Case Report

From Multiple Dermatofibroma to Spindle Cell Sarcoma
Different Types of Fibrohistiocytic Tumors in Same Patient

Tuğba Kevser Uzunçakmak,1* MD Ayşe Serap Karadağ,1 MD Bengü Nisa Akay,2 MD Ayşe Bahar Ceyran,3 MD Necmettin Akdeniz,1 MD

Address: 1 Istanbul Medeniyet University School of Medicine Goztepe Training and Research Hospital Department of Dermatology
2 Ankara University School of Medicine Department of Dermatology
3 Istanbul Medeniyet University School of Medicine Goztepe Training and Research Hospital Department of Pathology Istanbul, Turkey
E-mail: drtugbakevser@gmail.com
Corresponding Author: Dr. Tugba Kevser Uzuncakmak, Istanbul Medeniyet University, Goztepe Research and Training Hospital, Dermatology Istanbul, Turkey

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Abstract

Introduction

Dermatofibroma (DF) is a common, benign tumoral proliferation of histiocytes, fibroblasts and myofibroblasts in dermis and/or subcutaneous fat tissue. Etiopathogenesis of dermatofibroma is still unclear and trauma or damage to the superficial dermis is the most common suspected mechanism. Nowadays, through cytogenetic studies and clinical progression of some variants of DF (such as relapsing and metastasis), it is accepted to be a neoplastic lesion [1].

Clinically, dermatofibroma is usually characterized by solitary, pink-brownish papular lesion involving lower extremities and rarely seen in multiple (>15), congenital, familial, eruptive and giant forms. Multiple DF can be
seen in congenital clustered form or in acquired form which could be associated with autoimmune diseases, immunosuppression, different malignancies and drug usage [1,2,3,4,5,6,7].

To our knowledge our case is the first report of concomitantly seen benign and malignant fibrohistiocytic tumours in the literature.

Case Report

A 49- year-old male admitted to our outpatient clinic with reddish to brown, mildly painful lesions on his back, bilaterally hips and lower extremity evolved slowly in two years time (Figures 1a and b). Dermatological examination revealed one brownish nodular lesion on his right hip, two hyperpigmented papulonodular lesions ranging in size from 1 to 2 cm on the left hip and a pink papular lesion on the superior of scar tissue of a previous surgery on left subscapular area. Dermatoscopic examination of the nodular lesion on his right hip revealed white structureless area, white lines reticular over entire the lesion and light brown large clods distributed unevenly between the holes of white lines reticular while the lesion localized on the trunk revealed pink structureless area, white lines reticular and dotted vessels (Figures 2a and b). We performed two punch biopsies from these lesions with the piliminary diagnoses of dermatofibroma, dermatofibrosarcoma protuberans and cutaneous metastasis. Histopathological examination revealed DF in both lesions. Microscopically, tumor was composed of fibroblast like spindle cells, histiocytes and blood vessels in varying proportions. More cellular areas exhibited a storiform pattern of interwoven, fascicled spindle cells. Tumors were typically poorly demarcated. No cytologic atypia and mitotic activity were present (Figures 3 a, b and c). He doesn’t have an autoimmune disease, drug use or immunosuppression history. On his history we learned that, he had three surgeries for soft tissue tumours in 2012 from his left leg and left lung. Excisional biopsy of the lesion on the left leg was consistent with low grade fibromyxoid sarcoma (Figure 4a and b). Following the operation scanning imaging for a probable metastasis, showed two nodular lesions with a diameter of 4x4, 5x4 cm in the thorax computerized tomography. Histopathological examination of the left lung lobectomy material was consistent with solitary fibrous tumour/intraparanchimal hemangiopericytoma (Figure 5a and b). He is routinely under follow-up by Oncology Department and follow up imaging procedures with 6 months intervals showed no further metastasis.

Discussion

Dermatofibroma is one of the most common mesenchymal tumours which is also known as
benign fibrous histiocytoma and fibroma sim-
plex [1]. The most common clinical presenta-
tion of a dermatofibroma is a solitary hyper-
pigmented papulonodular lesion involving
lower extremities. In addition to this usual pre-
sentation, several DFs (<5) or multiple DFs
(>15) can be observed less commonly. Multiple
DFs can be classified into two groups: conge-
nital clustered and acquired forms. Acquired
form has been reported in association with
pregnancy, several systemic diseases (atopic
dermatitis), immunosuppression (HIV infection,
immunosuppressant drug usage), autoimmune
diseases (lupus erythematosus, myastania
gravis, Hashimoto thyroiditis) and malignancies
(leukemia and myelodysplastic syndrome) [1,2,
3,4,5,6,7,8]. In consequence of related con-
comitant entities with DF, immune mecha-
nism are suspected in the etiopathogenesis.
Low grade fibromyxoid sarcoma is a rare, cyto-
logically bland malignant neoplasm with alter-
nating fibrous and myxoid stroma with

Figures 2a and b. a. Light brown large clods distributed unevenly between the holes of white lines retic-
ular while the lesion localized on the trunk revealed pink structureless area, white lines reticular and
dotted vessels b. White structureless area, white lines reticular over entire the lesion

Figures 3a, b, and c. a. Microscopic appearance of
dermatofibrom on right hip at low power. H.E.
x40. b. At high power, microscopically tumor
was composed of fibroblast like spindle cells, his-
tiocytes and blood vesels. H.E.x200. c. Negative
immunreactivity of tumor cells and positive im-
munreactivity of endothelial cells for CD 34. Im-
munostaining for CD 34. x100

Figures 4a and b. a: Histopathological examination
of the left lung lobectomy material was consistent
with solitary fibrous tumour/intraparanchimal
hemangiopericytoma. H.E.x40. b. Microscopic
appearance of tumor at high power view. Cytolo-
gically banal spindle cells that are arranged hap-
hazardly in a densely collagenous matrix. The thin
parallel strands of collagen set this lesion apart.
H.E.x200
low-grade/low malignant potential [9]. Differential diagnosis of LGFMS includes lesions showing spindle cell proliferations with myxoid pattern with or without fibrous component such as myxomas, neurofibroma, fibromatosis, malignant peripheral sheath tumour, and fibrous histiocytoma [9]. Solitary fibrous tumors (SFTs) of lung is another rare and benign, primary soft tissue tumors with mesenchymal origin that arise from the submesothelial tissue [10]. This tumour can also occur in other sites including the lung, liver, orbit, nasal passages, skin, thyroid, and gastrointestinal tract. The finding of positive immunoreactivity for CD34 and bcl-2 and negative immunoreactivity for cytoplasmic keratin can confirm the presence of SFTs [10]. Although DF is known as a benign lesion, rarely local recurrence and metastasis can occur. In our patient several DFs have appeared slowly in two years time concomitantly with other fibrocytic tumours within a wide range of oncogenic potential. To our knowledge an association as seen in our case has not been reported in the literature before. We also present this case because of rare occurrence of multiple benign and malignant fibrohistiocytic tumours concomitantly.

References