<https://doi.org/10.47660/CBR.2020.17219>

PANSCLEROTIC MORPHEA MIMICKING DIFFUSE SYSTEMIC SCLEROSIS: A CASE-REPORT

Rodrigo da Silveira Vasconcelos^{1*}, Carla Baleeiro Rodrigues Silva¹, Aline Capellato Dias Baccaro¹, Osvaldo Luiz Camata Junior¹, Giovanni Homero Jacome Verdugo¹, Samuel Katsuyuki Shinjo¹, Diogo Souza Domiciano¹

1.Universidade de São Paulo, São Paulo (SP), Brazil.

*Corresponding author: rodrigo.silveira.vasconcelos@gmail.com

BACKGROUND

Localized scleroderma is a condition that must be differentiated from systemic sclerosis (SS) in clinical practice. It is more common in children than adults. Linear morphea is the most common form of the disease. Pansclerotic (deep) morphea is a less common subtype and can be misdiagnosed as diffuse SS. We herein report a case of an adulthood presentation of a pan sclerotic scleroderma mimicking SS.

CASE REPORT

A 55-year-old woman presented with 8-month history of dermatosis involving the trunk and extremities. The disease began as sudden onset of peripheral oedema affecting both hands and feet and progressed with extensive and symmetrical cutaneous thickening that had also compromised the trunk, mainly abdomen. The patient reported a 20% loss of body weight since the onset of symptoms and denied other systemic complaints and a family history of a similar illness, as well as cutaneous trauma. The skin induration affected dorsal surface of the hands and spared fingers. The skin surface became smooth and shiny. On physical examination, it was noteworthy the extensive cutaneous thickening that restricted range of motion of the knees and ankles. Some areas, especially abdominal region, had indurated skin plaques that alternated with areas of normal skin. There was no evidence of Raynaud phenomenon, telangiectasia or digital pitting/ulcers. Laboratory evaluation revealed: PCR 17 mg/L and VHS 16 mm, antinuclear antibody (ANA) negative, no eosinophilia, with no other changes. Nail fold capillaroscopy was normal and no evidence of internal organ affection or fasciitis, as well as normal age-appropriate neoplastic screening, were found. The patient was diagnosed with Pansclerotic morphea and treated with daily prednisone 20 mg and subcutaneous methotrexate 15 mg/week was introduced.

CONCLUSION

Pansclerotic morphea is an extremely rare form of localized scleroderma in adults and is a diffuse scleroderma mimicker. Differential diagnosis of diffuse SS is challenging. Careful physical examination of the skin should be done to identify the erythematous indurated confluent plaques. Moreover, absence of Raynaud phenomenon, scleroderma pattern on nail fold capillaroscopy and serious internal organ affection are clues to the correct diagnosis. Being assertive when excluding SS has an important prognostic and therapeutic impact.