Letters to the Editor

Scrotum and testicular calcinosis in juvenile dermatomyositis (JDM). A report of two cases

Sirs,

Juvenile dermatomyositis (JDM) is a systemic disease characterized by inflammation of the skeletal muscle and skin. The disease is initially marked by the presence of vasculitis and later by the development of calcinosis (1).

It is known that calcinosis is a common and debilitating complication of JDM. It is also a hallmark of the disease, occurring mainly in the pediatric population. The frequency of calcinosis in JDM is 10% to 70%, generally affecting subcutaneous tissues, muscles, fascial planes and tendons, usually found on elbows, knees, digits and extremities (2-4). Four subtypes have been described in literature: calcinosis circumscripta (superficial plaques or nodules in the skin or subcutaneous tissues), tumoural calcinosis or universalis (larger nodular deposits that may extend to deeper layers of tissue including the muscle), calcinosis along fascial planes of muscles and tendons and extensive hard calcium deposition over all body surface areas (5). Remarkably, the known risk factors for calcinosis include delayed treatment and severe disease (6).

We report two cases of JDM with scrotum and testicular calcinosis. One of them had partial regression after alendronate, and the other one was refractory to colchicine, diltiazem and alendronate, and improved with intravenous immunoglobulin (IVIG).

Case 1: A 6-year-old boy was diagnosed with JDM (7) with the presence of heliotrope rash and Gottron's papules, muscle weakness, increased AST, ALT and LDH serum levels, inflammatory infiltrate and perifascicular atrophy at muscle histopathology and characteristic electromyo-graphy changes. He had severe disease activity, cutaneous vasculitis and extensive calcinosis since he was 2 years old. He was treated with prednisone, methotrexate, cyclosporine, thalidomide, colchicine and diltiazem. Despite the therapy, he maintained a chronic illness course. Remarkably, during the follow-up, he developed a swollen and tender mass in the left testis. The scrotum and testicular magnetic resonance imaging (MRI, high-resolution T2-weighted fast spin-echo sequence) showed increased edema and calcifications in the subcutaneous tissue of the perineum area and on the left scrotum (Fig. 1A). At that moment, the disease was active: the Childhood Myositis Assessment Scale (CMAS) (8) was 42 (range of score: 0-52) and the Disease Activity Score (DAS) (9) was 6 (range of score: 0-20). He received alendronate (70 mg/week) for 6 months before the association of IVIG (2g/kg/month). The scrotum calcinosis was refractory to preliminary therapy and improved after 6 months of association with IVIG as observed in the scrotal Doppler ultrasound (US) with the 14-MHz sector scanner (Fig. 1B).

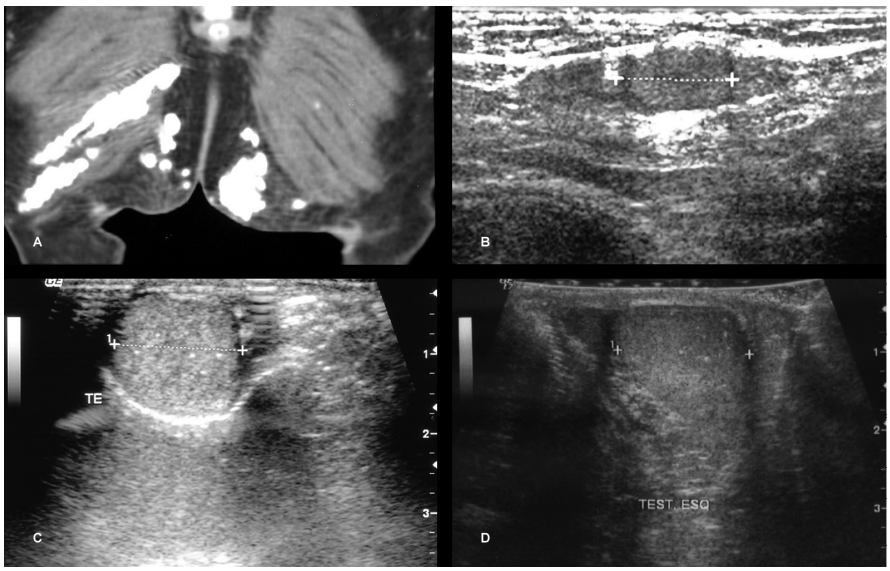

Fig. 1. (**A**): MRI high-resolution, T2-weighted fast spin-echo sequence shows increased edema and calcifications in the subcutaneous tissue of the perineum area and the left side scrotum.
(**B**): Doppler US image shows the absence of calcifications in the scrotum and testicle.
(**C**): Doppler US image shows small calcifications in both testicles in spite of the lack of shadows.
(**D**): Doppler US image shows the decrease of calcification sizes in both testicles.

Case 2: A 13-year-old boy was diagnosed with JDM (7) with the presence of heliotrope rash and Gottron's papules, muscle weakness, increased CK, LDH and AST serum levels and inflammatory infiltrate and muscular perifascicular atrophy in the histopathology when he was 10 years old. The JDM course was monocyclic and remission was acquired simply with prednisone therapy during a period of two years. His current JDM scores were normal: CMAS was 52 (range of score: 0-52) and DAS was 0 (range of score: 0-20). Interestingly, tender scrotum was reported and the testicular and scrotum Doppler US 14-MHz sector scanner found small calcifications in both testicles in spite of the lack of shadows (Fig. 1C). He received alendronate (70 mg/week) and had partial regression of testicular calcinosis in a period of 18 months (Fig. 1D).

Calcinosis is a devastating complication of JDM that can cause significant debility and severe pain, join contracture, skin ulcers and muscle atrophy (10).

As far as we know, these are the first inflammatory scrotum and testicular lesion case reports in JDM, suggesting scrotum and testicular calcinosis. These patients had tender scrotum and the diagnoses were established with testicular MRI or US. Please note that testicular involvement had been rarely described in patients with JDM (11) or adults with dermatomyositis (DM) (12, 13). Jalleh *et al.* (11) reported testicular necrotizing vasculitis in a 7-year-old boy with JDM who had a swollen and tender scrotum. In addition, primary testicular cancer, like a testicular germ cell tumour, was only reported in two adults with DM (12, 13).

Calcinosis occurs despite aggressive therapy and could be more related to disease severity than to therapeutic approach, as observed in the first case (10). In addition, none of the therapies for calcinosis have been consistently effective. Diltiazem, bisphosphonates and IVIG may be beneficial, as evidenced in our patients (10).

The US is usually the initial imaging modality for evaluating the calcification of testicles, epididymis and scrotum. MRI could be used as a problem-solving tool when US findings are inconclusive and cannot differentiate between solid mass, inflammatory or vascular abnormality (14). Epididymitis and testicular vasculitis were not observed in either case. The increased edema on a T2 weighted image on MRI, in the first case, and the lack of a shadow on the ultrasound, in the second case, usually do not indicate calcinosis. However, this finding could suggest another undefined inflammatory lesion in the testicle.

Therefore, the presence of tender scrotum in JDM patients could be associated with scrotum or testicular calcinosis, and the scrotum/testicular MRI or US should be performed. Furthermore, there is still uncertainty regarding fertility issues in the medium or long-term follow up of patients with JDM. Future studies should be conducted to evaluate the testicular function by serial semen analyses in the JDM population.

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Competing interests: none declared.

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