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Case Letter

Association of Shulman fasciitis, generalized morphea, and thyroid cancer

Siham Boularbah, Sara Elloudi, Sabrina Oujdi, Zakia Douhi, Meryem Soughi, Hanane Baybay, Fatima Zahra Mernissi

Department of Dermatology, University Hospital Hassan II, Fes, Morocco

Corresponding author: Siham Boularbah, MD, E-mail: sihamboularbah1902@gmail.com

Sir,

Shulman fasciitis (SF) is a rare scleroderma syndrome that may occur at any age. The onset is often brutal. A triggering factor, such as intense physical effort or trauma, is noted in 30% to 46% of cases [1]. It may be isolated or associated with certain autoimmune diseases, lymphoid and myeloid malignancy, or solid cancer [2]. Herein, we report a case associating Shulman fasciitis with generalized morphea and a thyroid tumor.

A 65-year-old patient presented with isolated edema on both ankles persistent for six months occurring after a physical effort, then extending to the four limbs and the trunk. The evolution was marked two months later by a progressive hardening of the skin without Raynaud's phenomenon. A dermatological examination revealed diffuse cutaneous sclerosis on the integument (Fig. 1a) with the limitation of the opening of the mouth (Fig. 1b), with no pulpitis or scleroderma pattern on dermoscopy. An osteoarticular examination revealed irreducible flexion of the knees and hands. A biological assessment revealed hyperleukocytosis with hypereosinophilia at 5000 elements/mm^3. An immunological assessment was negative, in particular AAN, anti-SC70, and anti-centromere. MRI of the limbs found thickening with contrast of the muscle fascia. A cutaneomuscular and deep fascia biopsy at the level of the right anterior tibialis revealed a thinned and straight epidermis. The dermoepidermal interface was slightly modified. The dermis was sclerous and punctuated with numerous polymorphonuclear eosinophils with an essentially perivascular arrangement. The muscle adipose and fascia fragments examined were infiltrated by lymphocyte and plasma cells. At the end of these clinico-biological and radiological elements, the diagnosis of Shulman fasciitis associated with morphea was established.

A paraneoplastic assessment was in favor of neoplasia of the thyroid. The patient was started on bolus corticosteroid therapy, then switched to oral corticosteroid therapy. The skin became increasingly supple. Methotrexate was planned after managing the neoplastic problem, yet the patient was lost from follow-up.

The physiopathology of SF remains poorly known, which is sometimes in a paraneoplastic context, sometimes in an autoimmune context [3], initially indurated, symmetrical, and edematous associated with hypereosinophilia, then progressive sclerosis of the skin and diffuse subcutaneous tissues. The hands and face are generally spared [1,3]. There is no Raynaud's phenomenon or autoimmune markers. MRI of the muscles and fascia describes increased acute fascia signal intensity in 80% of cases. A muscle and fascia biopsy shows fascia thickening with inflammatory infiltrates composed mainly of CD8+ T cells and eosinophils. It is necessary to underline the importance of the normality of the epidermis and dermis as it alone makes it possible to distinguish fasciitis of Shulman from morphea [4]. Indeed, around one-third of cases of EF are associated synchronously or metachronously with morphea [5]. In our patient, the anatomo-clinical, biological, and radiological signs compatible with Shulman fasciitis, yet the epidermo-dermal

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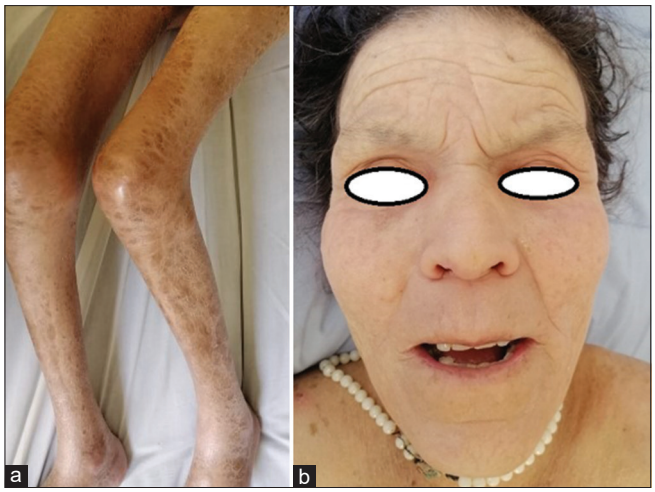

Figure 1: (a) Severe cutaneous sclerosis with bilateral blessum. (b) Limitation of mouth opening to two fingers.

anomalies and the extensive fibrosis strongly resemble a generalized morphea. This association is explained by some authors as a form of overlap and a transition between these two highly distinct entities.

The other frequently described associations with EF are hemopathies [2]. We found thyroid neoplasia which had been rarely described in the literature [5].

Bolus corticosteroid therapy, then oral corticosteroids remain the main treatment in the edematous phase and may be combined with an antifibrotic agent, such as methotrexate [1,3]. Other therapies have occasionally been used successfully [3]: ciclosporin, infliximab, and hydroxychloroquine. Our patient responded partially to corticosteroid therapy because she consulted in the edematous phase with hypereosinophilia found by biological assessment, which is seen mainly during this phase.

Consent

The examination of the patient was conducted according to the principles of the Declaration of Helsinki.

The authors certify that they have obtained all appropriate patient consent forms, in which the patients gave their consent for images and other clinical information to be included in the journal. The patients understand that their names and initials will not be published and due effort will be made to conceal their identity, but that anonymity cannot be guaranteed.

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