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Case Report

A Rare Case of Extensive Lower Limb Myonecrosis in Catastrophic Antiphospholipid Syndrome: Multimodality Imaging Findings

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Abstract

Catastrophic antiphospholipid syndrome (CAPS) is a rare and life-threatening thrombotic disorder characterized by diffuse microvascular thrombosis leading to multiorgan failure. Musculoskeletal involvement is exceedingly rare, with only isolated cases of muscle infarction reported in the literature. We report the case of a 44-year-old man with CAPS involving the heart, kidneys, and liver who developed acute bilateral lower-limb pain, swelling, and functional impairment. Multimodality imaging revealed extensive lower-limb myonecrosis and played a pivotal role in establishing the diagnosis while excluding the main differential diagnoses. This case highlights the crucial role of imaging, particularly MRI, in the diagnosis of ischemic myonecrosis complicating CAPS, especially as muscle biopsy, although considered the reference standard, is invasive and associated with infectious complications that limit its use. It also expands the spectrum of musculoskeletal manifestations of this disease.

Keywords: *Catastrophic Antiphospholipid Syndrome; Antiphospholipid Syndrome; Myonecrosis; Muscle Infarction; Magnetic Resonance Imaging; Computed Tomography, Stipple Sign*

Introduction

Catastrophic antiphospholipid syndrome (CAPS) is a rare and severe form of antiphospholipid syndrome characterized by widespread thrombotic microangiopathy involving multiple organs over a short period of time. Despite advances in diagnosis and treatment, CAPS remains associated with substantial morbidity and mortality [1,2].

The clinical manifestations of CAPS are heterogeneous and predominantly involve the kidneys, lungs, central nervous system, heart, and skin. Musculoskeletal involvement is exceedingly uncommon, with only isolated reports of skeletal muscle infarction and myonecrosis available in the literature [2,3].

Case Presentation

A 44-year-old man with no significant past medical history was admitted to the Department of Internal Medicine for the management of catastrophic antiphospholipid syndrome (CAPS) with multivisceral involvement, including cardiac, renal, and hepatic manifestations. The diagnosis of CAPS was established based on rapidly progressive multiorgan dysfunction associated with positive antiphospholipid antibodies.

One week after admission, the patient developed progressive bilateral lower limb pain, swelling, and functional impairment, associated with diffuse erythema and multiple bullous skin lesions. Physical examination revealed marked bilateral lower extremity edema, more pronounced on the left side, with diffuse tenderness and marked limitation of limb movement, without any history of trauma or preceding infectious symptoms.

Given the concern for an acute vascular complication, emergency CT angiography of the lower extremities was performed. Imaging demonstrated marked enlargement of the left leg compared with the contralateral side, with diffuse edema involving multiple muscular compartments. Extensive areas of muscular hypoenhancement were observed, surrounded by peripheral curvilinear enhancement following intravenous contrast administration. Associated subcutaneous edema and inflammatory changes of the adjacent soft tissues were also noted. No evidence of arterial occlusion, venous thrombosis, soft-tissue gas, or organized fluid collection was identified.

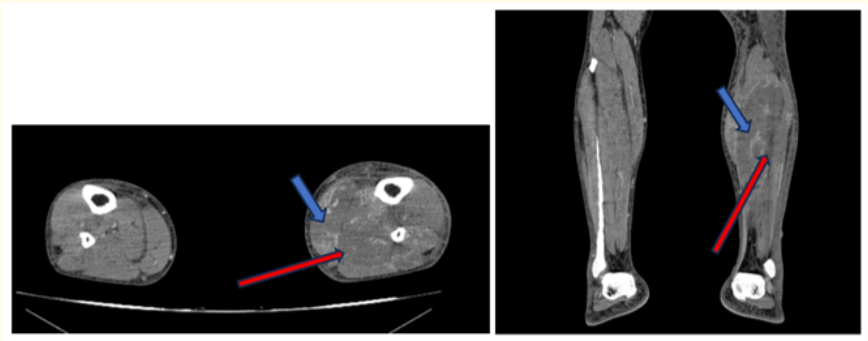

Figure 1: *Contrast-enhanced CT of the lower limbs. Axial (A) and coronal (B) images show extensive non-enhancing defects involving multiple muscle compartments of the left lower limb (red arrow), with peripheral curvilinear enhancement (blue arrow) outlining the necrotic muscle, consistent with ischemic myonecrosis.*

Magnetic resonance imaging (MRI) was subsequently performed for further lesion characterization. T2-weighted and STIR sequences demonstrated extensive intramuscular edema involving all muscle compartments of the left lower extremity, as well as the adjacent fascial planes and subcutaneous soft tissues. Post-contrast T1-weighted images revealed intense peripheral muscular enhancement surrounding large areas of non-enhancing muscle tissue, consistent with extensive myonecrosis.

In addition, multiple punctate and curvilinear enhancing foci were identified within the non-enhancing muscle segments, corresponding to the characteristic "stipple sign" of acute myonecrosis. This finding is thought to represent residual viable myofibers or inflammatory vascular structures embedded within necrotic muscle tissue.

In the setting of catastrophic antiphospholipid syndrome with multivisceral involvement, these imaging findings were highly suggestive of extensive ischemic myonecrosis secondary to diffuse thrombotic microangiopathy.

The patient was subsequently treated with anticoagulation and immunosuppressive therapy as part of the management of CAPS. Clinical and imaging follow-up was performed to assess disease progression and therapeutic response.

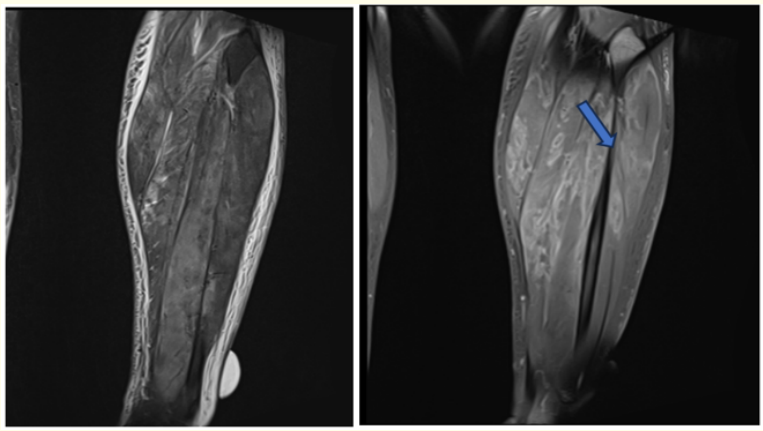

Figure 2: *MRI of the left lower limb. Coronal T2-weighted STIR image (A) demonstrates diffuse muscle edema involving multiple muscle compartments, associated with extensive subcutaneous edema. Contrast-enhanced fat-suppressed T1-weighted image (B) shows multiple non-enhancing areas of muscular necrosis surrounded by peripheral enhancement, with numerous punctate enhancing foci within the necrotic muscle, producing the characteristic "stipple sign" (Blue arrow).*

Discussion

Catastrophic antiphospholipid syndrome (CAPS) represents the most severe form of antiphospholipid syndrome. It is characterized by diffuse small-vessel thrombosis leading to rapidly progressive multiorgan failure. The currently accepted diagnostic criteria include involvement of at least three organs, systems, or tissues, development of manifestations within one week, histopathological evidence of small-vessel occlusion when available, and the persistent presence of antiphospholipid antibodies [2,4].

Muscle involvement is an exceptionally rare manifestation of CAPS. Its pathophysiology is thought to result from diffuse microvascular thrombosis leading to prolonged ischemia, muscle infarction, and ultimately myonecrosis. Similar mechanisms have been proposed in diabetic myonecrosis, in which microangiopathy and a hypercoagulable state play a major role. However, the specific role of antiphospholipid antibodies in the development of muscle infarction remains poorly understood because of the extreme rarity of this complication and the frequent coexistence of other factors promoting muscle ischemia, particularly diabetic microangiopathy [5-7].

Only a few cases of muscle infarction associated with antiphospholipid antibodies have been reported in the literature. Palmer and Greco described the first two cases in patients with type 1 diabetes mellitus and positive antiphospholipid antibodies. Subsequently, Boulman., *et al*. reported a similar case in a patient with type 2 diabetes mellitus and positive lupus anticoagulant, whereas Nishikai., *et al*. described muscle infarction in a non-diabetic patient with positive anticardiolipin antibodies, leading to the diagnosis of probable antiphospholipid syndrome [3,8,9].

Clinically, acute myonecrosis should be suspected in patients presenting with non-traumatic muscle pain associated with swelling and functional impairment, particularly in the setting of a systemic thrombotic disorder. Because the clinical and laboratory findings are nonspecific, imaging plays a pivotal role in establishing the diagnosis [6,7,10].

Computed tomography may demonstrate muscle enlargement, decreased attenuation of the affected muscles, and adjacent soft-tissue edema. However, MRI remains the imaging modality of choice owing to its superior soft-tissue contrast resolution. T2-weighted and STIR

images typically demonstrate diffuse hyperintense signal abnormalities involving the affected musculature, subcutaneous tissues, and subfascial planes, consistent with extensive edema. Following gadolinium administration, contrast-enhanced images accurately delineate areas of myonecrosis, which appear as non-enhancing regions with peripheral rim enhancement, reflecting reactive hyperemia at the margins of the necrotic muscle. Within these necrotic areas, multiple punctate, linear, or curvilinear enhancing foci may be observed, producing the characteristic "stipple sign". Initially described in diabetic myonecrosis and rhabdomyolysis, this sign is thought to represent residual viable muscle fibers or preserved vascular structures within necrotic tissue and constitutes a valuable imaging feature suggestive of ischemic myonecrosis. Nevertheless, diffuse T2/STIR hyperintensity remains nonspecific and may also be encountered in infectious or inflammatory myopathies, traumatic muscle injury, compartment syndrome, subacute denervation, early myositis ossificans, rhabdomyolysis, abscesses, and skeletal muscle metastases. In this context, recognition of the stipple sign is particularly useful in narrowing the differential diagnosis [10-14].

In our case, the diagnosis of myonecrosis was established on the basis of multimodality imaging findings in conjunction with the clinical and biological features of CAPS. Recognition of muscle involvement is of particular importance, as it may represent an additional marker of disease severity and widespread thrombotic burden [5,6].

Although muscle biopsy remains the histopathological gold standard, demonstrating muscle fiber necrosis associated with inflammatory cell infiltration, it is rarely required when imaging findings are characteristic. Because it is invasive and carries risks of infection, delayed wound healing, and worsening of muscle injury, biopsy should be reserved for cases with inconclusive imaging findings or when an infectious or neoplastic process cannot be confidently excluded [7,10,15].

The treatment of CAPS relies on a multimodal approach combining anticoagulation, high-dose corticosteroids, plasma exchange, intravenous immunoglobulins, and, in refractory cases, immunosuppressive agents or biologic therapies. Management of myonecrosis is primarily conservative, with surgery reserved for patients who develop compartment syndrome, secondary infection, or complicated necrosis. Early diagnosis is crucial, as delayed treatment is associated with a poorer prognosis [2,4,16].

Conclusion

Myonecrosis is an exceptionally rare manifestation of catastrophic antiphospholipid syndrome. MRI plays a pivotal role in identifying muscle necrosis and differentiating it from its numerous mimickers. Awareness of this unusual presentation may facilitate prompt diagnosis and initiation of life-saving therapy in patients with multivisceral CAPS.

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