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Postinflammatory Cutis Laxa: A Case Report

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Abstract Cutis Laxa (CL) is a rare disease in which the skin loses its elasticity and hangs in large folds. It is an inherited or acquired connective tissue disorder. We report the case of a 29 year-old woman followed up since 4 years for a dermatomyositis treated with glucocorticosteroids and methotrexate. She was hospitalized in February 2012 for fever, arthralgia, pelvic and shoulder muscle weakness with myalgia, malar rash, thrombocytopenia, leucopenia and lymphocytopenia Immunological tests showed Antinuclear Antibodies (ANA) (+) to 1/640, AC anti-DNA (+) and AC anti-SSA (+). Histology of the salivary glands showed grade III lymphocytic sialadenitis. The systemic lupus erythematosus and secondary Sjögren's syndrome were diagnosed. The patient was treated with glucocorticostroids, methotrexate, nivaquine and bissolvon. One year later, the patient presented a skin aging that began in hands which has expanded rapidly in the face. The skin biopsy confirmed the diagnosis of a "Cutis Laxa". The esthetic treatment is proposed.

Keywords: *Acquired cutis laxa, systemic disease, systemic lupus erythematosus, dermatomyositis, Sjögren's syndrome*

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1. Introduction

Cutis laxa (CL) is also known as dermatolysis, dermatomegaly, chalazoderma, pachydermatocele, dermatochalasia, and elastolysis [1].

CL represents a heterogeneous group of connective tissue disorders that may be acquired or inherited. It can have a generalized or local form.

It is often preceded by cutaneous inflammatory eruption (ie, urticaria, eczema, erythema multiforme). Frequently, there is an important internal organ involvement: the gastrointestinal, pulmonary and cardiovascular systems can be concerned.

Redundant skin is often most noticeable on the neck, hands, and groin, but can also be seen on the face, creating a premature aging appearance.

Clinically, there is redundant skin with a wrinkled and loose appearance. Histologically, degenerative changes in the dermal elastic fibers were observed. Light microscopy reveals few or absent elastic fibers in the dermis [2].

Of the few reports on this rare disorder, authors have speculated about an immune-mediated destruction of elastic fibers, and monoclonal gammapathies, such as multiple myeloma or heavy chain deposition disease have a recognized association with CL.

We report through one observation an exceptional association: Acquired CL and mixed connective tissue disease.

2. Case Report

A 29-year-old Tunisian woman having the history of dermatomyositis diagnosed 4 years ago was treated with glucorticosteroids and methotrexate. Her disease was stable with a clinical and biological remission until 2012. In February 2012, she was admitted in the internal medicine department for fever, arthralgia, proximal muscle weakness, diffuse myalgia, eyelid erythema, a "butterfly" rash.

Physical examination revealed skin rash, metacarpophallangeal joints arthritis and motor deficit of the shoulder and pelvic muscles. Ophthalmologic examination confirmed eye dryness: Shirmer's test, performed without anesthesia (under than 5 mm in 5 minutes) and Tear Break up Time (less than 10 seconds).

Biological tests confirmed the lymphopenia and thrombopenia and showed increased inflammatory biological parameters (CRP, ESR).

Immunological tests showed a high rate (1/640) of ANA and positive anti-DNA, anti-SSB and anti-SSA antibodies. The biopsy of the accessory salivary glands showed grade III lymphocytic sialadenitis according to Chisholm classification.

The systemic lupus erythematosus and secondary Sjögren's syndrome associated to dermatomyositis confirm the diagnosis of mixed connective tissue disease.

Thus, she was treated with glucocorticosteroids, methotrexate, nivaquine and bissolvon. The evolution was marked by the regression of disease activity signs.

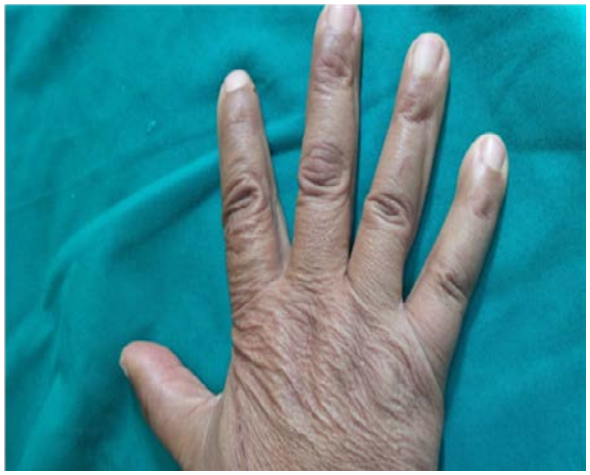

Figure 1. Loose and pendulous skin of all fingers

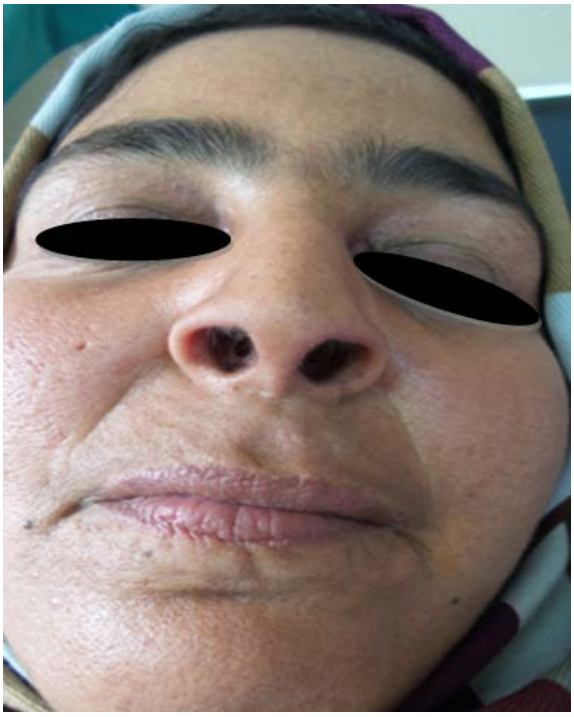

Figure 2. Loose, pendulous skin of the face; the patient appeared to be much older than her real age

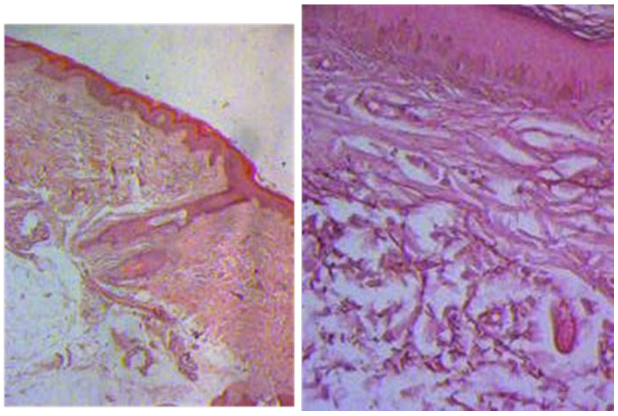

Figure 3. Pathological examination of the biopsy material showing fragmentation and destruction of elastic fibers (HE x 100; Ocrein x 100)

One year after, the patient developed skin loosening, deep wrinkles aging that began in hands which quickly spread to the face. Physical examination revealed loose, pendulous skin of all fingers, hands and the face (Figure 1, Figure 2). She appeared to be much older than her real age (Figure 2). Pathological examination of the biopsy material showed the fragmentation and destruction of elastic fibers (Figure 3).

The diagnosis of "Cutis Laxa" was chosen because of the clinical and histological data. Physical and morphological examination (chest X-ray; echocardiography; gastroscopy …) didn't show any visceral affection. Protein electrophoresis ruled out gammapathy particularly multiple myeloma. There was no history of drug ingestion. Botox injection was proposed.

3. Discussion

CL is a rare connective tissue disorder characterized by wrinkled skin with loss of elasticity. It can be hereditary or acquired, generalized or localized [3,4]. Our patient is suffering from acquired and localized CL.

Its pathophysiology is not well understood. Hypotheses include abnormal copper metabolism or copper deficiency, decreased serum elastase inhibitor (α 1-antitrypsin), low lysyl oxidase activity, and immune-mediated mechanisms. In one case, anti-elastin antibodies were detected [5,6].

Acquired CL has been described in association with drug ingestion, inflammatory skin disorders, and neoplasms. Reviewing the literature, we noted that diseases that have caused acquired cutis laxa include chronic urticaria, erythema multiforme, erythema perstans, vesicular eruptions, eczema, and Sweet's syndrome [7,8,9]. Drugs such as penicillin, penicillamine, and isoniazid have also been implicated [10,11].

Reports of Acquired CL associated with multiple myeloma, plasmacytoid lymphoma, systemic lupus erythematosus, nephritic syndrome with and without sarcoidosis, necrobiosis lipoidica, syphilis, and Lyme disease can be found in the literature [12-17].

The basic histological lesion is localized in the connective tissue related to electively elastic fibers. However, other lesions in the dermis can be observed. In our case, pathological examination showed the fragmentation and destruction of elastic fibers (Figure 3).

The lesions of elastic fibers may also affect other organs than the skin and cause vascular or visceral lesions. Our patient showed no extra-cutaneous affection.

Acquired CL is more common in adults. It progresses in a cephalocaudal direction [18,19,20]. In our case, symptoms began in hands which quickly spread to the face (Figure 1- Figure 2).

The disease most commonly found in association with CL is monoclonal gammapathy particularly multiple myeloma. Since 1976, a dozen cases of this association was reported [21-25].

Lewis FM et al. and Vegnil S et al. reported, respectively in 1993 and 2003, a patient with acquired CL in association with sarcoidosis [26,27].

In 2002, Rongioletti F et al. described the first case of the acral localization of the acquired form of CL associated with severe rheumatoid arthritis [28].

In our knowledge, our case represents the first reporting the association mixed connective tissue disease /CL.

In all the reported sightings, skin aging is preceded by erythematous scaly lesions. This was not the case of our patient.

Acquired CL is not transmitted genetically. However, one of the areas of research conducted by Siefring ML et al is whether certain individuals may have a genetic predisposition to develop CL after certain exposures [29].

Nowadays, there is no yet aneffective therapy for CL. Its treatment based on plastic surgery such as cosmetic surgical procedure (rhytidectomy) [30,31].

4. Conclusion

CL is a rare connective tissue disease, the first obvious symptom is a skin slackening. The late appearance of CL should suggest the acquired form which makes the prognosis worse.

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