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Multiple dermatofibromas in a female with systemic lupus erythramatosus on immunosuppressive medications. Case report and a brief literature review

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Abstract

Background: Multiple dermatofibromas (“DFs”) are defined by the presence of 15 lesions in the same patient or the development of five to eight DFs over the period of 4 months. Fifty-six percent of multiple DFs are associated with other diseases. The most common associated disease is systemic lupus erythematosus (“SLE”) followed by immunodeficiency virus (“HIV”) infection. *Main observation:* We report a case of a 25-year-old Saudi Arab female with SLE on immunosuppressive drugs with multiple DFs. *Conclusion:* The most common association with multiple DFs is SLE followed by HIV. Most of the patients with SLE were on immune suppressive medications. Dermatologists, rheumatologists, surgeons and internists should note that patients with SLE who are on immune suppressive medications are at risk of developing multiple DFs.

Keywords: Dermatofibromas; Systemic lupus erythramatosus; HIV

1. Case presentation

A twenty five year old Saudi Arab female with systemic lupus erythematosus (SLE) having an antinuclear antibody 1:320 homogenous pattern, double stranded DNA antibody recorded >1000 IU/ml, and anti-Smith antibodies equal to 174 units. Anti-Ro/SSA antibodies and anti-La/SSB antibodies were negative. The disease was controlled with prednisone 10 mg daily, tacrolimus 0.5 mg every 12 h, and hydroxychloroquine 200 mg daily. The patient visited the dermatology clinic complaining of a new onset of multiple asymptomatic skin growths over a period of six months. On skin examination, six rounded brown nodules were noted over the superior shoulder (Fig. 1), medial and lateral arm, back and posterior thigh. Histopathological examination of one of the lesions showed epidermal hyperplasia, dermal proliferation of fibroblasts, and histiocytes with intervening thick collagen bundles consistent with dermatofibroma (Figs. 2 and 3).

2. Discussion

Dermatofibroma also known as fibrous histiocytoma is a dermal tumor formed by proliferation of fusiform cells in the dermis. The cells are a variable combination of fibroblasts, collagen, blood vessels and histiocytes

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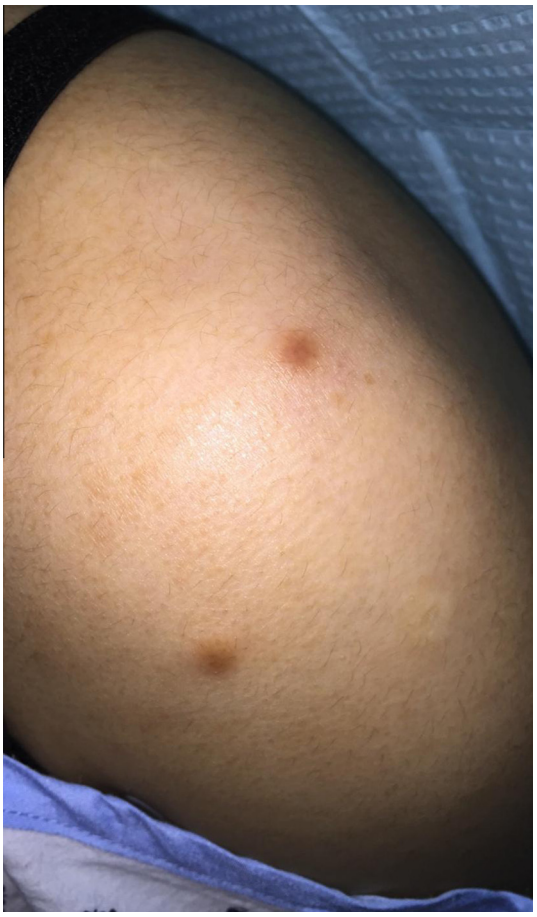

Figure 1. Two rounded brown nodules on the shoulder.

(Weedon, 2002). Clinically, the lesions are usually tan brown rounded firm papules ranging from 3 mm to 2 cm. They can also be pink-red (32.8%), skin colored (23%), or, in rare circumstances, blue in color (2.45%). The most common location is the lower extremities of young females (Han et al., 2011). When lateral pressure is applied to the skin surrounding DFs, the lesions become depressed from the level of the skin (dimple sign or Fitzpatrick's sign) (Naversen et al., 1993). The dermoscopic patterns of DFs are variable with the most common being a central white scar-like area with a delicate pigment network at the periphery (Zaballos et al., 2008).

The histological variants include fibrocollagenous DF (40.1%) which is formed mainly from collagen and fibroblast-like cells in an irregular or whorled pattern, histiocytic (13.1%) with angulated epithelioid cells, cellular (11.5%) with larger numerous fibroblasts that can occasionally infiltrate fat, aneurysmal (7.4%) with prominent pseudovascularity, angiomatous (6.5%) with the presence of small branching vessels in a collagenous stroma, sclerotic (6.5%) with hyalinized dense collagen, monster (4.9%) with bizarre atypical giant cells, palisading (1.6%) with Verocay-like bodies resembling a Schwannoma, and keloidal characterized by the presence of thick collagen and multinucleated giant cells with hemorrhage and hemosiderin deposits (Han et al., 2011; Rapini, 2005). Typically, DFs express positivity for actor XIIIa, HMGA1, and HMGA2 (high mobility group AT-hook 1 and 2. CD34 is negative in DFs (Li et al., 2004).

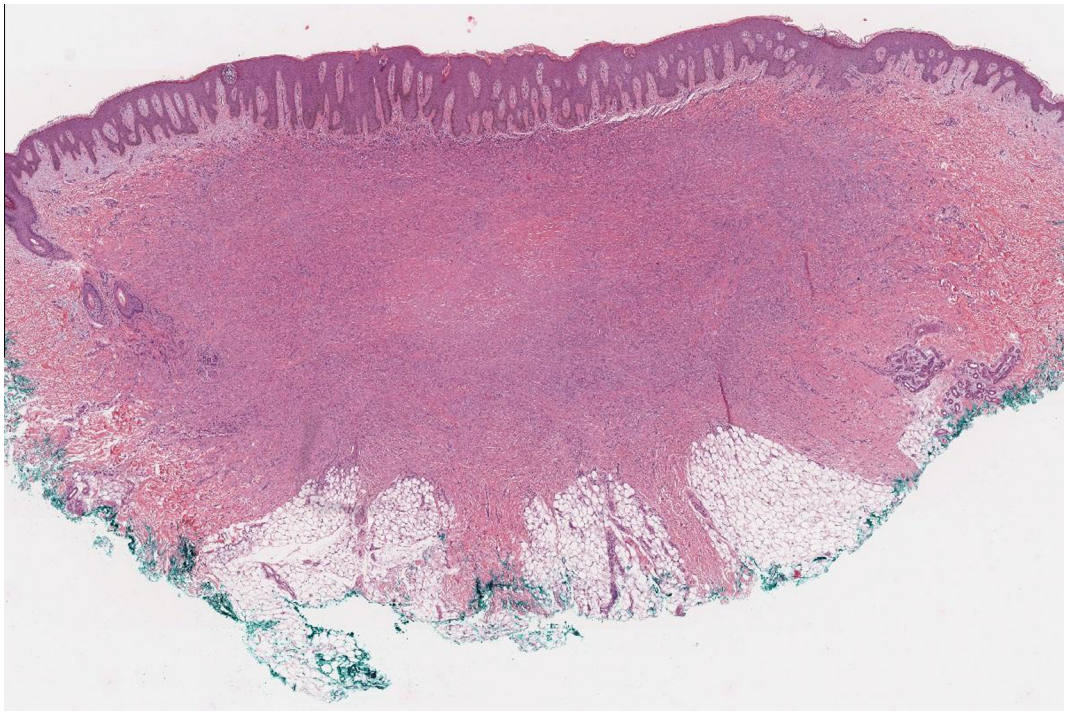

Figure 2. Dermatofibroma with overlying hyperplastic epithelium (hematoxylin and eosin, 33× magnification).

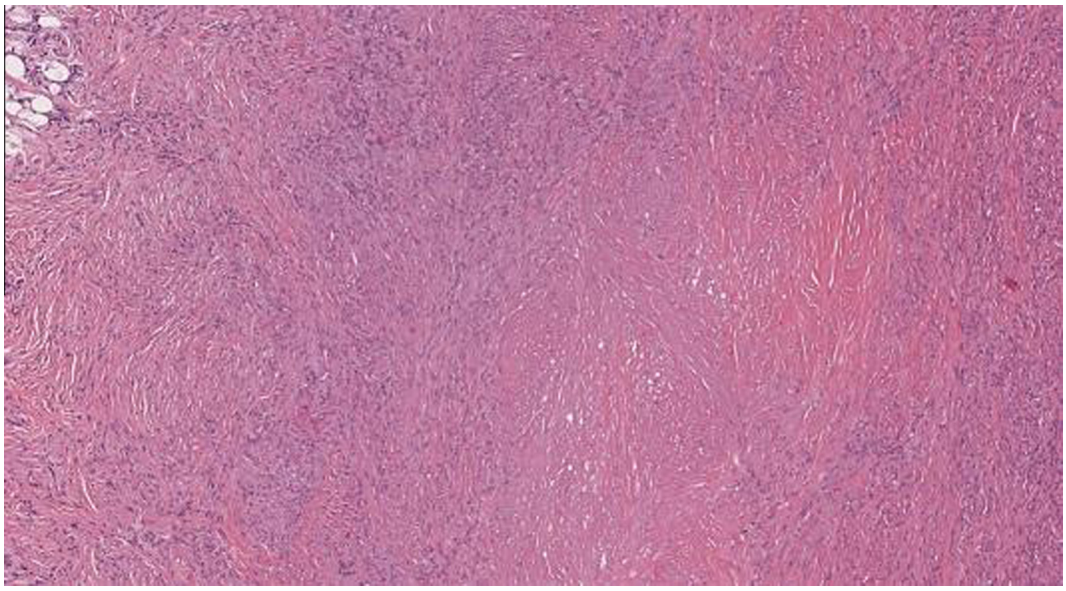

Figure 3. Spindle cell proliferation in storiform pattern (hematoxylin and eosin, 197× magnification).

The definition of multiple DFs remains controversial. Fifty percent of cases reported were more than 15 lesions. However, less than this number with the same associations was also reported. Five to eight DFs developing over the period of 4 months is also included in this definition. The term eruptive DFs is used when the lesions appear over a 4-month period.

Fifty-six percent of multiple DFs are associated with disease. The commonest diseases include SLE (46%), followed by immunodeficiency virus (HIV) infection (32%). Most of patients with SLE were on immune suppressive medications. Other diseases associated with multiple DFs include myasthenia gravis, pemphigus vulgaris, Sjogren's syndrome, hydronephrosis, sarcoidosis and Sézary syndrome (Niiyama et al., 2002a,b, 2001; Massone et al., 2002; Tsunemi et al., 1995; Lu et al., 1997; Kanitakis et al., 2000; Gualandri et al., 1999). Familial and congenital multiple DFs are very rare but have been reported (Pinto-Almeida et al., 2013; Marque et al., 2013). Immunodeficiency, autoimmune diseases or the intake of immunosuppressive drugs should be ruled out in patients with multiple DFs.

The lesions represent reactive proliferation of fibroblasts usually secondary to minor trauma or an insect bite (McKee et al., 2005). Basic fibroblast growth factor and platelet-derived growth factor levels were elevated in the serum of patients with SLE with multiple DFs, which stimulates fibroblast proliferations, and thus explains the increased risk of dermatofibroma development (Yamamoto et al., 1995). The role of immune suppressive medications in the development of multiple DFs is still unknown but could be explained by the fact that DFs develop through an abortive immune reactive process which can be triggered by drugs that down-regulate T cells (Nestle et al., 1995).

Because the lesions are benign, treatment is not necessary. Some lesions do undergo spontaneous slow resolution. Nevertheless, treatment options include surgical excision, pulse dye laser, or carbon dioxide laser (Alonso-Castro et al., 2012; Wang and Lee, 2006; Wang et al., 2013).

3. Conclusion

The most common association with multiple DFs is found to be SLE followed by HIV infection. Most of patients with SLE who developed multiple DFs were on immune suppressive medications. Dermatologists, rheumatologists, surgeons and internists should be aware of this association and the clinical appearance of the lesions in order to reassure patients and explain the possible clinical course.

Conflict of interest

None.

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