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

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Abstract

Observation: Dermatofibroma is a benign fibrohistiocytic tumour of skin which is also known as benign fibrous histiocytoma. It is usually seen in adults in lower extremities and as a solitary, pink to brownish papulonodular lesion that may vary in number and size. Multiple dermatofibromas are rarely seen and have been reported as congenital clustered dermatofibromas or acquired associated with autoimmune diseases, immunsuppression and different malignancies in the literature.

A 49 year-old male patient with multiple reddish to brown, rigorous lesions on his back and hips with a history of two years. He had three surgeries for soft tissue tumours on his left leg and left lung two years ago which were consistent with low grade fibromyxoid sarcoma and solitary fibrous tumour/ hemagiopericytoma histologically. Dermatoscopic examination of papulonodular lesions were consistent with dermatofibroma. In regard to his past medical history, punch biopsies were performed to both lesions. Histopathological examinations of the lesions were both consistent with dermatofibroma. Multiple dermatofibroma and sarcoma association has not been reported in the literature before. We also present this case because of rare occurence of multiple benign and malignant fibrohistiocytics tumours concominantly.

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, immunsuppression, different malignancies and drug usage [**1,2,3,4,5,6,7**].

To our knowledge our case is the first report of concominantly seen benign and malignant

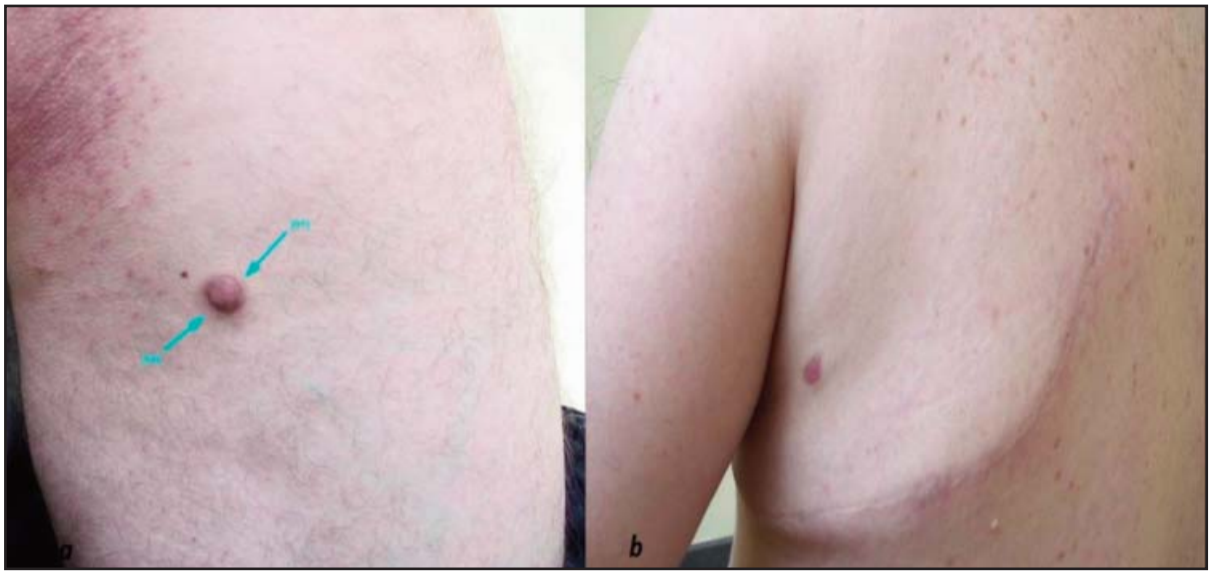
fibrohisticytic tumours in the literature.

Case Report

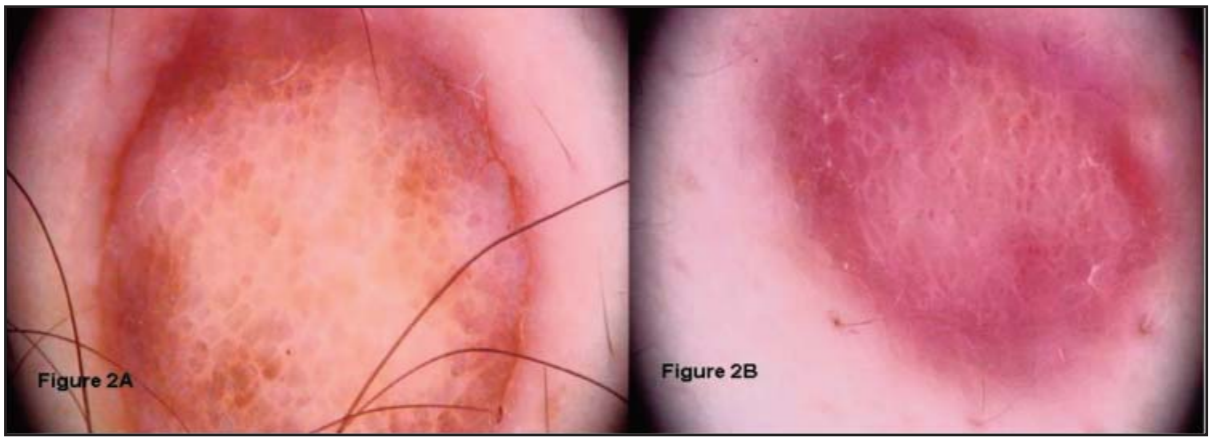
A A 49- year-old male admitted to our outpatient clinic with reddish to brown, mildly painful lesions on his back, bilaterally hips and lower extremity evolved slowly in two years time (**Figures 1a and b**).Dermatological examination revealed one brownish nodular lesion on his right hip, two hyperpigmented 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. Microscopycally, tumor was composed of fibroblast like spindle cells, histiocytes and blood veesels 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 immunsuppression 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

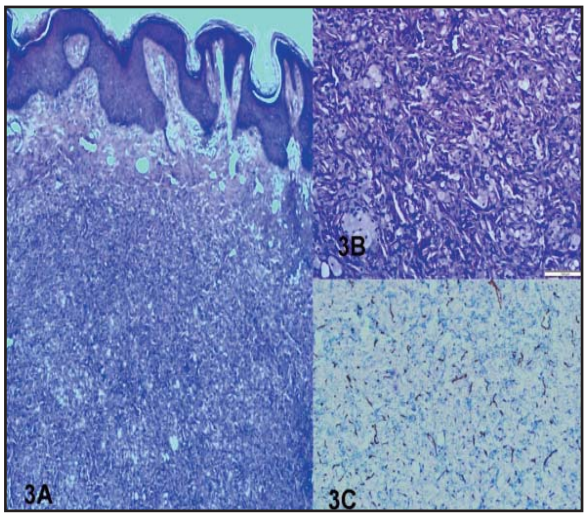


Figures 1a and b. **a**. Reddish to brown, mildly painful nodular lesion lower extremity **b**. Reddish to brown, mildly painful nodular lesion on his back

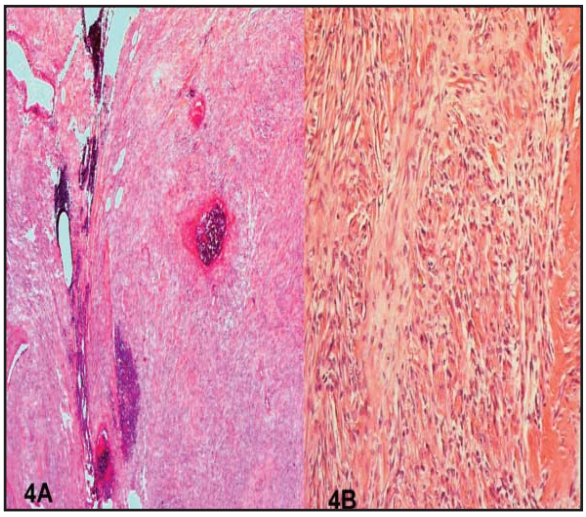


Figures 2a and b. a. Light brown large clods distributed unevenly between the holes of white lines reticular while the lesion localized on the trunk revealed pink structureless area, white lines reticular and dotted vessels **b**. White structureless area, white lines reticular over entire the lesion

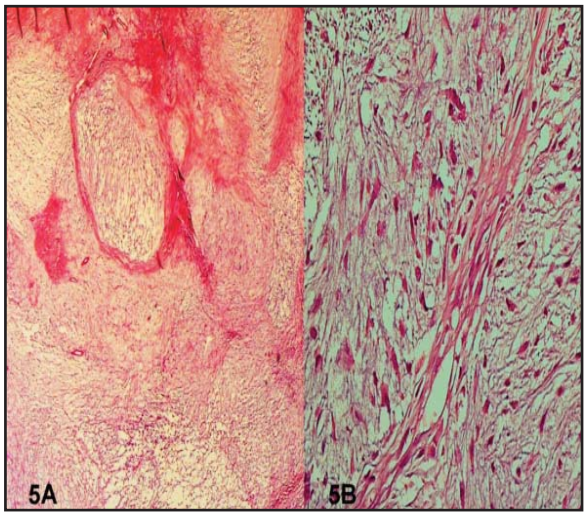
benign fibrous histiocytoma and fibroma simplex [1]. The most common clinical presentation of a dermatofibroma is a solitary hyper pigmented papulonodular lesion involving lower extremities. In addition to this usual presentation, several DFs (<5) or multiple DFs (>15) can be observed less commonly. Multiple DFs can be classified into two groups: congenital clustered and acquired forms. Acquired form has been repeorted in association with pregnanacy, several systemic diseases (atopic dermatitis), immunsuppression (HIV infection, immunsuppressant drug usage), autoimmune diseases (lupus erythematosus, myastania gravis, Hashimato tyroiditis) and malignancies (leukemia and myelodysplastic syndrome) [1,2,3,4,5,6,7,8]. In consequence of related concominant entities with DF, immune mechanism are suspected in the etiopathogenesis. Low grade fibromyxoid sarcoma is a rare, cytologically bland malignant neoplasm with alternating fibrous and myxoid stroma with



Figures 3a, b, and c. a. Microscopic appearance of dermatofibrom on right hip at low power. H.E. x40. **b**. At high power, microscopycally tumor was composed of fibroblast like spindle cells, histiocytes and blood veesels. H.E.x200. **c**. Negative immunreactivity of tumor cells and positive immunreactivity of endothelial cells for CD 34. Immunostaining 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. Cytologically banal spindle cells that are arranged haphazardly in a densly collagenous matrix. The thin paralel strands of collagen set this lesion apart. H.E.x200



Figures 5a and b. a. Microscopic appearance of low grade fibromyxoid sarkom of the left leg at low power. Extremely hypocellular myxoid noduler areas and hypersellular, interwoven spindle cell areas are seen. H.E. x40. **b**. At high power view, mild to moderately cytologic atypia may be seen. H.E.x400

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 immunreactivity for CD34 and bcl-2 and negative immunreactivity for cytoplasmic keratin can confirm the presence of SFTs [**10**]. Although DF is a known as a benign lesion, rarely local recurrence and metastasis can occur. In our patient several DFs have appeared slowly in two years time concominantly with other fibrocytic tumours within a wide range of oncogenic potential. To our knowledge an ass ociation as seen in our case has not been reported before in the literature. Hardy JD has reported case series of eight patient in 1987 who have different fibroblastic proliferations [**11**]. Our case supports this report with his different types of fibroblastic tumoral lesions. Multiple dermatofibroma and sarcoma association has not been reported in the literature before. We also present this case because of rare occurence of multiple benign and malignant fibrohistiocytics tumours concominantly.

References

1. Calonje E. Soft-Tissue Tumours and Tumour-like Conditions. Rook's textbook of Dermatology'de. Ed.Burns T, Breathnach S, Cox N, Griffiths C. Wiley-Blackwell, United Kingdom. Eighth edition. 2010; 56: 56.16–56.18.
2. Kimura Y, Kaneko T, Akasaka E, et al. Multiple eruptive dermatofibromas associated with Hashimoto's thyroiditis and myasthenia gravis. Eur J Dermatol 2010; 20: 538-539. PMID: 20406729
3. Yagami A, Akamatsu H, Suzuki K, et al. Multiple dermatofibromas in a patient with atopic dermatitis. Dermatology 2004; 208: 351-353.PMID: 15178922
4. Kanitakis J, Carbonnel E, Delmonte S, Livrozet JM, Faure M, Claudy A. Multiple eruptive dermatofibromas in a patient with HIV infection: case report and literature review. J Cutan Pathol 2000; 27: 54-56. PMID: 10660133
5. Zaccaria E, Rebora A, Rongioletti F. Multiple eruptive dermatofibromas and immunosuppression: report of two cases and review of the literature. Int J Dermatol 2008; 47: 723-727. PMID: 18613883
6. Caldarola G, Bisceglia M, Pellicano R. Multiple eruptive plaque-like dermatofibromas during anti-TNFα treatment. Int J Dermatol 2013; 52: 638-641. PMID: 22804251
7. Santos-Juanes J, Coto-Segura P, Mallo S, et al. Multiple eruptive dermatofibromas in a patient receiving efalizumab. Dermatology 2008; 216: 363. PMID: 18292655
8. Bhattacharjee P, Umar SA, Fatteh SM. Multiple eruptive dermatofibromas occurring in a patient with myelodysplastic syndrome. Acta Derm Venereol 2005; 85: 270-271. PMID: 16040422
9. Indap S, Dasgupta M, Chakrabarti N, Agarwal A. Low grade fibromyxoid sarcoma (Evans tumour) of the arm. Indian J Plast Surg 2014; 47: 259-262. PMID: 25190926
10. Zhu Y, Du K, Ye X, Song D, Long D. Solitary fibrous tumors of pleura and lung: report of twelve cases. J Thorac Dis 2013; 5: 310-313. PMID: 23825765
11. Hardy JD. The ubiquitous fibroblast. Multiple oncogenic potentials with illustrative cases. Ann Surg 1987; 205: 445-455. PMID: 3034175