CASE REPORT

Multiple eruptive myxoid dermatofibromas

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ABSTRACT

Multiple eruptive dermatofibromas are a rare presentation of dermatofibroma and have been associated with altered immunity. A rare case of multiple eruptive myxoid dermatofibromas (MEMDFs), characterized by marked stromal mucin deposition, is reported herein; additionally, an in-depth discussion on the implication of altered immunity is presented. Because MEMDFs can be an initial manifestation of systemic lupus erythematosus, it is necessary for dermatologists to perform a skin biopsy for pathological diagnosis and a comprehensive survey for autoimmunity, infectious diseases, especially human immunodeficiency virus infection, hematologic diseases, and malignancies, or other immunodeficiency conditions when the patient is diagnosed with MEMDFs.

Introduction

Dermatofibromas are common benign fibrohistiocytic lesions that are most often solitary and occur on the legs of middle-aged women.1 Multiple eruptive dermatofibromas (MEDFs) are a rare presentation of dermatofibroma and have been associated with altered immunity, such as in the case of human immunodeficiency virus (HIV) infection and autoimmune diseases treated with immunosuppressive therapy.2 Myxoid dermatofibroma, characterized by marked stromal mucin deposition, is an extremely rare variant of dermatofibroma, found only in 0.4% of all dermatofibromas.3 Only one single case of multiple eruptive myxoid dermatofibromas (MEMDFs) in a healthy woman was published in the literature.4 Herein, we report another rare case of MEMDFs as an initial manifestation of systemic lupus erythematosus (SLE).

Case report

A 51-year-old woman presented with 20 flesh-colored to pinkish asymptomatic papules that developed within 2 months (Figure 1). The lesions were located on her arms and legs, and varied in size from 3 mm to 8 mm. Differential diagnoses included eruptive xanthomas, dermatofibromas, neurofibromas, and lymphoproliferative diseases. A skin biopsy was performed, which showed a dermal nodule of fibrohistiocytes and thin collagen fibers accompanied by lymphocytes in a myxoid stroma (Figure 2). Abundant mucin was demonstrated in the lesion using Alcian blue stain, with and without hyaluronidase digestion. An immunohistochemical study showed that the spindle tumor cells were positive for factor XIIIa, CD68, CD44, and CD163; weakly positive for NK1/C3; and negative for S-laminin, caldesmon, and collagen type IV. Neurogenic and smooth muscle tumors were excluded. The immunophenotype was compatible with dermatofibroma. Because the patient had multiple lesions with profound mucin deposits, MEMDFs were diagnosed. Laboratory investigations indicated leucopenia (leukocyte count 3000/μL), speckled antinuclear antibody pattern (1:160 titer), hypocomplementemia (C3 at 77.50 mg/dL and C4 at 4.63 mg/dL), positive antidualle-strand DNA antibody, positive anti-Smith antibody (anti-Sm), positive anti-Ro/SSA antibody, and equivocal anti-La/SSA antibody. Other laboratory findings, including rapid plasma reagin and HIV tests, were negative or within normal limits. MEMDF as an initial manifestation of SLE was diagnosed. The patient was treated with prednisolone and hydroxychloroquine. The number of skin lesions continued to increase in the 1st month, but started resolving 3 months later without recurrence.

Discussion

A survey of literature for studies on dermatofibromas published between 1962 and 2014 reveals that only 62 patients were reported...
with MEDFs. Among the reported cases, the predominance of middle-aged female patients [39/62 (62.9%); mean age 38.3 years] is worth noting. Most patients had associated diseases [50/62 (80.7%)], and among them nearly half had autoimmune diseases [24/50 (48.0%) with SLE predominance [18/24 (75.0%)]. Sjögren syndrome, dermatomyositis, Graves disease, myasthenia gravis, rheumatoid arthritis, and pemphigus vulgaris were also reported as the autoimmune diseases. Most [22/24 (91.7%)] MEDFs patients associated with an autoimmune disease were women, and all [18/18 (100%)] MEDFs patients with SLE were adult women. Other associated diseases were HIV infection, hematologic diseases (e.g., chronic myelogenous leukemia, myelodysplastic syndrome, and multiple immunoglobulin A myelomas), metabolic diseases (e.g., diabetes, hypertriglyceridemia), and inflammatory diseases (atopic dermatitis, psoriasis, and sarcoidosis), among others; furthermore, it is was also reported to be associated with kidney transplantation, and pregnancy. Ten of the 11 MEDFs patients with HIV infection were male and the only patient having both HIV infection and SLE was a female.

Survey findings indicate that MEDFs occur mostly before or concurrently with the diagnosis of other associated diseases or altered immune status, and exacerbate or improve after treatment with immunosuppressants. Previous reports suggested that both the diseases and their treatments, such as treatment with corticosteroids, cyclophosphamide, azathioprine, cyclosporine A, methotrexate, hydroxychloroquine, monoclonal antibodies, and other chemotherapeutic agents, are related to the occurrence of MEDFs. Immunosuppression, autoimmunity, and immune restitution may all predispose to the development of MEDFs.

The pathogenesis of dermatofibroma remains unclear. It is controversial whether it is a true neoplasm or a reactive hyperplasia. An abortive immunoreactive process of infiltrating T cells responding to an unknown antigen presented by dermal dendritic cells was postulated. Increased dermal mucin deposition is a
feature of lupus erythematosus and other degenerative-inflammatory processes. Because of the rarity of MEMDFs cases, it is unclear whether MEMDFs is a specific reactive or neoplastic process or just a coincidental finding with mucin deposition, and this needs further investigations.

Dermatologists must be aware that MEDFs may imply altered immunity and MEMDFs might be an initial manifestation of SLE. A skin biopsy for pathological diagnosis and a comprehensive survey for autoimmunity, infectious diseases, especially HIV infection, hematologic diseases and malignancies, or other immunodeficiency conditions are necessary when MEMDFs are diagnosed.

References


