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Letter to the Editor

Cutis laxa (elastolysis) in a patient with Sjögren's syndrome

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Dear Editor,

Cutis laxa (CL), or elastolysis, is a rare inherited or acquired connective tissue disorder where the skin becomes inelastic and hangs loosely in folds. Sjögren's syndrome (SS) is a chronic inflammatory disease characterized by diminished lacrimal and salivary gland function. SS may be a primary disease and may accompany other rheumatologic diseases. To our knowledge, the simultaneous occurrence or the accompaniment of these two diseases has never been reported before.

A 56-year-old female patient applied to our outpatient unit with dry mouth, dry eye, and bilateral edema on the hands. In the physical examination, there was no arthritis, fever, tenderness, or erythema. In the laboratory evaluation, the anti-nuclear anticore (ANA) was positive, with a titer of 1/320, and extractable nuclear anticore (ENA SsA) was also positive. The Schirmer test score on both sides was 5 millimeters/5 minutes, and the minor salivary gland biopsy score was grade 3 with the Chisholm ve Mason system. The patient was diagnosed with SS, but to explain the swelling on the hand, a biopsy was done, and the microscopic examination revealed hyperkeratosis in the epidermis, histiocytes, and a few multinuclear giant cells in the dermis (Figure 1). Verhoeff staining demonstrated the complete loss of elastic fibers in the middle and upper dermis (Figure 2). The histologic features were compatible with elastolysis (cutis laxa) and giant cell reaction. Hydroxychloroquine and steroid combination therapy was offered to the patient, and after 6 weeks, the range of motion of the hand joints got better, and now, the patient can eat by herself and can also wear her dresses.

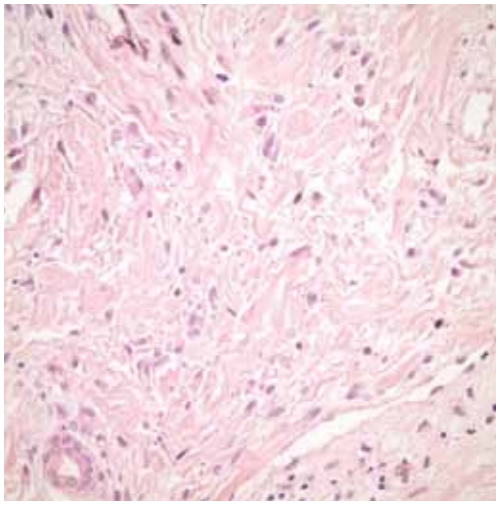

Figure 1. Histiocytes and giant cells with 1-3 nuclei in between collagen fibers. H&E, 40X

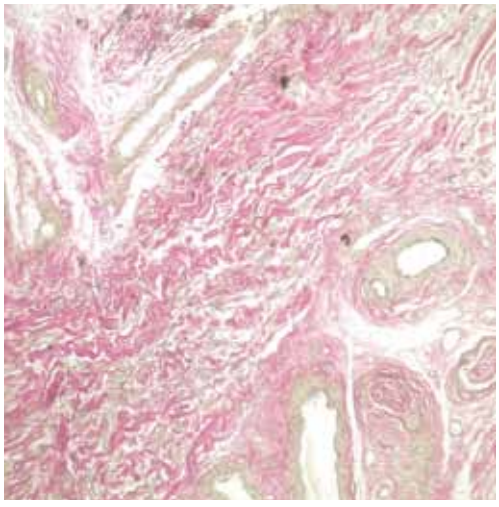

Figure 2. Loss of elastic fibers replaced by histiocytes. 20X

Kittridge A et al. (1) stated that nearly half of patients with SS have dermatologic manifestations, such as dry skin, purpura, or urticaria-like lesions, which are mostly underemphasized and confer an increased risk for the development of multisystem vasculitis and non-Hodgkin lymphoma.

Fox RI et al. (2) have reported that the frequency of skin vasculitis in patients with SS ranges from 9%-30% and that the other rare dermatologic manifestations are subcutaneous amyloid, erythema multiforme-like, erythema perstans-like, and erythema nodosum-like lesions.

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In conclusion, the coexistence of CL and SS in this patient may be by chance, but it must not be forgotten that connective tissue disorders may have different dermatological presentations, and some may cause life-threatening complications, such as lymphoma and vasculitis; therefore, a better dermatologic evaluation is necessary.

Informed consent: Written informed consent was obtained from patients who participated in this case.

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References

1. Kittridge A, Routhouska SB, Korman NJ. Dermatologic manifestations of Sjögren syndrome. J Cutan Med Surg 2011; 15: 8-14.
2. Fox RI, Liu AY. Sjögren's syndrome in dermatology. Clin Dermatol 2006; 24: 393-413. [CrossRef]