Scleredema associated with Sjögren's syndrome*

João Alves[1]
Tiago Judas[1]
Tiago Ferreira[1]
Diogo Matos[1]
Elvira Bártolo[1]

DOI: http://dx.doi.org/10.1590/abd1806-4841.20153779

Abstract: Scleredema adultorum of Buschke is a rare disorder characterized by diffuse swelling and non-pitting induration of the skin usually involving the face, neck, arms and upper trunk. It has been associated with previous infectious diseases, diabetes, paraproteinemia and, more rarely, malignant neoplasms or autoimmune disorders. We report the case of a 30-year-old man who presented with a 2-year history of scleredema. Further investigation led to the diagnosis of primary Sjögren's syndrome. The association between scleredema and autoimmune disorders has been rarely seen. To our knowledge, there are no other reports describing the association between primary Sjögren's syndrome and scleredema adultorum of Buschke.
Keywords: Autoimmune diseases; Mucinoses; Scleredema adultorum

INTRODUCTION

Scleredema adultorum (SA) of Buschke is a rare disorder characterized by diffuse swelling and non-pitting induration of the skin usually involving the face, neck, arms and upper trunk.[1] Traditionally, it is classified into three types: type 1 is usually preceded by a febrile infectious episode; type 2 is associated with paraproteinemia and type 3 is associated with diabetes mellitus.[2] Very rare cases have been associated with malignant neoplasms or autoimmune disorders.[3,4]

CASE REPORT

A 30-year-old man has presented with a 2-year history of progressive, diffuse, painless and non-pruritic cutaneous induration of the arms and shoulders with some functional limitation of the upper limbs. He had no medical history and no other complaints in the first consultation. On physical examination, we observed a discrete erythema and a non-pitting induration of the skin of arms, shoulders and superior dorsum (Figure 1). Differential diagnosis included scleredema, scleromyxedema and scleroderma. A cutaneous biopsy was performed (Figure 2) and showed thickened collagen fibers separated by clear spaces filled with mucin (Figures 3 and 4). The diagnosis of SA of Buschke was assumed based on clinical and histopathological features. In subsequent consultations, we observed that the skin induration had slightly increased. Moreover, the patient experienced symmetric migratory polyarthralgias mainly involving the knees and shoulders. He had also been experiencing dry eye sensation for 5 months. He denied xerostomia, fever, fatigue, weight loss or others symptoms. Laboratory analysis revealed discrete thrombocytopenia (119 000) with otherwise normal hemoglobin, leukocytes and erythrocyte sedimentation rates and C-reactive protein values. Fasting glycemia and total serum protein were normal, as well as protein electrophoresis, urinalysis, and renal, hepatic and thyroid function. A strong positivity to anti-SSA antibodies and a positive ANA (1/1280) were immunologically detected. Anti-DNAds, anti-SSB, rheumatoid factor, ACE, An-

Received on 17.06.2014
Approved by the Advisory Board and accepted for publication on 10.07.2014
* Study conducted at the Garcia de Orta Hospital – Almada, Portugal.
Financial Support: None.
Conflict of Interest: None.

[1] Garcia de Orta Hospital – Almada, Portugal.

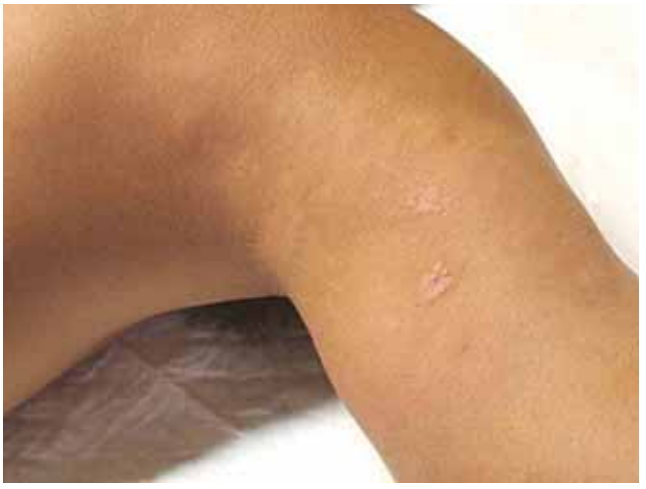

Figure 1: Clinical appearance of the right arm. Cutaneous induration was evident only on palpation

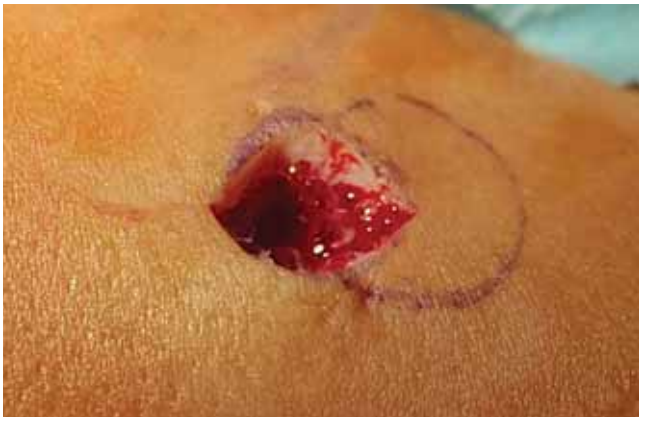

Figure 2: A jelly material was identified in the skin biopsy

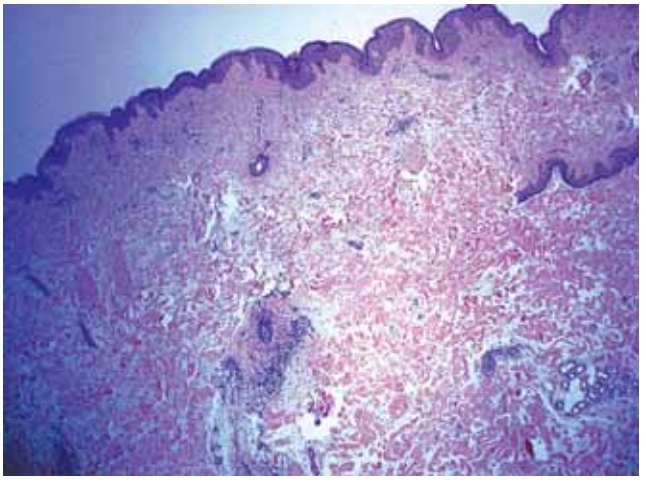

Figure 3: Thickened collagen fibers separated by clear spaces on the dermis (hematoxylin and eosin 40x)

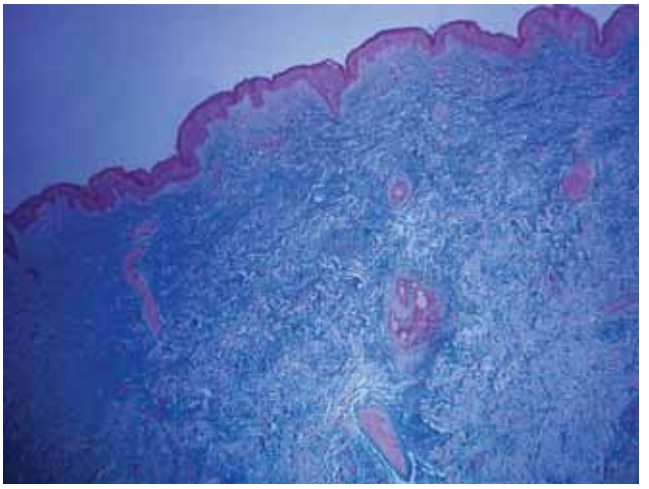

Figure 4: Thickened collagen fibers separated by mucin (alcian blue 40x)

tistreptolysin-O, anti-SM, ANCA and anti-SCL 70 were negative. Complement level was normal. Serologic tests to hepatitis B and C, HIV and syphilis were negative. Radiographs of the shoulders and knees were normal. A Schirmer's test revealed a moderate to severe impairment of tear function (<5 mm, normal range ≥ 15 mm). Scintigraphy of the salivary glands was compatible with Sjögren's syndrome. The diagnosis of primary Sjögren's syndrome was established according to the American-European Consensus Criteria.[5] The patient was treated with hydroxychloroquine 400 mg PO daily. One month after onset of treatment, we already observed improvement in pain symptomatology and stabilization of skin changes.

DISCUSSION

SA is a rare disorder of unknown etiology which was first described by Buschke in 1900.[1] It may resolve spontaneously within 2 years or have a progressive and persistent course with potential fatal outcome.[1,2] The association between autoimmune disorders and SA has been rarely seen. To our knowledge, only one report associates SA with Sjögren's syndrome, namely that of a patient with secondary Sjögren's syndrome (with concomitant rheumatoid arthritis).[4] Our patient could possibly be the first case of SA associated with primary Sjögren's syndrome. Therefore, in addition to diabetes and dysproteinemia, this case highlights the importance of investigating other associations, specifically autoimmune disorders, ensuring its proper management and treatment. ❑

REFERENCES

1. Lewerenz V, Ruzicka T. Scleredema adultorum associated with type 2 diabetes mellitus: a report of three cases. J Eur Acad Dermatol Venereol. 2007;21:560-1.
2. Beers WH, Ince A, Moore TL. Scleredema adultorum of Buschke: a case report and review of the literature. Semin Arthritis Rheum. 2006;35:355-9.
3. Manchanda Y, Das S, Sharma VK, Srivastava DN. Scleredema associated with carcinoma of the gall bladder. Br J Dermatol. 2005;152:1373-4.
4. Miyagawa S, Dohi K, Tsuruta S, Shirai T. Scleredema of Buschke associated with rheumatoid arthritis and Sjögren's syndrome. Br J Dermatol. 1989;121:517-20.
5. Vitali C, Bombardieri S, Jonsson R, Moutsopoulos HM, Alexander EL, Carsons SE, et al. Classification criteria for Sjögren's syndrome: a revised version of the European criteria proposed by the American-European Consensus Group. Ann Rheum Dis. 2002;61:554-8.

MAILING ADDRESS:
João Vítor Pina Alves
Av. Torrado da Silva
2801-951 Almada
Portugal
E-mail: alves.joaovitor@gmail.com

How to cite this article: Alves J, Judas T, Ferreira T, Matos D, Bártolo E. Scleredema associated with Sjögren's syndrome. An Bras Dermatol. 2015;90 (3 Suppl 1): S81-3.