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Case Report

Leg muscle involvement in polyarteritis nodosa (limited form)—A rare case report ☆

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ABSTRACT

Polyarteritis nodosa is a necrotizing arteritis involving small and medium vessels. Polyarteritis nodosa can have variable presentations depending upon the organ involved. It can either present as a diffuse disease or a limited form confined to a particular organ. Isolated muscular involvement in this disease is rare, which may present as myalgia, nonspecific fever, weight loss or even as claudication. The imaging pattern on ultrasound and MRI can help diagnose this condition in the background of clinical history and muscle biopsy is confirmatory. We present a case of 15 years old boy who presented with fever, weight loss, myalgia in leg region. MRI and ultrasound examination showed perivascular inflammation in calf muscles with a characteristic "cotton wool" pattern of enhancement on contrast study.

Introduction

Polyarteritis nodosa (PAN) is a necrotizing arteritis predominantly involving medium and small muscular arteries, without glomerulonephritis or vasculitis in arterioles, capillaries, or venules [1,2]. PAN can affect a variety of organs and systems as arteries are distributed ubiquitously throughout the body. Systems and organs frequently involved include the peripheral nervous system, skin, gastrointestinal tract, kidneys, and skeletal muscles [3]. PAN has protean manifestations, and its presentation may be a systemic disease or be confined to a single organ (limited form) [2]. There is an inherent diagnostic challenge to diagnose limited PAN, as it is rare and has a nonspecific presentation.

Patients with musculoskeletal system involvement of PAN present clinically as myalgia, arthralgia, polymyositis-like syndrome, asymmetric nondeforming polyarthritis, and, less commonly with intermittent claudications, acute leg ischemia, or myopathy [2]. Skeletal muscle involvement of PAN may occur as a constituent of systemic PAN or as a limited form.

Abbreviations: PAN, Polyarteritis nodosa.

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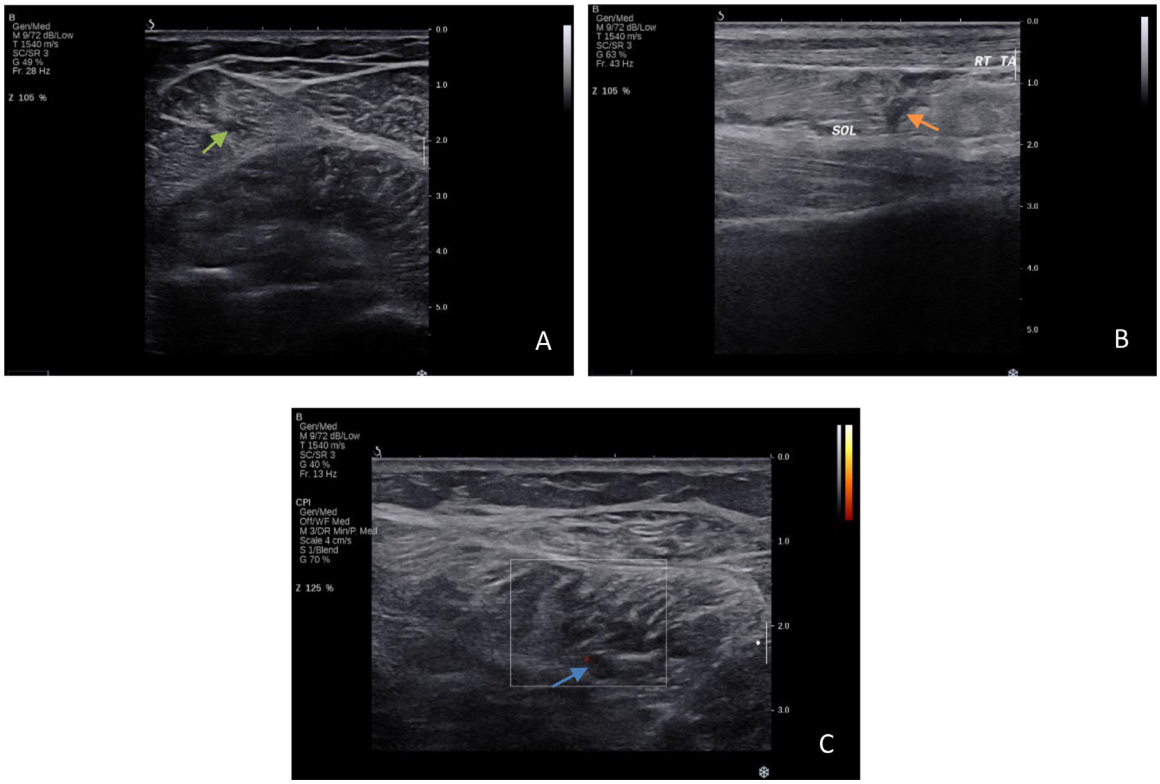

Fig. 1 – High-resolution ultrasound images showing ill-defined hypoechoic areas in gastrocnemius (light green arrow in A), and soleus (orange arrow in B) muscle. Hypoechoic area is present surrounding an intramuscular vessel (blue arrow in C) showing perivascular inflammation.

Patients with the limited form are known to present with pain, swelling, and tenderness of the lower leg [4]. In these cases, the clinical picture may mimic those of deep vein thrombosis, soft-tissue infection, or even compartment syndrome.

MRI findings of skeletal muscle involvement in PAN have been reported in several individual cases. Various studies have reported different patterns of muscle edema (as diffuse or patchy T2/short tau inversion recovery [STIR] sequence signals) and/or contrast enhancement [4–12]. One case series described MRI findings of 3 patients with vasculitis restricted to limbs, in which 2 cases were of PAN [9]. In a case series of 8 patients with lower extremity muscle involvement in PAN, Kang et al. [15] described the enhancement pattern commonly seen in these patients as “cotton wool enhancement” pattern.

Case report

A case of 15 years male, presented with complaints of on and off fever, loss of weight, and arthralgia for 6 months. Over the past 2 months, the patient also developed complaints of pain in bilateral legs predominantly in the calf region, which aggravated during the fever episodes. On examination, his temperature was 37.8°C, pulse rate 90/min, blood pressure 116/72 mm Hg, and respiratory rate 16/min. All peripheral pulses were palpable and abdominal examinations were normal. He had no demonstrable rash or erythema over the skin or face.

The initial laboratory results were as follows: Hb 9.96 g/dL, total leukocyte count 12,600/mm^3, polymorphs 71%, serum calcium (ionic) 1.18 mmol/L, serum urea 18 mg/dL, normal liver function test, normal kidney function test, presence of blood in urine with RBC 6/hpf, angiotensin-converting enzyme 55.83 mg/L (reference range 12-68), raised erythrocyte sedimentation rate 55mm/h, mildly raised C-reactive protein. Hepatitis B, hepatitis C, and HIV serology tests were negative. Cold agglutinin, antinuclear antibody, and antineutrophil cytoplasmic antibody were all negative. 2D echo, ultrasound abdomen were within normal limits.

High-resolution ultrasound of both legs done using 5-12 MHz linear probe revealed multiple ill-defined hypoechoic lesions centered around blood vessels in bilateral gastrocnemius, soleus and tibilas anterior muscles (Fig. 1). These lesions suggested perivascular inflammatory changes within the muscles. Doppler ultrasound of both legs showed no evidence of luminal narrowing or thrombosis/plaque within the vessels.

MRI of both legs done on 1.5 T MRI machine, showed multifocal intramuscular ill-defined patchy areas of high signal on T2-weighted images, STIR and proton density fat suppressed sequences, involving bilateral gastrocnemius, soleus and tibialis anterior muscles (Fig. 1). These areas were isointense on precontrast T1-weighted images. On postcontrast T1-weighted images, there were small multifocal ill-defined enhancing areas noted centered around blood vessels, with fuzzy outline (Fig. 2).

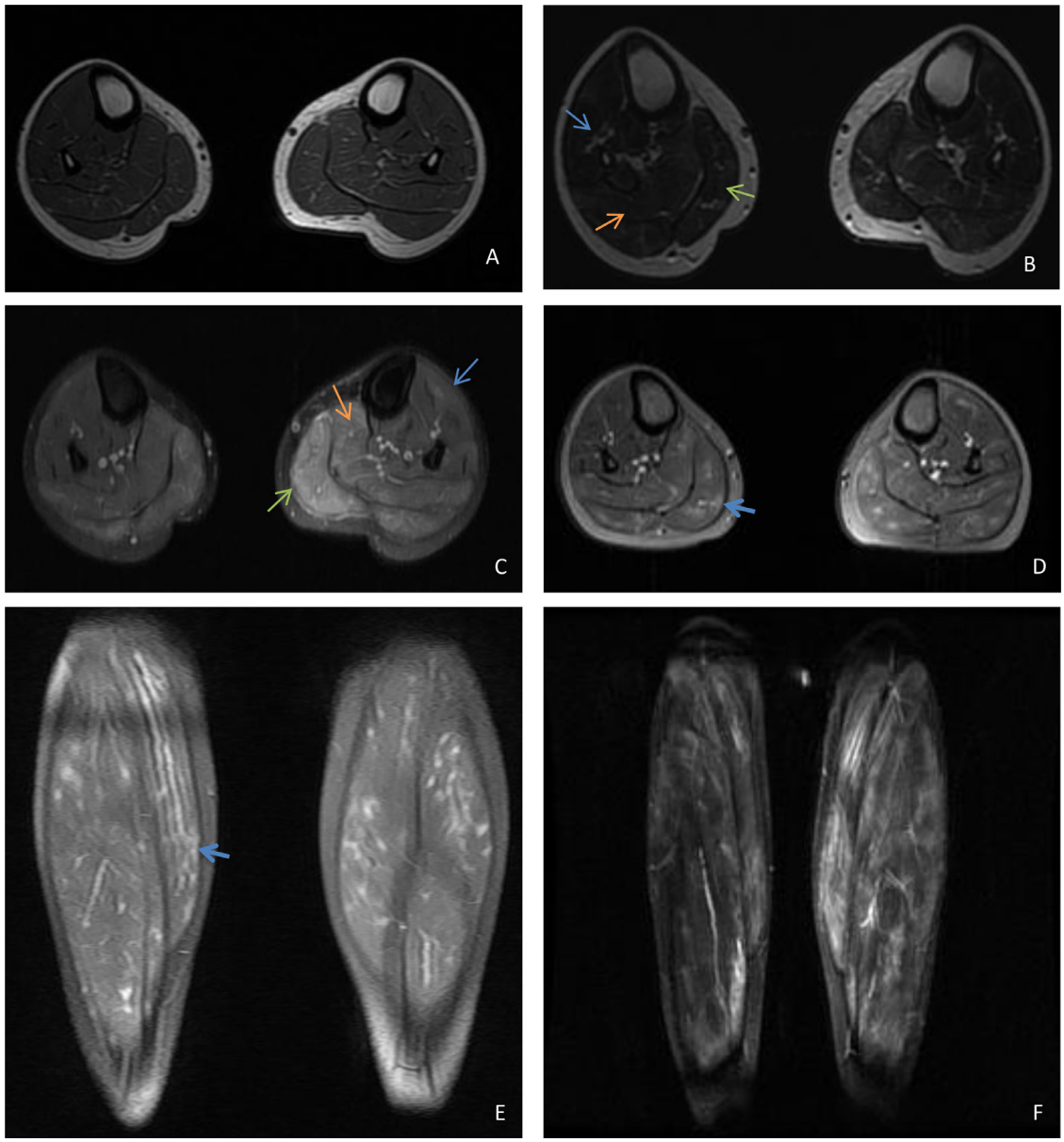

Fig. 2 – Axial T1 weighted image showing no signal abnormality (A), axial T2 weighted image (B), axial PDFS (C), coronal PDFS (F) shows patchy areas of high signal involving tibialis anterior (blue arrow), gastrocnemius (green arrow) and soleus (orange arrow) muscles of both legs. Post contrast axial (D) and Coronal (E) images show fluffy nodular enhancing areas around the intramuscular vessels termed "cotton wool appearance." PDFS, proton density fat suppressed.

Muscle biopsy was performed and the patient was diagnosed with PAN confined to both legs.

The patient was treated with oral steroids. There was resolution of fever and muscular pain after 2 weeks of treatment. The patient is kept on follow up in OPD.

Discussion

The MRI findings of isolated skeletal muscle involvement in PAN have been sparsely reported. Previously reported cases [4–12] showed either patchy or diffusely increased signal intensity of the involved muscles on T2-weighted or STIR images. The high signal on T2-weighted images is likely due to edema, probably due to an increase in intracellular or extracellular free water [9,13].

Kang et al. [15] showed the enhancement pattern of muscle involvement in PAN where they demonstrated fluffy enhancing lesions centered on branching vessels on contrast-enhanced images, for which they coined the term "cotton-wool appearance." Our case also showed similar enhancement pattern in the leg muscles of both limbs. The pathophysiology of skeletal muscle involvement of PAN has been explained in several ways, including ischemia resulting from occlusion of blood vessels, involvement of muscle fibers by the inflamma-

tion around the blood vessels, and muscle atrophy due to extensive damage of the peripheral nerves [14,15].

Biopsy specimens of the involved muscle in PAN have been reported to show areas of atrophy, degeneration of muscle fibers, and myonecrosis, which is indicative of ischemic injury, which thus supports the hypothesis [14,16].

The cotton-wool appearance was explained by the deteriorated blood flow due to the occlusion or narrowing of blood vessels leading to a sluggish diffusion of the contrast media into the extravascular space, which exhibits a nodular appearance centered on blood vessels [15].

The fluffy nodular enhancing lesions may not be a specific feature of PAN. Schulze et al. [16] showed similar ill-defined focal enhancing lesions in the muscles in Behcet disease and familial Mediterranean fever. Akansel et al. [17] reported a case of localized myositis in a patient with Behcet disease, which appeared as focal enhancing lesions in the affected muscles. To date, muscle biopsy remains the gold standard for diagnosis of muscle involvement of PAN. However, the recognition of signal alteration patterns in muscles may help suggest a diagnosis of PAN, especially when no systemic stigmata of vasculitis are present.

Conclusion

PAN should be considered in differentials in cases of patchy muscle signal changes on MRI, with fluffy nodular enhancing lesions centered on vessels on contrast-enhanced images.

Patient consent

A written informed consent was obtained from the patient for publication of this case, images and the clinical details.

REFERENCES

[1] Jennette JC, Falk RJ, Bacon PA, Basu N, Cid MC, Ferrario F, et al. 2012 revised International Chapel Hill consensus conference nomenclature of vasculitides. Arthritis Rheum 2013;65:1–11.

[2] Colmegna I, Maldonado-Cocco JA. Polyarteritis nodosa revisited. Curr Rheumatol Rep 2005;7:288–96.

[3] Hernández-Rodríguez J, Alba MA, Prieto-González S, Cid MC. Diagnosis and classification of polyarteritis nodosa. J Autoimmun 2014;48:84–9.

[4] Nakamura T, Tomoda K, Yamamura Y, Tsukano M, Honda I, Iyama K. Polyarteritis nodosa limited to calf muscles: a case report and review of the literature. Clin Rheumatol 2003;22:149–53.

[5] Yang SN, Cho NS, Choi HS, Choi SJ, Yoon ES, Kim DH. Muscular polyarteritis nodosa. J Clin Rheumatol 2012;18:249–52.

[6] Hofman DM, Lems WF, Witkamp TD, Putte VD, Bijlsma JW. Demonstration of calf abnormalities by magnetic resonance imaging in polyarteritis nodosa. Clin Rheumatol 1992;11:402–4.

[7] Révelon G, Rahmouni A, Jazaerli N, Godeau B, Chosidow O, Authier J, et al. Acute swelling of the limbs: magnetic resonance pictorial review of fascial and muscle signal changes. Eur J Radiol 1999;30:11–21.

[8] Ahmed S, Kitchen J, Hamilton S, Brett F, Kane D. A case of polyarteritis nodosa limited to the right calf muscles, fascia, and skin: a case report. J Med Case Reports 2011;5:450.

[9] Gallien S, Mahr A, Rety F, Kambouchner M, Lhote F, Cohen P, et al. Magnetic resonance imaging of skeletal muscle involvement in limb restricted vasculitis. Ann Rheum Dis 2002;61:1107–9.

[10] MacDonald WB, Blake MP. Periostitis and localized myositis in polyarteritis nodosa. Clin Nucl Med 2004;29:703–5.

[11] Esteva-Lorenzo FJ, Ferreiro JL, Tardaguila F, de la Fuente A, Falasca G, Reginato AJ. Case report 866: pseudotumor of the muscle associated with necrotizing vasculitis of medium- and small-sized arteries and chronic myositis. Skeletal Radiol 1994;23:572–6.

[12] Reading PJ, Hudgson P, Johnson MA, Jenkins A. A sartorial challenge. Lancet 1999;354:996.

[13] May DA, Disler DG, Jones EA, Balkissoon AA, Manaster BJ. Abnormal signal intensity in skeletal muscle at MR imaging: patterns, pearls, and pitfalls. RadioGraphics 2000;20:S295–315.

[14] Matsubara S, Mair WG. Ultrastructural changes of skeletal muscles in polyarteritis nodosa and in arteritis associated with rheumatoid arthritis. Acta Neuropathol 1980;50:169–74.

[15] Kang Y, Hong SH, Yoo HJ, Choi JY, Park JK, Park J, et al. Muscle involvement in polyarteritis nodosa: report of eight cases with characteristic contrast enhancement pattern on MRI. AJR Am J Roentgenol 2016;206(2):378–84.

[16] Schulze M, Kotter I, Ernemann U, Fenchel M, Claussen CD, et al. MRI findings in inflammatory muscle diseases and their noninflammatory mimics. AJR 2009;192:1708–16.

[17] Akansel G, Akgoz Y, Ciftci E, Arslan A, Demirci A. MRI findings of myositis in Behcet disease. Skeletal Radiol 2004;33:426–8.