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Acute sarcoid myopathy: a case report and literature review

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Abstract. Sarcoidosis has a worldwide distribution and a large clinical spectrum, in which pulmonary involvement is the main manifestation (more than 90% of cases); nevertheless, extrathoracic symptoms can predominate in the clinical picture and may even be the first manifestation. Skeletal muscle involvement is one of them, usually chronic and silent with poor response to treatment with glucocorticoids. However, in some cases, it has an acute presentation. We present a case of a 61-year old man who was being evaluated for abrupt proximal lower limb weakness in whom a diagnosis of sarcoidosis was previously performed. *(Sarcoidosis Vasc Diffuse Lung Dis 2016; 33: 413-415)*

Key words: sarcoidosis, myopathy, granulomatous disease

Introduction

Sarcoidosis has a worldwide distribution, however racial or geographic influences difficult to establish its prevalence and incidence. Nevertheless, we can say that this entity is more common in Nordic and African-American populations (1). It shows a peak of incidence before 40 years of age (2) and a slight female predominance (3).

The main manifestation of sarcoidosis is pulmonary involvement, with a presence higher than 90% in most series (4); however, in some cases, the clinical picture may be dominated by symptoms of extrathoracic origin. We report a case of acute sarcoid myopathy, an infrequent reference in literature.

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Case presentation

A 61-year-old Caucasian man was admitted to the hospital because of proximal lower limb weakness for the last week. At first, he felt mild myalgia and difficulty climbing stairs. A few days later, he could not get out of chair without help and moderate asthenia was noted. Eight days after the onset of symptoms, the patient attends the Emergency Department. He had never developed fever, dyspnea, skin manifestations or localizing symptoms of underlying malignancy. He has been diagnosed with sarcoidosis 6 years earlier, when a biopsy of hyperpigmented maculopapular skin lesions located on the face and upper back revealed noncaseating granulomas. At that moment, a chest X-ray examination showed bilateral hilar lymphadenopathy with mild pulmonary infiltrates and minimal splenomegaly was described in an abdominal ultrasound. His medical history also included hypertension, which was well controlled on medication, and he had been cholecystectomized. His usual treatment was prednisone 7.5 mg daily, candesartan cilexetil 16 mg daily and calcifediol 12 mcg every 48 hours.

On the day of admission, physical examination revealed strength 4+/5 in right deltoid with mild atrophy, bilateral proximal weakness in psoas (3+/5) and otherwise was normal. Analytical testing showed elevated creatine phosphokinase (CPK) levels up to 834 mg/dl. Renal function, sodium, potassium, calcium, phosphate, magnesium, glucose, thyroid and parathyroid hormones, blood count, C-reactive protein (CRP), erythrocyte sedimentation rate (ESR), antinuclear antibodies and complement were normal; also serologies for *Brucella*, *Borrelia burgdorferi*, *Toxoplasma*, syphilis and Human Immunodeficiency Virus (HIV) were performed and their results were negatives. An electromyogram (EMG) revealed marked myopathic signs in upper and lower extremities without nerve involvement.

Subsequently, the patient underwent a muscle biopsy on the left quadriceps, revealing noncaseating epithelioid granulomas, dense interstitial CD4 and CD8 lymphocytic infiltrate, with predominance of the former, and degenerative changes in striated muscle (Figure 1).

The study concluded with a chest radiograph in which no changes were noted over the previous, an electrocardiogram that showed no conduction disturbances, an echocardiogram that revealed absence of pulmonary hypertension or pericardial involvement, and magnetic resonance (MRI) of lower limb that detected diffuse involvement of the posterior compartment muscles of both thighs. STIR sequence drew signs of fatty infiltration and an increased signal compatible with intramuscular and perifascial edema.

Finally, the patient showed rapid improvement after the increasement of steroid theraphy (three boluses of 250 mg of methylprednisolone followed by deflazacort 60 mg daily). Azathioprine was added later and the dose of glucocorticoids was gradually reduced.

Discussion

Sarcoidosis is a disease of unknown cause, characterized by the presence of non-caseating granulomatous inflammation in several organs, mainly involving the intrathoracic lymph nodes and the lungs. Silent skeletal muscle involvement in sarcoidosis is common (50-80% of cases) (5), but extremely rare as a clinical manifestation (less than 3%) (6).

Classically, sarcoid myopathy has been divided into three variants. The first and most common is the chronic form (6,7), similar in clinical to muscular dystrophies. It develops slowly, often evolves in years and is characterized by proximal muscle weakness of the lower limbs, with slight elevation of CPK and/or acute phase reactants. Sarcoid cardiac involvement is often associated with this pattern (6,8).

Another possibility is an acute presentation similar to a picture of myositis, accompanied by weakness, myalgias and elevated muscle enzymes, CRP and ESR. Sometimes acomppanied fever and arthritis, as well (6). It is the rarest variety (9). Finally, we can find a nodular variant with the presence of one or multiple painful lumps (6-8).

The gold standard diagnostic technique is the biopsy, revealing noncaseating granulomas in up to 80% of cases (6). Furthermore, the EMG can yield false negative if infiltrates and granuloma formation are not closed to excitable units (9). On the other hand, while MRI is not useful in the diagnosis of chronic and acute variants, it describes some char-

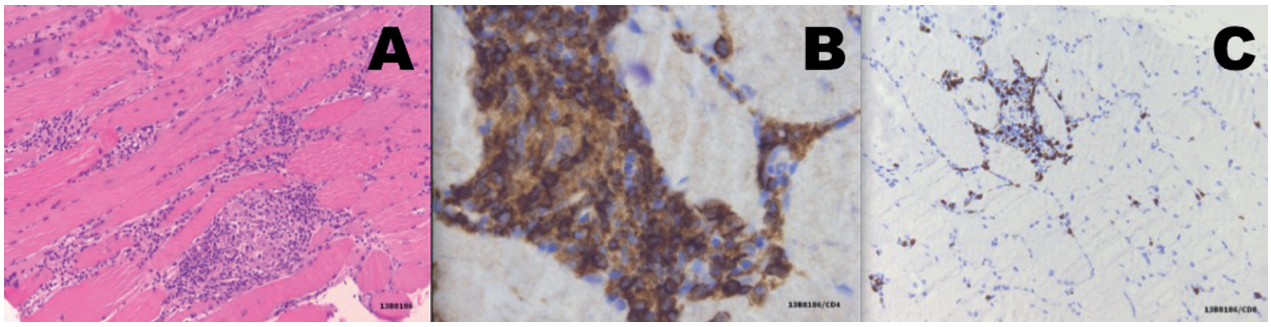

Fig. 1. A) Sarcoid granuloma (hematoxylin-eosin staining). B) Immnuhistochemical staining for CD4 lymphocytes. C) Immnuhistochemical staining for CD8 lymphocytes

acteristic signs of nodular variant, such as the "black star" pattern in the axial or the "three stripes" pattern in the coronal and sagittal axes (10). Anyway, the main role of this technique is as a locator of the most profitable area for the muscle biopsy.

Regarding treatment, the acute form responds well to glucocorticoids versus poor or irregular response of the other patterns (8). In those patients who have an inadequate response to or unable to taper their corticosteroids, other immunosuppressive drugs are used (6).

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