An Unusual Soft Tissue Tumor: Dermatofibrosarcoma Protuberans: Differential Diagnosis

Nadir Görülen Bir Yumuşak Doku Tümörü: Dermatofibrosarkom Protuberans

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Geliş Tarihi/*Received:* 28.05.2008
Kabul Tarihi/*Accepted:* 18.09.2008

The article was presented as a poster in 5th EADV Symposium, İstanbul/ Turkey May 22-25, 2008

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ABSTRACT Dermatofibrosarcoma protuberans is a rare, locally aggressive, dermal and subcutaneous fibrohystiocytic tumor that commonly appears in adult patients. The etiopathogenesis of dermatofibrosarcoma protuberans remains unclear. It is characterized by slow growth and a high rate of recurrence but limited potential for metastasis. The most common presentation is an indurated plaque with red-brown exophytic nodules on the trunk and proximal extremities. The diagnosis is made by histopatological examination. Wide excision is the conventional treatment of dermatofibrosarcoma protuberans. Herein we describe a case of a 39-year-old woman who has had a giant dermatofibrosarcoma protuberans on the abdominal wall for fifteen years and was treated with surgical excision.

Key Words: Dermatofibrosarcoma; soft tissue neoplasms

ÖZET Dermatofibrosarkom protuberans sıklıkla erişkinlerde görülen, lokal agresif, dermal ve subkutanöz dokudan kaynaklanan nadir bir fibrohistiositik tümördür. Etiyopatogenezi tam olarak anlaşılamamıştır. Yavaş büyüme hızı, sık rekürrens ve nadir metastazla karakterizedir. En sık rastlanan formu gövde ve proksimal ekstremitelerde yerleşen indüre plak üzerindeki ekzofitik tümöral lezyonlardır. Tanı histopatolojik inceleme ile konur. Konvansiyonel tedavisi geniş eksizyondur. Karın duvarında 15 yıldır devam eden büyük nodüler lezyona dermatofibrosarkom tanısı konan ve cerrahi eksizyonla tedavi edilen 39 yaşında bir kadın olgu sunulmaktadır.

Anahtar Kelimeler: Dermatofibrosarkom; yumuşak doku neoplazmları

Turkiye Klinikleri J Med Sci 2009;29(1):285-7

Dermatofibrosarcoma protuberans (DFSP) is a rare, locally aggressive dermal and subcutaneous fibrohystioctyic tumour that commonly appears in adult patients. It accounts for less than 0.1% of all malignant neoplasms and represents 2-6% of all soft tissue sarcomas.[1] We present a case of giant DFSP because of its rare occurrence.

A 31-year-old woman presented with a history of multiple asymptomatic swellings on her abdominal wall approximately for 15 years. Her self and family history was unremarkable. Dermatological examination revealed firm, livid colored four tumerous lesions, localized to a well-defined, indurated, red-brown, morphea-like plaque of 10 x 20 cm diameters above the umbilicus (Figure 1). Routine laboratory investigations were within normal ranges and chest X-ray and abdominal computerized tomography reve-

aled no abnormalities. On the histopathological examination, a tumoral lesion extending to the subcutis was determined. There was no grenz zone between the epidermis and the tumor. The tumor composed of interwoven bundles of spindle cells with plump nuclei arranged in a storiform or cartwheel pattern (Figure 2). Immunostaining of CD34 was strongly positive (Figure 3). A diagnosis of DFSP was made. Tumerous lesions were excised together with the indurated plaque. There was no occurrence during the 1 year follow-up period.

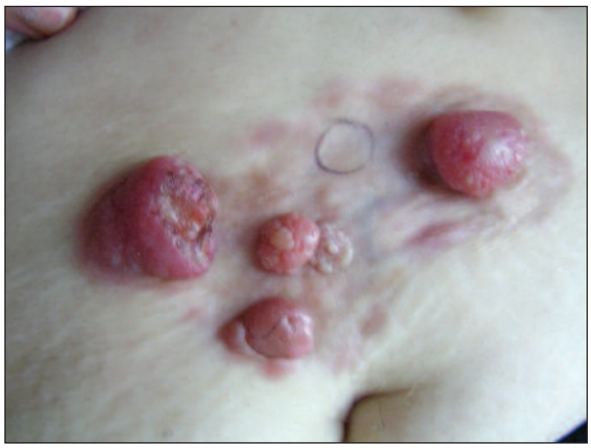

FIGURE 1: Tumerous lesions, localized to well defined morphea-like plaque.

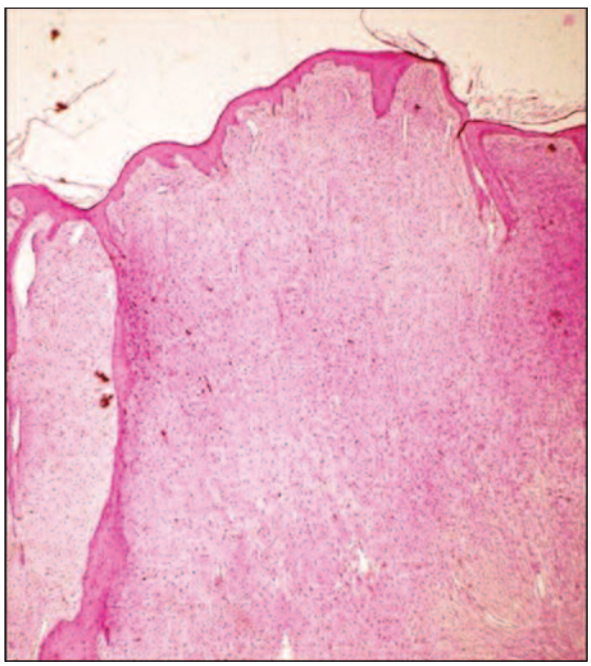

FIGURE 2: Tumoral infiltration extending to the subcutis (HE, x10).

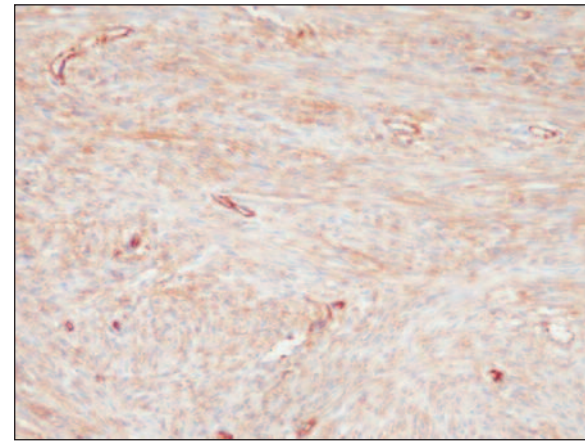

FIGURE 3: Immunostaining of CD34 x, 20.

DISCUSSION

The histiogenesis of DFSP remains unclear. Fibrohistiocytic, fibroblastic, periadnexial dendritic and neural-related differentiation have all been suggested.[1] Reports indicate that 33% of patients had a history of prior trauma, which may be related to the site of occurrence or the nodular appearance of the tumor.[2] DFSP arises from the arrangement of chromosomes 17 and 22, with the fusion between the collagen type 1α1 gene (COLIA1) and platelet-derived growth factor (PDGB) β-chain expression and activation of PDGF receptor β (PDGFRb) protein tyrosine kinase.[3,4]

The initial clinical presentation of DFSP is characterized by a slow growing raised asymptomatic lesion; it eventually enters a more rapid growth phase that may result with one or more nodules. Due to its indolent onset, the patient may present for evaluation when the tumor is several centimeters in size, like our patient. She has waited for 16 years for a dermatological examination. The clinical morphology of DFSP is variable. The most common form is a firm, indurated plaque with red-brown exophytic nodules as in our patient. It also may present with nontuberant forms like atrophic, violaceous lesions resembling morphea, anetoderma, sclerosing basal cell carsinoma, angioma-like lesions or scar.[1,5] Although DFSP is highly invasive it rarely metastasizes even after local recurrence. We did not determine any metastases and recurrence after 1 year. As in our

patient, the most common site of this lesion is the trunk as in our patient, followed by the extremities.

Diagnosis is made histopathologically. DFSP usually exhibits dense, atypical, spindle shaped fibrocytes arranged in a characteristic cartwheel pattern. The proliferation usually infiltrates the subcutaneous adipose tissue. In immunohistochemical examination, CD34 is a useful marker to distinguish DFSP from other soft tissue proliferations like deep penetrating dermatofibroma and cellular benign fibrous histiocytoma. DFSP, in contrast to dermatofibroma, reacts negatively towards anti-factor XIII.[1,5] Our patient had typical histopathological appearance and CD34 staining was positive.

Wide excision remains the mainstay of treatment for DFSP with an expended local recurrence rate of less than 10%.[6] Mohs surgery was reported to be an extremely effective method of resection, with a low rate of local recurrence.[1,6] Adjuvant radiotherapy, chemotherapy and imatinib may be helpful to decrease local recurrence rates.[1] In our patient, wide excision was performed.

In conclusion, DFSP is an uncommon cutaneous malignancy and may be confused with several benign and malignant tumors. In clinical settings, indolent and slow growing tumors should be considered suggestive for DFSP and a histopathological examination should be made.

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