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Case Report

A rare case of dermatofibrosarcoma protuberans of the forefoot

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ABSTRACT

Dermatofibrosarcoma protuberans is an extremely rare, potentially malignant tumor type that usually presents on the trunk or proximal extremities. The clinical presentation includes a gradually enlarging painless plaque-like or nodular lesion of the skin with surrounding red to blue discoloration. The diagnosis is based on clinical presentation, computed tomography or magnetic resonance imaging, and biopsy with histologic analysis. An early and timely diagnosis improves chances of complete surgical resection thus improving prognosis. Herein, we present a rare case of dermatofibrosarcoma protuberans with the hopes that its addition to the literature will aid in the earlier recognition of future patients and help prevent this potentially curable disease from becoming deadly.

Introduction

Dermatofibrosarcoma protuberans (DFSP) accounts for approximately 1%-6% of all soft-tissue tumors [1,2]. It has an annual incidence of 4.2 per million [3]. Although there have not been many extensive studies performed that identify the differences in the incidence of DFSP across race and sex, preliminary data points toward DFSP being approximately twice as common in blacks as compared with whites and equally distributed between males and females [3]. The tumor is found to be located on the trunk in 40%-50% of cases, the chest and shoulders in 30%-40% of cases, and the proximal portion of the limbs in 10%-15% of cases. Some studies report a greater frequency of distally located DFSP in children. One study of 27 cases, reports that 14.8% of childhood DFSP was located on the hands or feet [4]. It presents most frequently between the ages of 20 and 50 years [1]. Clinicians should be made aware that DFSP is known to occur among children. Because it occurs less commonly in this patient population, it is frequently misdiagnosed or underdiagnosed.

Case report

A 14-year-old boy with a history of a soft-tissue mass on the dorsum of his left foot since age 5 presented to the hospital

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because of a markedly increased growth rate of the mass over the last 3 months, see Figure 1. During the same period, the mass began eluting a serous fluid through separated skin margins over the 2nd and 3rd toes. He had developed areas of skin loss on the lateral aspect of the foot overlaying the 5th metatarsal and the anterolateral aspect of his ankle in an approximately vascular distribution. Physical examination confirmed a large ulcerating mass over the dorsum of the left foot with decreased sensation of the overlying skin. Magnetic resonance imaging (MRI) confirmed a 10 × 15 × 18-cm ovoid mass on the dorsum of the left foot, see Figure 2. Incisional biopsy results were consistent with DFSP, see Figure 3. After the biopsy results, surgical removal of the lesion was carried out to remove the locally invasive tumor.

Discussion

DFSP is a fibrohistiocytic tumor of intermediate malignancy characterized by a nodular cutaneous mass. It is most frequently located on the trunk and proximal extremities and has a propensity for recurrence. Because of its indolent growth, it is hypothesized that these tumors frequently arise during childhood but only become apparent during young adulthood [5]. Giant cell fibroblastoma (GCF) is considered to be the juvenile form of DFSP [1]. Initially, it manifest as a firm, plaque-like lesion of the skin with surrounding red to blue discoloration. Rarely, these tumors present as an area of atrophy or small subcutaneous nodules rather than a plaque [5].

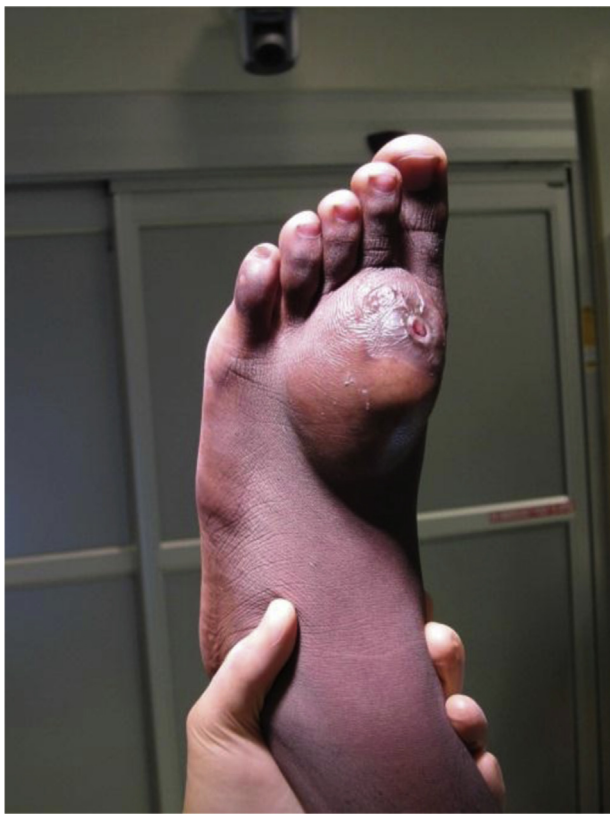

Fig. 1 – A large fungating mass present preoperatively on the left foot of a 14-year-old boy.

Prior trauma is reported in up to 20% of cases and larger lesions can ulcerate, bleed, and become painful.

The tumor is characterized histologically by surface bound CD34 and the absence of factor XIIIa, which are used to differentiate it from other soft-tissue tumors [6]. Molecular characterization of DFSP has identified an association with the chromosomal translocation t(17;22)(q22;q13) and with supernumerary ring chromosomes containing material from chromosomal regions 17q22 and 22q13 accompanied by simple chromosome trisomies. These genetic aberrations fuse the COL1A1 and PDGF beta genes, resulting in PDGF beta being under the transcriptional control of the COL1A1 promoter. This gives rise to an overexpression of PDGF beta, which leads to the constitutive activation of the platelet-derived growth factor subunit B (PDGFB) receptor, a type III tyrosine kinase receptor leading to autocrine stimulation and tumorigenesis [1,7,8].

Multiple histologic variants of DFSP exist. These variants include myxoid DFSP, the Bednar tumor, the atrophic variant of DFSP, and GCF. The myxoid variant is characterized by the presence of moderately cellular areas made up of stellate or fusiform cells with abundant accumulation of hyaluronidase-sensitive mucin in the intercellular space. The Bednar tumor has a pigmented storiform appearance. The atrophic variant of DFSP is characterized by reduced thickness of the dermis and replacement of much of the dermis and subcutis by spindle cells. Finally, the GCF is often myxoid and punctuated by pseudovascular tissue spaces being lined by multinucleated giant cells [1].

Presumptive diagnosis of DFSP can usually be made based on clinical appearance alone do to its superficial location and characteristic findings. However, the ease of diagnosis of less superficial tumors can be enhanced through the use of imaging techniques. The radiologic appearance of DFSP is characterized by a nodular soft-tissue mass involving the skin and subcutaneous adipose tissue with a lack of mineralization [9]. The case we present here provides unique radiologic images that can be used as comparisons for physicians that are treating patients with potential DFSP.

Both computed tomography (CT) and MRI can be used to define the underlying structure of a given lesion. In 1 study of 14 cases, CT generated minimally enhanced images in 3 of the cases, heterogeneously enhanced images in another 3 cases, and homogenously enhanced images in 8 cases. In the same study, all 14 lesions imaged with MRI were T1-isointense to muscle. Ten lesions were T2-hyperintense and 4 were T2-isointense to T2-hypointense to muscle. T1 postcontrast enhancement patterns ranged from mild to markedly heterogeneous or markedly homogeneous [10].

MRI has stood out as the modality of choice for margin definition in planned operative procedures for DFSP because of the increased resolution available with higher Tesla (3-4T) magnets as compared with CT. MRI also surpasses CT in its utility for imaging DFSP because of the ability to produce images with unique imaging protocols: T1, T2, frequency-selective fat suppression, and short tau inversion recovery (STIR). DFSPs are usually hypointense to fat on T1-weighted images and hyperintense to isointense to fat on T2-weighted images. Specialized techniques such as frequency-selective fat suppression and STIR are frequently used to accentuate the pathology on MRI [11].