CASE STUDIES

A case of giant cell arteritis with lower extremity myositis on MRI

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Abstract

Giant cell arteritis (GCA), seen mainly in elders and rarely in children, has a female predominance. The present study discusses a case of biopsy-proven GCA with associated bilateral lower extremity myositis diagnosed on MRI and normal muscle enzymes. There are case studies on the occurrence of polymyalgia rheumatica (PMR) in GCA, but the involvement of lower limbs has not been reported. This report reiterates the importance of high clinical suspicion in the setting of occult and atypical presentations to avoid diagnostic delays.

Keywords: giant cell arteritis (GCA), temporal arteritis, polymyalgia rheumatica (PMR), myositis

Introduction

Giant cell arteritis (GCA) is a well-documented systemic vasculitis affecting medium- and large-sized vessels, most commonly of the head and neck.[1-4] The disease is predominant in the elderly, with an average age of onset of 70 years. The prevalence is estimated to be 1 in 500 individuals of >50 years of age.[5] The disease is more common in women, Caucasians, and those of European descent. Classic symptoms include headache, scalp tenderness, and jaw claudication, and their specificity is greater than 99% when present in combination.[6] Sudden vision loss is one of the hallmark features and it occurs in up to one-fifth of patients.[7]

The overlap between GCA and polymyalgia rheumatica (PMR) has been well recognized, so patients may also present with hip and shoulder girdle pain, weight loss, and fatigue. The present case focuses on a rare association of GCA with concurrent myositis.

Case report

A 77-year-old Caucasian male was admitted to the hospital with intractable lower extremity pain and swelling. His medical history revealed: type II diabetes mellitus, an indeterminate thyroid nodule and PMR, which had been diagnosed 12 years prior. At that time, he had hip and shoulder pain, which resolved within 24 hours of treatment with oral prednisone. Seven years later, the patient experienced recurrent episodes of PMR, which again responded rapidly to steroids. Since then, he had remained asymptomatic and did not receive any systemic steroids.

Three months prior to the current admission, he had recurrence of shoulder and pelvic girdle pain. He was on steroid tapers almost continuously throughout this period, ranging from doses of prednisone 10 mg to 40 mg daily. Three weeks prior to the admission, he developed acute pain in both lower extremities (left greater than right), bilateral lower extremity swelling and difficulty walking. He had transient vision loss in his right eye induced by bright lights for the same duration. He also reported fever of around 102 °F, drenching night sweats, and unintentional weight loss of about 20 pounds for weeks to months. He denied having symptoms of jaw claudication or scalp tenderness. White blood cell count (WBC) was 18.0×10^9/L, erythrocyte sedimentation rate (ESR) was 91 mm/hr, and creatine phosphokinase (CPK) was 10 IU/L. Arterial duplex scans of the lower extremities revealed ankle-brachial indices of 1.04 (right) and 1.1 (left), with moderate disease of the right posterior tibial artery.

He was evaluated by an ophthalmologist and found to have normal fundoscopic and slit lamp examinations with 20/20 vision, normal intraocular pressures and intact visual fields. He had bilateral pedal edema, diffuse tenderness to palpation in his legs, and 4/5 strength with left hip flexion. Repeat inflammatory markers revealed an ESR of 112 mm/

hr, and C-reactive protein (CRP) of 26.6 mg/dL (upper limit normal 0.75). Anti-nuclear antibody (ANA), anti-neutrophil cytoplasmic antibody (ANCA), C3 and C4 complement, and rheumatoid factor were within normal limits. Carotid artery Doppler revealed no hemodynamically significant stenosis, and venous Dopplers of the lower extremities were negative for thrombus. MRI of the lower extremities revealed diffuse intramuscular T2 hyperintensity involving all three compartments in the left calf, representing a non-specific myositis (Fig. 1), with similar findings in the right leg. Biopsy of the right temporal artery revealed granulomatous changes consistent with GCA (Fig. 2). He was started on methylprednisolone 1 gm intravenously for 3 days and subsequently changed to prednisone 30 mg twice daily. He had a rapid improvement in his pain and lower extremity edema upon increasing prednisone dosing. Follow-up conducted at first and third month post discharge demonstrated normalization of ESR and CRP. He remained on a slow steroid taper for one year, currently at a dose of prednisone 7 mg daily. It has been planned to continue tapering over several more months. He has had no recurrence of leg pain or GCA symptoms.

Discussion

MRI muscle edema may be the result of inflammatory myopathies, infections, trauma, muscle infarction or denervation. In vasculitis, MRI findings typically show a focal patchy muscle involvement.[8] The present case describes

Fig. 1: Magnetic resonance imaging of the lower extremities demonstrating diffuse intramuscular hyperintensity bilaterally on coronal T2 (a) and axial STIR (b) sequences with deep fascial and subcutaneous edema

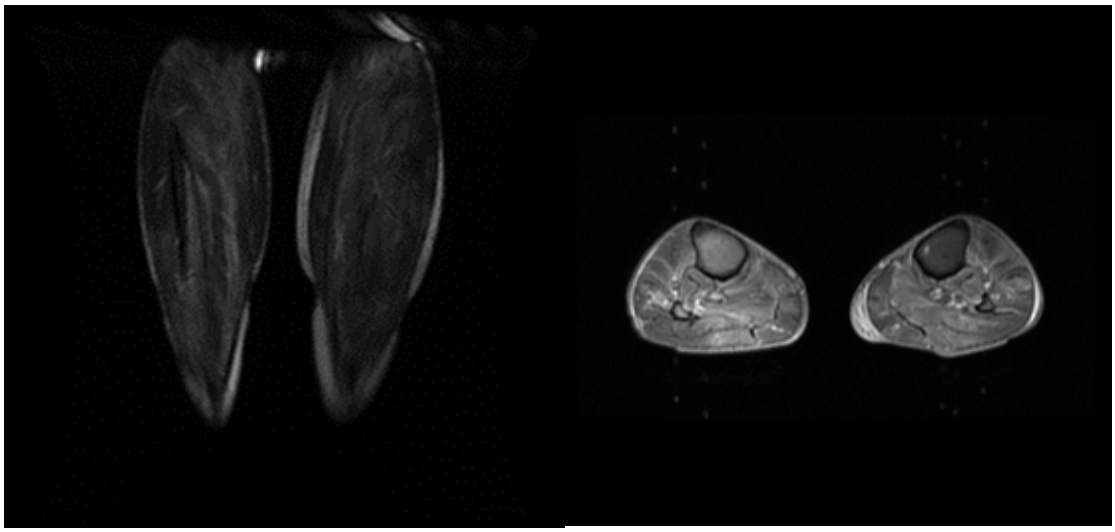
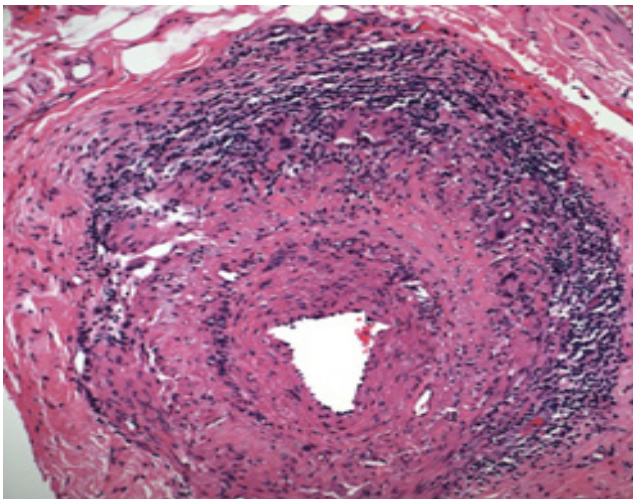


Fig. 2: Temporal artery biopsy demonstrating intramural inflammation with giant cells and some fibrosis



an elderly male patient presenting with intractable lower extremity pain and swelling, who had biopsy-proven GCA as well as non-specific myositis of the lower extremities on MRI, and normal muscle enzymes. Vasculitis has been shown to occur uncommonly in muscle biopsy specimens of patients presenting with a variety of conditions, either as a primary process or secondary to an underlying inflammatory myopathy.[9] However, an association between myositis and GCA has not been well-documented. To the best of our knowledge, there is only one case in the existing literature of concomitant occurrence of GCA with overt myositis, which was incidentally identified during post mortem.[10] However, there have been several reports of diffuse myositis on MRI, with normal or minimally elevated CPK and normal muscle biopsies in patients who were found to have polyarteritis nodosa (PAN), suggesting that MRI muscle edema can be seen diffusely in systemic vasculitides.[11,12] A muscle biopsy was not conducted in the present patient, as a dramatic improvement in his muscle symptoms was noted following the treatment for underlying GCA. This report illustrates the possibility that diffuse myositis on MRI with normal or minimally elevated CPK should raise clinical suspicion for an underlying vasculitis, including GCA in the appropriate patient population. Tissue biopsy, of either the affected muscle or an easily accessible involved vessel, should be performed to confirm the diagnosis.

In addition to documenting this atypical association, the present case is illustrative of the need for a high clinical suspicion in the presence of atypical manifestations to conclude the diagnosis of GCA. The present patient did not show any 'classic' features of GCA such as headache, jaw claudication or scalp tenderness. Moreover, it is crucial to remember that the sensitivities of these findings are quite poor, with negative predictive values on the order of 70%.[6]

Key message

Giant cell arteritis can be associated with a non-specific myositis, even in the absence of elevated muscle enzymes.

Competing interests

The authors declare that they have no competing interests.

Citation

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