in the literature, thus, the importance of this case report arises due to the rarity of this clinical and histopathologic entity.

Keywords: Dermatofibrosarcoma protuberans; Sarcoma; Soft-tissue sarcomas; Malignancy

Introduction

Dermatofibrosarcoma protuberance accounts for about 6% of all soft-tissue sarcomas [1]. This disease is more common in men than in women and has a peak incidence during the third decade [2]. It may occur at sites of previous trauma. Lesions ranging from 1 cm to more than 25 cm have been described in the literature. Dermatofibrosarcoma protuberance most commonly involves the trunk, accounting for about half of all cases. The extremities, followed by the head and neck, are the next most common sites, but tumors may occur on any part of the body [3]. DFSP was first described in 1924 by Darier and Ferrand as "progressive and recurring dermatofibroma". Dermatofibroma is a nodular cutaneous tumor characterized by a prominent storiform pattern [4]. DFSP is a locally aggressive tumor with a high recurrence rate, but it rarely metastasize [5]. Patients often ignore these tumors because of their slow growth so they are usually left untreated for several months [3]. There is usually little nuclear pleomorphism and only low to moderate mitotic activity. Conventional DFSP, usually have fewer than 5 mitotic figures/10 high-power fields [6]. DFSP is characterized by the nearly consistent presence of CD34, the human progenitor cell antigen, in a significant proportion of its cells [7-9]. We describe the histopathological findings of a case of this rare tumor with high mitotic activity (15 mitotic figures/10 high-power fields and high nuclear pleomorphism.

Case report

We describe a case of a 74- year old male patient who presented to the department of general surgery at The Marmara University Hospital in Istanbul-Turkey in January/2011 with active bleeding from a painless mass in his right arm which has been growing progressively

over a period of two years. There was no history of trauma. The patient did not complain of fever or weight loss. Physical examination revealed a 4x6x5 cm well defined mass lesion located in the right arm. The mass was sessile, bloody, mobile, non-tender, with an irregular ulcerative surface and fixed to the overlying skin. Routine laboratory tests were within the normal range. The patient was admitted to our department for further evaluation. MRI revealed an oval shaped, soft tissue mass, epicentered in the subcutaneous fat of the right arm. The lesion caused a prominent bulge in the right arm. The lesion was superficial and minimally hypointense to the biceps muscle in T1A sequences. T2A sequences revealed significant hyperintense solid mass (Figure 1). The underlying muscle and humerus bone were normal. X-ray imaging of the chest showed no evidence of metastasis. The patient underwent urgent surgery and the mass was en bloc resected under general anesthesia. No invasion into the underlying fascia was seen macroscopically during the operation. After a short recovery period the patient was discharged. Macroscopic pathological examination reported a solitary, protuberant, gray mass (5.5x5x4.5 cm) involving the subcutis and the skin. Areas of hemorrhage were seen in the tumor. Microscopically, spindle cell proliferation was seen (Figure 2). Cells had no conspicuous pattern of proliferation, cellularity and pleomorphism were moderate to high and mitotic figures were high (15 mitotic figures per 10 high power fields (Figure 3). A diffuse CD34 (Qbend-10, 1-100 dilution, Dako, Carpinteria CA, USA) positivity in the tumor cells was shown immunohistochemically (Figure 4). Detection of the CD34 antibody was performed on a Dako automated immunostainer with universal detection kit streptavidin biotin-alkaline phosphatase/red/detection system Dako after enzyme (CD34) induced antigen retrieval. Pathological diagnosis was dermatofibrosarcoma with an annotation of fibrosarcomatous changes like pleomorphism, cellularity and high mitotic figures. Although the surgical margin was considered negative macroscopically, it was positive in microscopic examination.

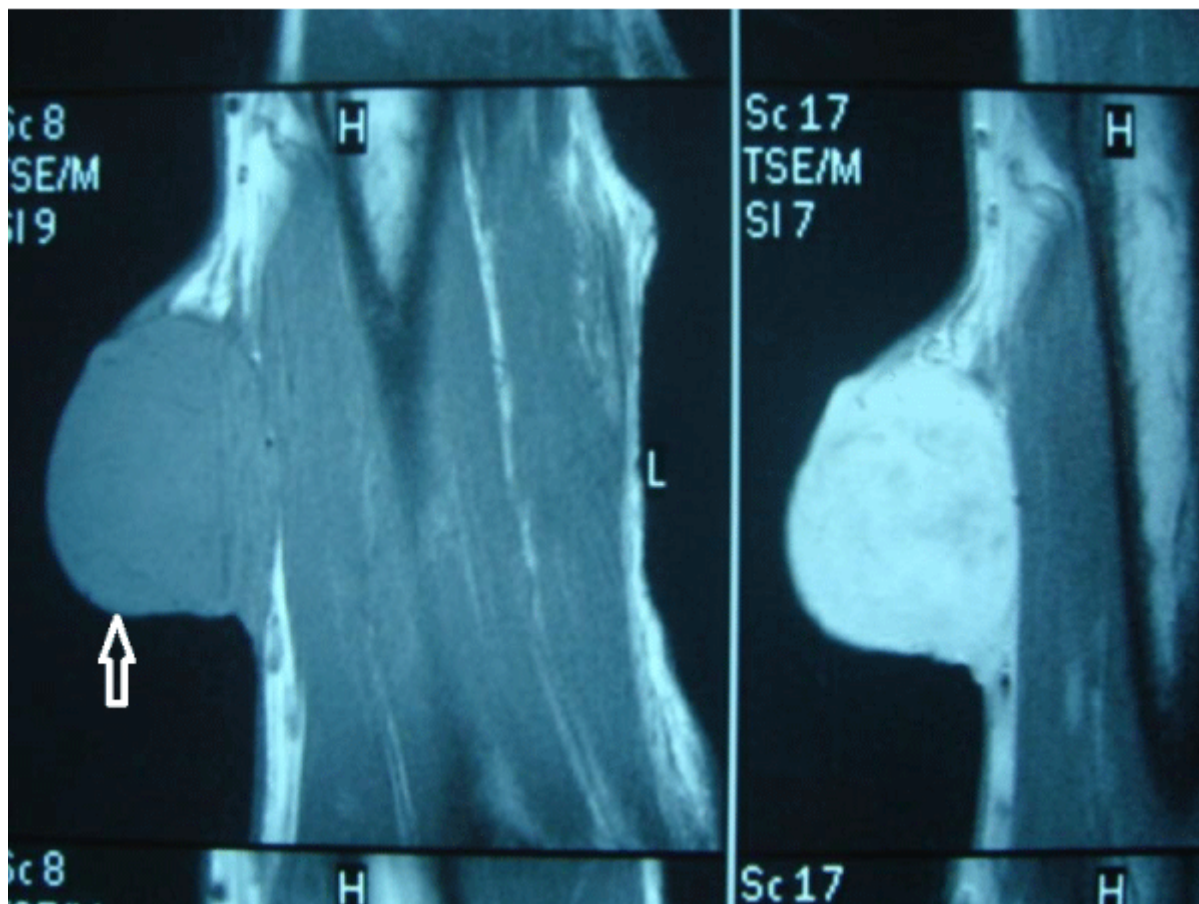


Figure 1: MRI of the left arm: (A) Coronal T1WI and (B) Axial T1WI showing an oval subcutaneous soft tissue mass (arrow) that is iso signal to the muscle.

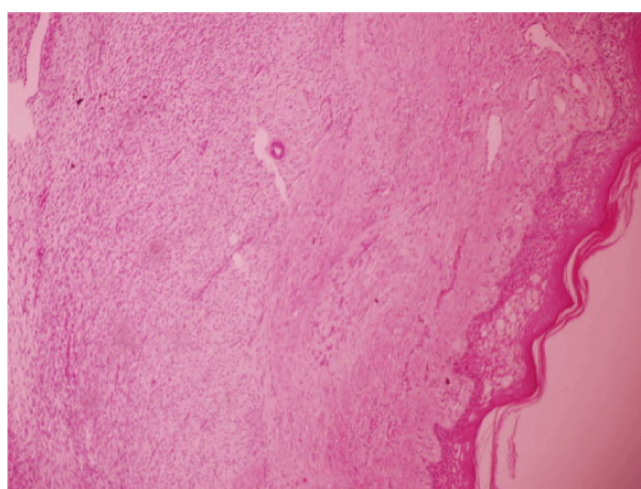


Figure 2: A superficial spindle cell proliferation was seen under light microscope. (Hematoxylin eosin, 40x).

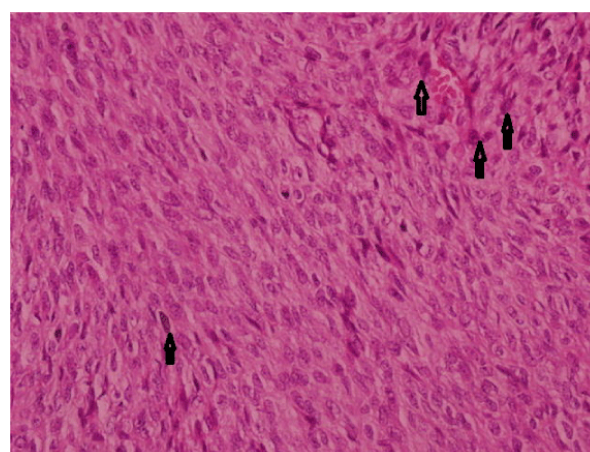


Figure 3: Photomicrograph of dermatofibrosarcoma showing moderate pleomorphism, high cellularity and frequent mitotic figures (arrows) (Hematoxylin eosin, 400x).

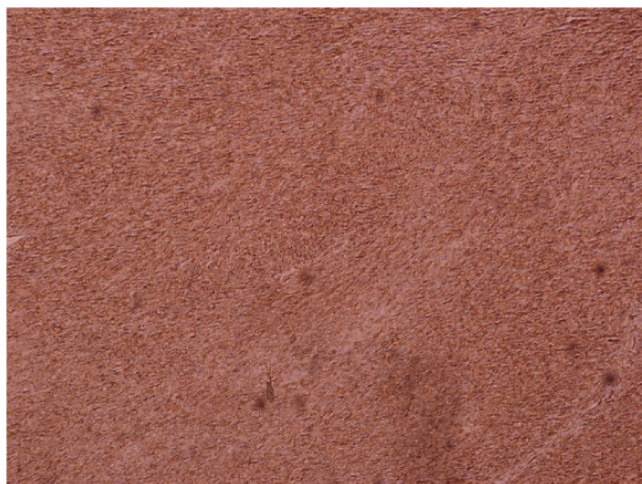


Figure 4: CD34 immunostain in a dermatofibrosarcoma. Diffuse cytoplasmic CD34 positivity was seen in tumor cells (CD34 antibody, counterstained with hematoxylin 40x).

Because of the positive resection margin, the patient received a total dose of 60Gy of radiotherapy starting 2 weeks after surgery. 3-year follow up showed no recurrence of the tumor. We learned that the patient died 3 years and 3 months after the operation because of an intracranial hemorrhage.

Discussion

Dermatofibrosarcoma protuberans (DFSP) is a relatively uncommon soft tissue neoplasm with an intermediate-to-low grade malignancy [10]. The tumor first appears as a single, red to bluish, blanchable, firm coetaneous nodule. During the late stages, the rate of growth accelerates, producing the characteristic protrusion from the skin. The growth rate is variable. Lesions may remain stable for many years or they may grow slowly with periods of accelerated growth. Local recurrences occur in 20-55% of the cases [10]. Conventional DFSP, usually have fewer than 5 mitotic figures/10 high-power fields [5]. DFSP is characterized by the nearly consistent presence of CD34, the human progenitor cell antigen, in a significant proportion of its cells. DFSP is genetically characterized by the t(17;22)(q22;q13), resulting in the fusion of alpha chain type 1 of collagen gene and platelet-derived growth factor beta gene. This translocation is present in 90% of DFSP and represents a very useful tool in the differential diagnosis of DFSP with other tumors with similar histology [11]. Reviewing the literature and to the best of our knowledge, the findings presented in this report are rarely seen histopathological findings of a case of this rare tumor with high mitotic activity (15 mitotic figures/10 high-power fields) and high nuclear pleomorphism and this is the first

case described histopathologically with these findings in Turkey. Reviewing the literature, we could find 24 reports containing 1422 patients with DFSP and 225 with fibrosarcomatous dermatofibrosarcoma protuberance (FS-DFSP) [12]. As a result, this case report is important due to the rarity of this clinical and histopathologic entity. Until this time only 225 cases of DFSP-FS were described in the literature.

In conclusion, dermatofibrosarcoma protuberance is a rare, usually superficial soft-tissue sarcoma, Conventional DFSP, usually have fewer than 5 mitotic figures/10 high-power fields. We have described rarely seen histopathological findings of a case of this rare tumor with high mitotic activity and high nuclear pleomorphism, which is described as a fibrosarcomatous dermatofibrosarcoma protuberance (FS-DFSP). Only few cases of DFSP-FS were described in the literature.

References:

1. Kransdorf MJ, Meis-Kindblom JM (1994) Dermatofibrosarcoma protuberans: radiologic appearance. *AJR Am J Roentgenol* 163: 391-394.
2. Shrimali R, Garg L, Setia V, Jain S (2002) Dermatofibrosarcoma protuberans (DFSP)-CT findings in DFSP-a rare skin tumor. *Indian J Radiol Imaging* 12: 357-358.
3. Torreggiani WC, Al-Ismaïl K, Munk PL, Nicolaou S, O'Connell JX, et al. (2002) Dermatofibrosarcoma protuberans: MR imaging features. *AJR Am J Roentgenol* 178: 989-993.
4. Darier J, Ferrand M. (1924) Dermatofibromes progressifs et récidivants ou fibrosarcomes de la peau. *Ann Dermatol Syphiligr* 5: 545-562
5. Rutgers EJ, Kroon BB, Albus-Lutter CE, Gortzak E (1992) Dermatofibrosarcoma protuberans: treatment and prognosis. *Eur J Surg Oncol* 18: 241-248.
6. Taylor HB, Helwig EB (1962) Dermatofibrosarcoma protuberans. A study of 115 cases. *Cancer* 15: 717-725.
7. Goldblum JR, Reith JD, Weiss SW (2000) Sarcomas arising in dermatofibrosarcoma protuberans: a reappraisal of biologic behavior in eighteen cases treated by wide local excision with extended clinical follow up. *Am J Surg Pathol* 24: 1125-1130.
8. Goldblum JR, Tuthill RJ (1997) CD34 and factor-XIIIa immunoreactivity in dermatofibrosarcoma protuberans and dermatofibroma. *Am J Dermatopathol* 19: 147-153.
9. Weiss SW, Nickoloff BJ (1993) CD-34 is expressed by a distinctive cell population in peripheral nerve, nerve sheath tumors, and related lesions. *Am J Surg Pathol* 17: 1039-1045.
10. Asuquo ME, Umoh MS, Ebughe G (2007) Dermatofibrosarcoma protuberance: case reports. *Ann Afr Med* 6: 80-83.
11. Llombart B, Serra-Guillén C, Monteagudo C, López Guerrero JA, Sanmartín O (2013) Dermatofibrosarcoma protuberans: a comprehensive review and update on diagnosis and management. *Semin Diagn Pathol* 30: 13-28.
12. Liang CA, Jambusaria-Pahlajani A, Karia PS, Elenitsas R, Zhang PD, et al. (2014) A systematic review of outcome data for dermatofibrosarcoma protuberans with and without fibrosarcomatous change. *J Am Acad Dermatol*: 1245- 1246.

This article was originally published in a special issue, entitled: "**Skin Cancer**", Edited by Dr. Homer S. Black, Baylor College of Medicine, USA