Introduction

Dermatofibrosarcoma protuberans (DFSP) is a rare, locally aggressive cutaneous mesenchymal skin tumor (1). Typically it presents as a solitary, slowly growing, painless cutaneous nodule. After a period of inactivity it can grow slow and then cause characteristic protuberant nodules. Most cases of observed in the third and fourth decades of life, but newborns and children can be affected (2). Several histological variants of DFSP have been described (3, 4). Here we report a patient with myxoid DFSP, rarely reported in literature.

Case Report

A 35-year-old man was presented with a 5x2 cm in size, violaceous, painless, enlarging nodular lesion on the lower abdomen (Figure 1). The lesion had been present for approximately nine years and was gradually increasing in size. In the affected area had no previous history of trauma or any preexisting skin lesions. Clinical examination showed erythematous to brownish nodule, with reddish plaque on lower abdomen. He denied any treatment of the lesion in the past. No other lesion was noted. The lymph nodes in the head and neck region and the axillae were nonpalpable. A skin biopsy was performed. On microscopic examination, the tumor consisted of spindle-shaped cells that were strongly positive for CD34 with a typical storiform pattern and myxoid stromal changes.

Histological examination revealed atypical spindle cells with myxoid changes in some areas. The atypical spindle-shaped cells, some of which infiltrating adipose tissue, showed storiform pattern. Five mitotic figures per 10 high power field were counted in the tumor. Immunohistochemical staining showed no staining with CD31, CD68 and S-100 (Figure 1). Diagnosis of DFSP was made based on the histological and immunohistochemical findings. The patient’s lesion was totally removed. There had been no recurrence during 1 year follow-up of patient.

Discussion

DFSP is a low grade spindle cell neoplasm of fibroblastic differentiation of the dermis and underlying soft tissue. Estimates of the overall incidence of DFSP in the United States are 0.8 to 4.5 cases per million persons per year (1, 2). It can be seen in all races. DFSP is found in similar distribution in men and women, although some large series suggest a slight male predominance (5). DFSP most commonly occurs between 20 and 50 years of age, although its can appear at any age. It occurs less frequently in children, and congenital forms of DFSP have been reported (2).

Pathogenesis of dermatofibrosarcoma protuberans (DFSP) is unknown. Laboratory studies have shown that chromosomal aberrations (translocation of chromosomes 17 and 22) may contribute to the pathogenesis of DFSP (6, 7). In 10-20% of patients with this tumor, trauma at the site seems to be incriminated. Surgical and burn scars, radiodermatitis, vaccinations and sites of central venous lines have all been reported as sites of DFSP (8-10). In our patient, in the affected area had no previous history of trauma or any preexisting skin lesions.

DFSP most commonly presents as a slow growing, asymptomatic, erythematous, flesh-colored, or violaceous plaque that slowly enlarges over months.
to years. It becomes raised, firm, and nodular (1). It also can present as a nonprotuberant, atrophic, violaceous lesion resembling morphea, sclerosing basal cell carcinoma, lipoma, dermatofibroma or scar (5).

The most common location for a DFSP is on the trunk and proximal extremities, generally on the chest and shoulders followed by the proximal extremities, head and neck. Lesion size usually ranged from 1-5 cm but can reach up to 20 cm in advanced cases (1).

The low-grade DFSP (85-90 %) is the most common condition. The remaining 10-15% constitute the high-grade variant, which is more frequently associated with local recurrence and metastasis (11, 12). The potential for distant metastasis is low (< 5%), most often to the lung, also to regional lymph nodes, bone, heart, and brain (5, 13, 14).

Definitive diagnosis requires a core needle or incisional biopsy. The correct diagnosis can be made by review of hematoxyline and eosin-stained sections, combined with immunohistochemical staining. DFSP is composed of dense, monomorphic cells with spindle-shaped nuclei that are arranged in a storiform or matlike pattern in the center of tumor nodules, and infiltrate the dermal stroma peripherally. Immunohistochemical expression of CD34 has been considered characteristic for the diagnosis of DFSP. Approximately 80%-100% of DFSP express this marker, although between 10% and 20% are negative. Factor XIIIa is very useful in the differential diagnosis between DFSP and cellular fibrous histiocytomas, as it is usually negative in DFSP (13,15).

Several histological variants of DFSP have been described including the Myxoid, Fibrosarcomatous, Pigmented (Bednar tumor), Giant cell fibroblastoma, Atrophic DFSP, Sclerosing DFSP and Granular cell types (3, 4). Myxoid DFSP doesn’t differ from conventional DFSP in terms of clinical characteristics or prognosis. It’s contain an abnormal type of connective tissue that is called myxoid stroma. Almost all cases are positive staining for CD34, and negative for other markers, such as S-100, desmin.
and actin (16). Our histopathology also showed typical features, characteristic of DFSP.

Treatment is primarily surgical, with chemotherapy and radiation therapy sometimes used. Wide surgical excision is the standard treatment in localized, resectable cases. Mohs micrographic surgery is the treatment of choice for recurrent tumors. The high recurrence rate for conventional Radiotherapy has been used as an adjuvant therapy after wide surgical excision or in those patients who have inoperable macroscopic disease. Imatinib is a tyrosine-kinase inhibitor used in the treatment of multiple cancers. It is gold standard for treatment of inoperable, recurrent and/or metastatic DFSP (17).

We presented a case of myxoid DFSP, rarely reported in literature. Dermatologists should be aware of this rare entity and should make wide excision and reduce risk of recurrence.

References