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A Rare Presentation of Muscle Involvement in Sarcoidosis: A Case Report

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Abstract: *Background*: Sarcoidosis can affect virtually any organ of the body, primarily the lungs, lymphatic system, skin or eyes, or a combination of these sites and it is characterized by the formation of non-caseating granulomas. Muscular involvement in sarcoidosis is usually asymptomatic or pauci-symptomatic, and its nodular form is exceptional. *Observation*: In this study, we report the case of a 36-year-old man who had been presenting with nodules in both calves for about two years which onset was progressive. These nodules were painless and deeply embedded in the gastrocnemius muscles. There were no functional complaints, and the general condition was preserved. The physical examination, especially at skin level was free of other clinical signs except the nodules. The PET-scan (positron emission tomography) revealed a few discretely hypermetabolic cervico-axillary lymph nodes and multiple bilateral mediastino-hilar adenopathies involving all lymph nodes, as well as multiple bilateral hypermetabolic pulmonary foci. Biopsy of accessory salivary glands (BASG) exhibited an epithelioid and gigantocellular granuloma without necrosis that confirm the diagnosis of systemic sarcoidosis. The search for another location, notably cardiac, ophthalmological, neurological, or renal, was negative. The respiratory function tests did not show any restrictive syndrome or abnormalities in the DLCO/VA ratio. It was therefore a systemic sarcoidosis with mediastino-pulmonary stage II, diffuse lymph node, splenic, and muscle involvement. *Conclusion*: This case shows the importance of BASG in the diagnosis of muscular sarcoidosis, in case of impossibility of performing a muscle biopsy.

Keywords: Muscular Nodules, PET Scan, Sarcoidosis, Dijon

1. Introduction

Sarcoidosis is a systemic inflammatory disease with a wide range of clinical manifestations, but its causes are quite unknown [1]. It can affect virtually any organ of the body, primarily the lungs, lymphatic system, skin or eyes, or a combination of these sites and it is characterized by the formation of non-caseating granulomas [1]. All around the world, its prevalence and incidence are variable. In Caucasians, the prevalence was 0.05% and the incidence was 8.1 [2]. Sarcoidosis is considered as a result of a dysregulated antigenic response to unknown environmental exposures in a genetically susceptible individual [1]. Mediastino-pulmonary involvement is widespread as it is found in 90% of patients. The most common extra-thoracic localizations are ophthalmic, cutaneous, articular, and hepatic (15 to 35% of cases) [3]. However, all organs can be affected. Muscular involvement is usually asymptomatic or pauci-symptomatic and its nodular form is exceptional [3]. This case report illustrates the presentation of a systemic sarcoidosis in a young patient with muscular nodules.

2. Observation

A 36-year-old man had been presenting with nodules in both calves for about two years which onset was progressive. These nodules were painless and deeply embedded in the gastrocnemius muscles. There were no functional complaints, and the general condition was preserved. The physical examination, especially at skin level was free of other clinical signs except the nodules. The patient had a medical and surgical history of a congenital aortic valve disease treated with a mechanical prosthesis, requiring long-term anti vitamin K treatment. The biological assessment did not show any inflammatory syndrome, nor any elevation of CPK. The immunological assessment showed nothing relevent. Serum angiotensin-converting enzyme (ACE) was elevated to 89 IU/L ($20 < N < 70$). Ultrasound confirmed the presence of five muscle nodules. Thoracoabdomino-pelvic CT revealed multiple bilateral mediastino-hilar adenopathies, a micronodular syndrome of perilymphatic and peribronchovascular distribution within both lungs with a discrete posteroapical predominance.

Muscle MRI was not feasible due to the mechanical valve. Moreover, the muscle biopsy was postponed, initially because of the anticoagulant treatment, and then because the patient refused.

The PET-scan (positron emission tomography) revealed a few discretely hypermetabolic cervico-axillary lymph nodes and multiple bilateral mediastino-hilar adenopathies involving all lymph nodes, as well as multiple bilateral hypermetabolic pulmonary foci. On the subdiaphragmatic level, there was a discrete, diffuse, and homogeneous fixation of the spleen and bilateral external iliac and left inguinal hypermetabolic nodes. Finally, at the musculoskeletal level, several focal nodular muscular hypermetabolisms were reported, predominantly in the lower limbs (Figures 1 and 2).

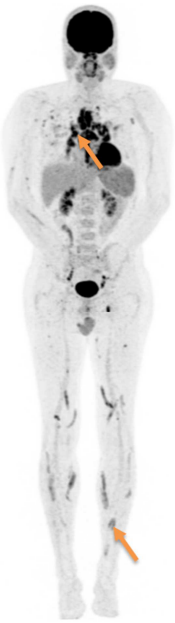

Figure 1. *PET-scan: hypermetabolism of mediastinal adenopathies and muscle nodules of the lower limbs.*

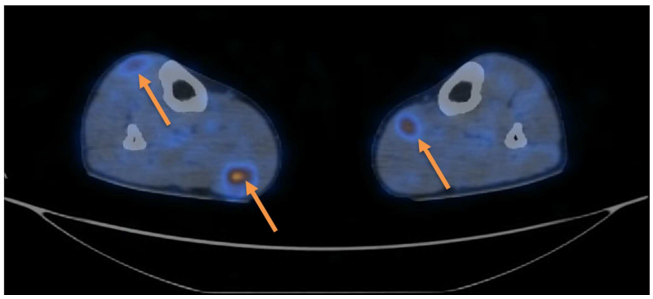

Figure 2. *PET-scan: Hypermetabolism of some muscle nodules of the lower limbs.*

Biopsy of accessory salivary glands (BASG) exhibited an epithelioid and gigantocellular granuloma without necrosis that confirm the diagnosis of systemic sarcoidosis. The search for another location, notably cardiac, ophthalmological, neurological, or renal, was negative. The blood calcium level was normal. The respiratory function tests did not show any restrictive syndrome or abnormalities in the DLCO/VA ratio. It was therefore a systemic sarcoidosis with mediastino-pulmonary stage II, diffuse lymph node, splenic, and muscle involvement.

Systemic corticosteroid therapy was initially considered but was rejected by the patient because of the fear of significant side effects. After 18 months of evolution, the muscle nodules had regressed, and the mediastino-pulmonary involvement had been stabilized.

3. Discussion

Muscle involvement in sarcoidosis was outlined in the early twentieth century with the first clinical form reported in 1908 by Licharew [4]. Subsequent studies have shown that this muscle involvement is frequent, but asymptomatic in many cases being usually diagnosed by using of muscle biopsy [3]. As for the symptomatic muscular sarcoidosis, it is a rare condition associated with multivisceral sarcoidosis involvement, and the need for long-term treatment [5, 6]. Muscle biopsy is not required for the diagnosis of the nodular forms of muscular sarcoidosis, especially the asymptomatic or pauci-symptomatic forms which exhibit good prognosis, as diagnosis can be obtained using simpler means. However, muscle biopsy is required if the examinations do not show clear suggestive lymph node and/or pulmonary lesions.

Three types of involvement have been described: chronic myopathy, acute myositis, and nodular forms [3]. Nodular forms are exceptional [3]. Nodules may be single or multiple within a muscle, ranging in size from a few millimeters to several centimeters, sometimes presenting a pseudotumoral appearance [3]. They are usually palpable and painless but may also present sometimes with myalgia or contractures. They predominate in the lower limbs, without motor deficits [7]. Muscle enzymes and EMG are usually normal [7]. MRI shows well-limited lesions with a central fibrous zone associated with an inflammatory peripheral zone that enhances after injection of contrast substances [8].

In isolated nodular involvement, biopsy is essential

because it is the only way to rule out other etiologies, particularly focal myositis, or nodular vasculitis. Performing systematic muscle biopsies carried out in the context of studies found histological muscle damage in 50 to 80% of cases [9, 10]. In our case, where muscle biopsy was not carried out, BASG was used to obtain histological evidence of sarcoidosis. BASG remains a very useful procedure in the diagnostic search for certain systemic diseases, in particular Sjögren's syndrome, sarcoidosis, and amyloidosis. In sarcoidosis, Michon-Pasturel et al [11] reported, for BASG, a diagnostic efficiency of 38.5% in total (36.4% in Löfgren's syndrome and 42.5% in the other forms, whether there is a dry syndrome or not). In the Baeteman et al study [12] the specificity of BASG was 100%, but its sensitivity was lower and depended on the etiology: 75% for Sjögren's syndrome, 67% for amyloidosis, 60% for sarcoidosis and 14% for other autoimmune diseases. In their study, Lopes AI et al had not found sufficient data to draw conclusions about the use of BASG in the diagnosis of sarcoidosis. They believe that this is probably due to the small sample size of suspected sarcoidosis patients in their study [13].

In our patient, elevated ACE was a good argument for sarcoidosis. Despite its relative value, ACE remains an interesting marker, with a sensitivity of 60-80% depending on the study [14]. Although the correlation is not very good, it increases with the mediastino-pulmonary stage, with higher ACE in stages II and III. The ACE is therefore of diagnostic interest, but also for evolutionary follow-up and prognosis [14].

Therapeutically, corticosteroid therapy is known to be effective in nodular forms of muscular sarcoidosis [7]. Corticosteroid therapy and methotrexate are the most commonly used treatments in the context of sarcoidosis when there is a therapeutic indication [3, 15]. Spontaneous regression is nevertheless possible, as in the reported case.

4. Conclusion

Muscle involvement in sarcoidosis is insufficiently described in the literature. They correspond to an infiltration of the muscle by the sarcoid granuloma. Nodular forms are exceptional. MRI and PET-scan are useful for lesion assessment and extension. Moreover, this clinical case shows us the importance of BASG in the diagnosis of muscular sarcoidosis, in case of impossibility of performing a muscle biopsy.

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