Case Report

Stiff skin syndrome rock-hard skin a report case of two sisters

Abstract
Stiff skin (SS) syndrome is a scleroderma-like disease characterized by the absence of involvement of internal organs and muscles in infancy or early childhood, with hardness of the skin, localized hypertrichosis, and limitation of joint mobility. Involvement of the hips and thighs is prominent, and localized hyperpigmentation may also attend the affected areas. Non-cutaneous findings may be accompanied by “tiptoe-fingertip gait” walking, scoliosis, restrictive breathing difficulty and growth retardation. Familial manifestation shows a genetic transmission of it. We present two sister who initially were diagnosed clinical and histopathological characteristics of SSS.

Keywords: scleroderma, scleroderma-likedisorder, stiff skin syndrome

Case report
A 15-year-old female patient was referred in dermatology for difficulty in walking, difficulty opening and closing her arms and legs, and excessive stiffness of the abdomen, hips and thighs. From the age of 8 complaints began to become clear. Over the time, skin stiffness and painful muscle spasms started to trigger by movement and coldness. Her family history revealed that there was kinship between the mother and father, and similar complaints were also found in the sister. Stiffness of the skin and subcutis was felt on palpation. Lesions were starting from the inguinal region and the thighs, which had collapsed and puffed hard palpated from the dense skin surface, had a cobblestone appearance. (Figure 1 A,B,C) Although 23-year-old sister had similar physical examination findings but sclerosis was less. ANA, ENA profile for scleroderma, EMG findings for muscle involvement were normal. Histopathological examination of both sisters showed; coarsening, disordered and homogenization of collagen fibers in papillary and reticular dermis. Decreased skin supplements and loss of fat pad in the dermis. (Figure 2 A,B) With clinical, pathological and laboratory findings the patients were diagnosed as “STIFF SKIN SYNDROME”. In order to reduce sclerosis, UVA-1 treatment was started at the external center. Sclerosis decreased significantly after 15 sessions and patients are still under treatment (Figure 3 A,B,C).

Discussion
Stiff Skin first is described by Esterly and Mc Kusik in 1971 as uncommon connective tissue disease similar to scleroderma. Generally internal organ and muscular involvement, vascular...
hyperactivity and immune system abnormalities are absent. There are some criteria that support this syndrome such as: hereditary transmission, stone-hard thickening of the skin predominantly in facia buttocks and thighs, restricted joint movements, variable hypertrichosis and absence of mucopolysaccharidosis in urine test. This syndrome is most commonly confused with scleroderma or morphea profunda but despite excessive sclerosis progression is very slow, laboratory findings are unremarkable, nail capillary microscopy is normal, raynaud’s phenomenon is not accompanied and muscle involvement is not observed. Skin induration of hard skin syndrome is not progressive, hardens and remains constant. Most of the sss patients histolopathologies showed a non-inflammatory, thickened and hyalinized fascia accompanied by dermal fibrosis.

**Conclusion**

Stiff skin syndrome is a rare syndrome with unknown etiology and pathogenesis, characterized by an early and insidious stony hard skin, especially in the thigh, hip and abdominal and joint mobility restriction. There is no effective treatment for this disease yet. Exercises and rehabilitative treatment are important to improve quality of life of the patient. Our patient was given UVA-1 treatment a set of exercises and is being followed-up periodically. Exercise therapy and phototherapy combination provided a significant improvement in our patients condition.

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**Conflict of interest**

The authors declare that there is no conflict of interest.

**References**