CASE REPORT

Atrophic dermatofibrosarcoma protuberans and enlargement with pregnancy: Case report and literature review

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Received 10 May 2011; accepted 10 September 2011
Available online 25 November 2011

KEYWORD
Dermatofibrosarcoma protuberans (DFSPs)

Abstract Dermatofibrosarcoma protuberans (DFSPs) are a slowly growing locally aggressive tumor of disputed histogenesis with low-grade malignancy and a marked tendency to local recurrence but rarely metastasize to distant sites. Few reports have suggested that DFSPs may enlarge more rapidly during pregnancy. This article describes a lesion of the atrophic variant of DFSP that increased in size during pregnancy which is an unusual presentation, and a review of the literature.

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1. Introduction

Dermatofibrosarcoma protuberans (DFSPs) are a rare fibroblastic mesenchymal skin tumor. The most commonly affected area is the trunk followed by the extremities, scalp, and neck. DFSP is a low-grade malignancy of the skin and subcutaneous tissues with low potential for the developing distant metastases.

The most common presentation is a firm, indurated plaque, often skin-colored with red brown exophytic nodules. It also can present as a non-protuberant, atrophic, violaceous plaque resembling morphea. The epidermis may be atrophic and focal ulceration may occur. Uncommon pigmented variant, called Bendar tumor, which is an exceedingly rare tumor accounts for 1–5% of all cases of DFSP. It is characterized by a usually scant (1–5% of cells) population of dendritic melanocytes within an otherwise typical DFSP. Less than 5% of DFSPs are associated with metastases and many of these show either a fibrosarcomatous component or, much more rarely, malignant fibrous histiocytoma [MFH] “like appearance” (Bisceglia et al., 1997).

There are only few reports in the literature describing dermatofibrosarcoma protuberans that enlarged considerably during pregnancy. This article describes a case of the atrophic variant of DFSP and enlargement of the lesion during pregnancy which is an unusual presentation.

2. Case presentation

A 30 year old Saudi lady presented on March 21st 2010 complaining of asymptomatic lesion over the left side of the
upper abdomen for 2 years that increased in size during pregnancy 9 months ago. There was no history of trauma, bleeding or discharge. She had \(6 \times 5\) cm firm subcutaneous tumor with an overlying central depressed atrophic brownish tethered morphea like skin, in the left side of the upper abdomen with a visible nodule at the upper edge of the lesion. The tumor is mobile and not fixed to deeper structures (Fig. 1) and lymph nodes were not palpable.

Routine blood count, renal function, liver function tests, ANA and CT chest were all normal.

Skin biopsy showed poorly circumscribed proliferation of spindle cells that are arranged in interlacing fascicles producing storiform pattern, infiltrating the subcutaneous fat and isolating adipocytes forming honey comb or swiss cheese pattern (Figs. 2 and 3). CD34 staining is strongly positive in the spindle cell (Fig. 4). Immunohistochemical studies were negative for factor XIIIa, estrogen and progesterone receptors.

3. Discussion

Dermatofibrosarcoma protuberans (DFSPs) were first described by Darier and Ferrand in 1924 and named by Hoffmann in 1925. The tumor usually occurs in adults of age 20–50 years. Rarely, DFSP has been reported in newborns and elderly individuals (Thornton et al., 2005).

The most common location is the trunk (62%), followed by the extremities (25%) and the head and neck regions (13%) (Lindner et al., 1999). DFSP is an uncommon skin tumor with a high potential for local recurrence if not adequately excised. Most local recurrences, which can occur in 20–49% of cases, are noted within the first 3 years after excision, but late recurrence has been reported as well (Gloster, 1996). The recurrence rate is the highest in DFSP of the head and neck because of cosmetic and functional restrictions of resecting large areas (Gloster, 1996). The greater the number of recurrences the more likely the tumor is to disseminate (Lindner et al., 1999). Metastases are rare. A review of 913 cases of DFSP described regional lymph node metastases in about 1% and distant metastases in approximately 4% (Gloster, 1996). The lungs are the most frequent site of metastases, but metastases to brain, bone, and heart have also been reported (Garcia et al., 1996). Metastases usually occur within 6 years (Lindner et al., 1999). DFSP is genetically characterized by the unbalanced chromosomal translocation t(17;22)(q21;q13), usually in the form of a supernumerary ring chromosome. The product of this chromosomal translocation...
is the chimeric gene COL1A1-PDGFB (collagen type I alpha 1-platelet-derived growth factor beta), which is amplified at low levels in the ring chromosome which triggers the proliferation of DFSP tumor cells through PDGF receptor tyrosine kinase (Abbott et al., 2006).

Imatinib is an oral class of medications called protein-tyrosine kinase inhibitors that inhibits the platelet-derived growth factor receptor (PDGF-R) signaling cascade which plays a crucial role in the pathogenesis and tumor growth of DFSP. Imatinib mesylate is indicated for the treatment of adult patients with unresectable, recurrent, and/or metastatic DFSP. A response rate of approximately 65% has been achieved among DFSP patients treated with imatinib (McArthur, 2007).

Mohs micrographic surgery (MMS) with a surgical margin of 2.5 cm deep to fascia may be the treatment of choice for DFSP, because of its high cure rate and maximal conservation of tissue. As reported in many studies the recurrence rate by MMS was 0.6–1.6%, while the average recurrence rate was 18% when DFSP was treated with wide excision (Gloster et al., 1996).

There are only few reports in the literature describing DFSP that enlarged considerably during pregnancy. In the report of Har-Shai et al. (1993) they presented 2 patients in whom dermatofibrosarcoma protuberans appeared and grew rapidly during pregnancy. Immunohistochemical studies were negative for estrogen and progesterone receptors. Fibrosarcomatous change during pregnancy had also been described by Cakir B arising in DFSP on the scalp (Cakir et al., 2003). Three additional cases of DFSP that showed accelerated growth during pregnancy were reported by Parlette et al. (1999). The tumors in all 3 patients, and 4 additional DFSPs from 2 male and 2 female subjects, showed expression of progesterone receptor.

Our patient is peculiar in that she showed considerable enlargement of the tumor during pregnancy. The immunohistochemical studies were negative for estrogen and progesterone receptors similar to the report of Har-Shai et al.

As with many other stromal neoplasms, DFSP appears to express low levels of hormone receptors, which may be one factor that accounts for their accelerated growth during pregnancy.

In conclusion the atrophic variant of DFSP could be misdiagnosed as morphea, therefore dermatologist should be aware of this uncommon but characteristic presentation of DFSP and a close follow up of the lesions during pregnancy for size changes, and for any sarcomatous deterioration within the tumor.

References


