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Case Report

Multiple Eruptive Dermatofibromas in a Patient with Primary Sjögren's Syndrome

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ABSTRACT

Multiple eruptive dermatofibromas (MEDF) are rare tumors and thought to be associated with the disturbances in the immune system. In our 40-year-old case, 5 nodules have developed in a 4-month period. The patient was diagnosed by a dermatologist to have MEDF, and referred to rheumatology outpatient clinic because of the symptoms such as dry mouth, and polyarthralgia. After clinical and laboratory evaluation, the diagnosis of primary Sjögren's syndrome (SS) was made. According to the best of our knowledge, this case is the first reported association between MEDF and primary SS. Therefore, when the diagnosis of MEDF is made, SS should also been included in the comprehensive evaluation of associated diseases.

Keywords: Sjögren's syndrome, dermatofibroma, dermal tumor, autoimmune diseases, fibrous histiocytoma

INTRODUCTION

Dermatofibroma (DF) is a common benign tumor among tumors of skin origin [1]. Although this tumor often occurs as a single lesion, more commonly on the upper extremities of young adults, the occurrence of multiple eruptive DF (MEDF) is rare. MEDFs have been associated with autoimmune diseases (AID), immunosuppressive therapy, HIV (human immunodeficiency virus) infection, hematological malignancies, pregnancy, and other conditions. Although the exact etiopathogenesis of MEDFs is not known, it is thought to be associated with the altered state of the immune system [2]. Sjögren's syndrome (SS) is a chronic inflammatory AID that often presents with dry eyes and mouth due to functional involvement of the exocrine glands [3]. SS consists of two forms: primary SS, characterized by sicca symptoms unrelated to AIDs; and secondary SS, which is characterized by symptoms associated with other AIDs, particularly systemic lupus erythematosus (SLE) [3]. We present a female patient who developed MEDF associated with primary SS.

CASE

A 40-year-old female patient applied to the dermatology clinic due to the presence of multiple asymptomatic papules and nodules developed in a four-month period. The patient has no known disease and no history of drug use. Clinical examination revealed five well-circumscribed, hyperpigmented, firm, brown and round nodules with a diameter of 3-10 mm in the left-right upper arm, right leg and abdomen (figure 1). In the skin biopsy performed, there are uniform spindle cells arranged in long fascicles parallel to the epidermis. Immunohistochemical examination showed the expression of factor XIIIa and vimentin. When evaluated together with the anamnesis, physical examination and skin biopsy it was found to be compatible with DF by the dermatologist. The patient diagnosed with DF was referred to the rheumatology clinic for the evaluation of AIDs. In the rheumatological evaluation, the patient had dry mouth, dry eyes and especially metacarpophalangeal and proximal interphalangeal joint pain of both hands for one year. No signs

of arthritis were found in the physical examination. In the Schirmer's test, the paper was wetted <5 mm in both eyes. In laboratory examination, antinuclear antibodies test was found to be positive (1:100 homogeneous). Extracted nuclear antigens were all negative. The patient was undergone a minor salivary gland biopsy which was reported as having focus score ≥1 and diagnosed as SS.

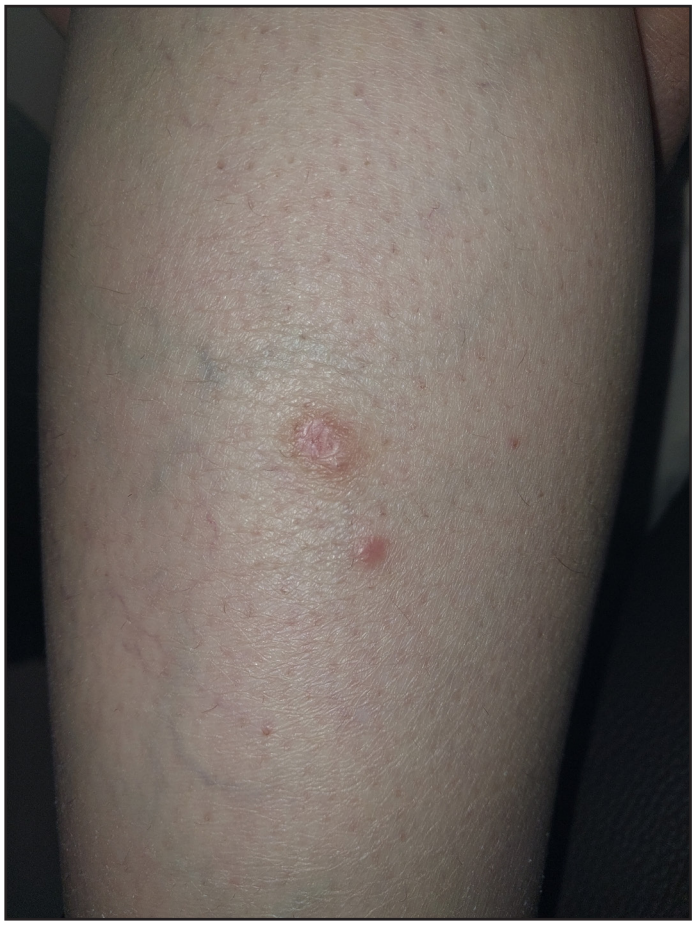

Figure 1. Hyperpigmented, brown and round dermal nodules are shown on the left-right upper arm, right leg and abdomen.

DISCUSSION

Dermatofibroma, also called benign fibrous histiocytoma, is one of the most common cutaneous soft-tissue lesions. It is known as a benign dermal proliferation of fibroblasts [4]. Although the pathogenesis is unknown, they can sometimes occur as a result of trauma or infection. DF occur most often in adults and are most commonly located on the lower extremities [4]. Diagnosis is usually based upon clinical appearance and history. If the lesion is longstanding, characteristically it should have no history of rapid change. Excision for histopathologic examination is indicated for any changing or bleeding lesion or when the lesion is suspicious for malignancy [5]. Usually, no treatment is required unless the lesion is symptomatic [6]. MEDF was first described in 1970 and is generally defined as the occurrence of at least 5 DFs within a four-month period. MEDF is significantly less common in patients with DF, and a significant proportion of MEDF is thought to be associated with a systemic condition [7]. The most frequently associated conditions associated with MEDF have been reported in the literature as AID (SLE, dermatomyositis), immunosuppressive drugs, HIV infection and hematological malignancies [8]. Of the MEDF considering its association with immune-mediated diseases or immune suppression states, it is thought to be strongly associated with immune system mechanisms [8]. Nestle et al. [9] states that DF arises as a result of an immune system in which dermal dendritic cells are prominent. According to this hypothesis, the development of MEDFs in immune-deficient conditions may be facilitated by inhibition of down-regulatory T cells; alternatively, MEDFs may develop as an exaggerated response to a putative pathogen that cannot be cleared by the suppressed immune system [9]. SLE is the most common autoimmune disorder associated with MEDFs. There are numerous cases of MEDF reported in patients with SLE or in patients with SLE and SS, but no cases of MEDF associated with primary SS have been reported [10]. Although we made the diagnosis of SS after the skin findings of DF in our case, the symptoms of SS had been begun months ago. Thus, MEDF might be considered as associated with SS. This case is one of the rare cases in the literature that emphasizes the potential relationship between primary SS and MEDF.

CONCLUSION

All these data show that the theoretical relationship between the incidence of MEDF and immunosuppression is not accidental. While any attempt so far to provide a solid interpretation has been purely speculative, dermatologists and rheumatologists should be aware that MEDF may be a sign of a disturbance in immune system, and all patients should evaluated for the presence of an associated systemic disease.

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DT. Data interpretation: SY. Drafting manuscript: MNK, SY. Revising manuscript: all authors.

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