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Case Report

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An unusual manifestation of fever, eosinophilia, and proximal muscle weakness: idiopathic eosinophilic myositis accompanied by eosinophilic vasculitis

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ABSTRACT

Eosinophilia-associated myopathies exhibit clinical and pathological heterogeneity. This is a man in his sixties who presented with fever accompanied by prodromal symptoms, as well as bilateral lower limb weakness and pain. There was evidence of peripheral eosinophilia, increased CPK levels, polyphasic muscle action potentials, and eosinophilic infiltration into the muscle with eosinophilic vasculitis present, without cutaneous or myofascial involvement. Serological and imaging studies were unremarkable, revealing no discernible secondary aetiology for eosinophilia or myositis. A diagnosis of idiopathic eosinophilic myositis accompanied by eosinophilic vasculitis was established. He received glucocorticoids and methotrexate, resulting in effective symptom relief.

Keywords: Idiopathic eosinophilic myositis, eosinophilic vasculitis, eosinophilia

INTRODUCTION

Eosinophilia-associated myopathies exhibit clinical and pathological heterogeneity. Idiopathic inflammatory myopathies are characterised by the presence of inflammatory infiltrates—specifically, lymphocytes and macrophages in muscle tissue. A pathogenic continuum exists that encompasses the involvement of muscle, fascia, and skin, with varied degrees of engagement across the different layers [1]. This type of illness is marked by the presence of peripheral and muscular eosinophilia. A man in his 60s presented with fever, eosinophilia, bilateral lower limb weakness, and pain, along with evidence of eosinophil infiltration into the muscle. He had no recognised aetiology and was diagnosed with idiopathic eosinophilic myositis and eosinophilic vasculitis.

CASE REPORT

A man in his 60s presented with a two-week history of fever and chills, accompanied by constitutional symptoms, arthralgia, myalgia, anorexia, and weight loss. He experienced bilateral lower limb weakness and soreness, resulting in impaired mobility. No variations were observed in lower limb weakness, upper limb weakness, facial muscle weakness, diplopia, or speaking difficulties.

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He experienced respiratory distress accompanied by a mild non-productive cough. There was no diarrhoea, gastrointestinal discomfort, alterations in urine output, painful urination, blood in urine, pleuritic chest pain, skin rashes, joint oedema, or exposure to pets, birds, or other occupational hazards. No previous history of using over-the-counter drugs, Ayurvedic treatments, statins, glucocorticoids, or other pharmaceuticals was noted. There was no previous history of allergies. Upon examination, he exhibited no fever and showed no signs of pallor, jaundice, lymphadenopathy, rashes, mouth ulcers, or oedema. The cardiovascular, pulmonary, and abdominal examinations were unremarkable.

The neurological assessment indicated intact cranial nerves and a normal upper limb evaluation. Bilateral lower limbs exhibited diminished strength, with a Medical Research Council (MRC) grade of 3/5 proximally and 4/5 distally. Tenderness was observed upon palpation, accompanied by diminished tone, reflexes, and sensory perception. His examinations disclosed indications of elevated white blood cell counts at 19.78 x 10^9/L (<8 x 10^9/L), with an eosinophilic preponderance of 6.725 x 10^9/L (<0.5 x 10^9/L) - 34% (<5%). Additionally, CRP was elevated at 256 mg/dL (<6), ESR was 25 mm/hour (<20), the urine complete report was normal, and both blood and urine cultures were negative. His serum creatinine was 1.11 (eGFR - 74) and 0.84 (eGFR - 96) mg/dL (<1 mg/dL), potassium was 3.8 mmol/L (<4), and sodium was 133 mmol/L (135-145), all of which were within normal limits. The liver profile indicates AST at 93 U/L (normal <40), ALT at 158 U/L (normal <40), ALP at 134 IU/L (normal <150), GGT at 22 U/L (normal <40), albumin at 2.1 g/dL (normal <4), and globulin at 3.2 g/dL (normal <5). His CPK level was 3097 U/L (normal range: <200).

The blood analysis indicated signs of an acute infection; however, the coagulation parameters were within normal limits: PT 1.02 (<1.5) and APTT 20.1 s (<35). Persistent eosinophilia prompted consideration of parasite infections, cancer, autoimmune illness, and EGPA. His screening for parasite infections (Toxocara, Toxoplasma, SAT, and filaria antibodies), cancer, and vasculitis (pANCA, cANCA, and ANA) yielded negative results. Given the proximal muscle weakness accompanied by high inflammatory markers, idiopathic inflammatory myositis, overlap myositis, and polymyositis were contemplated. The anti-Jo and PM-Scl antibodies were negative.

Table 1: Haematological, biochemical and microbiological investigations of the patient.

Investigation	Result	Reference range
White blood cell count	19.78 x 10^9/L	<8 x 10^9/L
Eosinophilia	6.725 x 10^9/L - 34%	<0.5 x 10^9/L (<5%)
CRP	256 mg/dL	<6 mg/dL
ESR	25 mm/hour	<20 mm/hour
UFR	Negative (No evidence of an active sediment)	
UPCR	Negative	
Blood culture	Negative	
Urine culture	Negative	
Serum creatinine (eGFR)	1.11 (74) – 0.84 (96)	< 1 mg/dL
Potassium	3.8 mmol/L	< 4.0 mmol/L
Sodium	133 mmol/L	135-145 mmol/L
AST	93 U/L	<40 U/L

ALT	158 U/L	<40 U/L
ALP	134 IU/L	<150 IU/L
GGT	22 U/L	<40 U/L
CPK level	3097 U/L	<200 U/L
PT	1.02	<1.5
APTT	20.1 s	<35 s
TSH	2.2 mIU/L	0.4 – 4 mIU/L
T4	0.9 ng/dL	0.8 – 1.2 ng/dL
ODST	30 nmol/L	< 50 nmol/L
ANA	Negative	
pANCA	Negative	
cANCA	Negative	
SAT	Negative	
Filaria antibody test	Negative	
Toxocara antibody test	Negative	
Toxoplasma antibody test	Negative	
Anti-Jo 1 antibody test	Negative	
Anti PM-Scl antobody test	Negative	

His ultrasound, transthoracic echocardiogram, contrast-enhanced computed tomography (CECT) of the thorax, abdomen, and pelvis, and high-resolution computed tomogram (HRCT) of the chest were normal. The ECG revealed indications of recently diagnosed atrial flutter. The endocrine panel results were as follows: TSH – 2.2 mIU/L (reference range: 0.4 - 4), T4 – 0.9 ng/dL (reference range: 0.8 – 1.2), and ODST – 30 (reference range: <50), all of which were within normal limits. Electromyography revealed multi-unit potentials with polyphasia in the muscles of both the upper and lower limbs. The muscle biopsy revealed skeletal muscle tissue characterised by dense interstitial inflammatory infiltration, including numerous eosinophils, a significant quantity of neutrophils, and a small number of histiocytes and lymphocytes.

Evidence of degenerating and repairing muscle fibres was seen. Numerous big cells of potential muscular origin and a limited number of Langerhans-type giant cells are observed. A well-formed granuloma was absent. A component of vasculitis accompanied by modest epimysial fibrosis was seen. The patient's muscle biopsy demonstrates eosinophil infiltration in the muscle (Fig. 1).

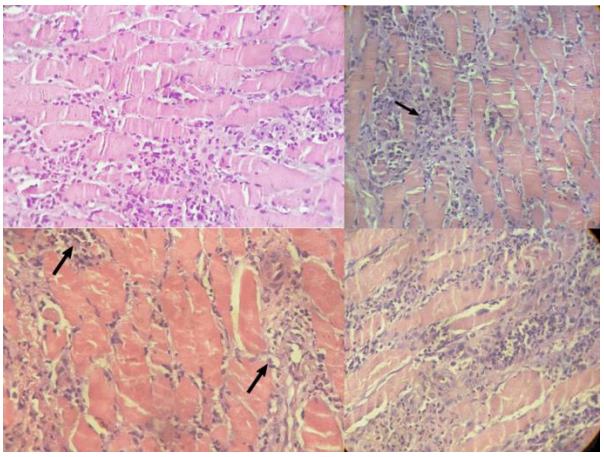

Figure 1: Muscle biopsy showing an abundance of inflammatory cells largely composed of eosinophils (arrows) in the perivascular and interstitial areas

He was initiated on intravenous antibiotics (ceftriaxone 2 g daily) and maintained on the antibiotic for a duration of ten days. Following the muscle biopsy results, he was initiated on high-dose intravenous methylprednisolone pulses of 1 gram daily for five days in the induction phase and

later transitioned to oral glucocorticoids, initially at 60 mg daily and gradually tapered off over a duration of six months. He was started on oral methotrexate (10 mg daily) in the maintenance phase as a steroid-sparing agent with ongoing monitoring of his liver transaminases. Six months following the initiation of glucocorticoids and the continuation of methotrexate, his myalgia diminished, and muscle strength was enhanced to mMRC-4+/5, accompanied by a steady decrease in CPK to 275 U/L (<200) and a reduction in eosinophil count to 0.6 x 10^9/L (<1 x 10^9/L) over a duration of six months.

DISCUSSION

Eosinophilic myositis represents an uncommon and clinically heterogeneous disorder within the broader spectrum of eosinophilia-associated myopathies. These conditions are characterized by inflammatory eosinophilic infiltration of skeletal muscle tissue, sometimes extending into the fascia or skin [1,2]. Based on the depth of tissue involvement, eosinophilic myopathies are subdivided into focal eosinophilic myositis (FEM), diffuse eosinophilic myositis (DEM), eosinophilic myofasciitis (EMF), and eosinophilic fasciitis (EF) [2]. The current case displayed distinct features of diffuse eosinophilic myositis accompanied by eosinophilic vasculitis, without cutaneous or fascial involvement. Such presentation is rare, as most published cases describe either isolated myositis or involvement of the fascia [1,3].

Clinically, our patient's initial manifestations; fever, hypereosinophilia, and proximal muscle weakness—correspond to the classic presentation reported in earlier literature [4]. However, eosinophilic vasculitis coexisting with idiopathic myositis has been rarely documented, with only a few reports describing such overlap in the absence of systemic eosinophilic granulomatosis or parasitic infection [5]. In contrast to the eosinophilic granulomatosis with polyangiitis (EGPA) described by Kokubu et al. [5], our patient lacked pulmonary, cutaneous, or neuropathic features, and serology for ANCA and other autoimmune markers was negative, further supporting a diagnosis of idiopathic eosinophilic myositis.

The diagnosis of eosinophilic myositis requires a combination of clinical, biochemical, electrophysiological, and histopathological findings [2,6]. Peripheral eosinophilia, elevated CPK levels, and EMG evidence of myopathic changes strongly suggest muscle involvement, but the definitive diagnosis relies on biopsy demonstrating eosinophilic infiltration [6,7]. In our patient, histopathology confirmed dense eosinophilic infiltrates with associated vasculitic changes and features of muscle fibre necrosis and regeneration hallmarks of inflammatory myopathy [7].

Compared with other reported cases, our diagnostic approach relied primarily on basic investigations and muscle biopsy, given the limited access to advanced imaging modalities such as MRI or PET-CT, which are frequently utilized in high-resource settings to delineate the extent of tissue involvement [6]. Despite these constraints, the diagnostic process was methodical, involving exclusion of parasitic, autoimmune, and neoplastic causes of eosinophilia through comprehensive laboratory evaluation. This approach mirrors the diagnostic strategies described in resource-rich contexts [1,2,5], yet it underscores that accurate diagnosis is achievable in low-resource environments through careful clinical reasoning and targeted investigations.

The early decision to pursue muscle biopsy was particularly valuable. In many tropical or resource-constrained regions, prolonged empiric antibiotic or antiparasitic treatment is often initiated when eosinophilia and systemic symptoms coexist. In this case, prompt biopsy avoided unnecessary therapy and directed management appropriately, illustrating an important lesson for clinicians in similar contexts.

Management of eosinophilic myositis is not standardized due to the rarity of the condition, but most case reports and reviews advocate high-dose corticosteroids as the initial therapy [1,9]. Corticosteroids typically induce rapid symptomatic and biochemical improvement by suppressing eosinophilic inflammation. Our patient received pulse intravenous methylprednisolone followed by an oral tapering regimen, consistent with previous reports by Fermon et al. [1] and Choi et al. [10].

However, prolonged corticosteroid monotherapy often leads to relapse and adverse effects, necessitating adjunctive immunosuppressive therapy. Methotrexate, azathioprine, and mycophenolate mofetil are commonly used as steroid-sparing agents [9,10]. In our case, methotrexate was selected due to its proven efficacy in idiopathic inflammatory myopathies, affordability, and established safety profile in Sri Lankan practice. The combination achieved sustained clinical remission, normalization of CPK, and resolution of eosinophilia within six months—comparable to outcomes reported in prior case series [9,10].

Strengths of management in this case include: early initiation of immunosuppressive therapy following histological confirmation, avoiding delays common in resource-limited setups; successful use of an accessible and low-cost regimen (prednisolone + methotrexate) that achieved durable remission and close monitoring of liver function and CPK to guide treatment safely despite limited access to advanced monitoring tools. Limitations, on the other hand, included: lack of MRI evaluation to quantify the extent of disease involvement; absence of cytokine or interleukin (IL-4, IL-5) assays, which can provide insights into disease activity in high-resource settings and unavailability of biologic therapies such as anti–IL-5 agents, which have shown efficacy in refractory eosinophilic myositis [9].

Despite these limitations, the patient's favorable clinical outcome underscores that effective disease control can be achieved with conventional therapy, reinforcing the value of early recognition and pragmatic management in constrained environments.

Eosinophilic myositis poses particular diagnostic and therapeutic challenges in resource-limited countries such as Sri Lanka, where laboratory infrastructure, imaging facilities, and access to biologics are often limited. This case emphasizes that a structured, stepwise diagnostic strategy, focusing on exclusion of secondary causes and reliance on histopathology can yield accurate diagnosis even without sophisticated tools. Moreover, affordable and widely available immunosuppressive agents such as methotrexate remain viable therapeutic options that can provide excellent long-term outcomes.

The broader implication for clinical practice is the importance of maintaining a high index of suspicion for idiopathic eosinophilic myositis in patients presenting with fever, myalgia, proximal muscle weakness, and persistent eosinophilia, particularly when common causes such as parasitic infections have been excluded. In such contexts, early biopsy and initiation of corticosteroid therapy can be life- and function-saving.
While this case demonstrates successful management, further research and case accumulation are needed to better define prognostic factors, optimal treatment duration, and the potential role of biologic therapy in refractory disease. Establishing regional or national registries for rare inflammatory myopathies could help delineate epidemiological patterns and outcomes in South Asian populations. Collaborative efforts between clinicians and pathologists will be critical to improve recognition and standardization of diagnostic and therapeutic protocols in eosinophilia-associated myopathies.

CONCLUSION

This case reinforces the importance of thorough diagnostic evaluation and awareness of rare eosinophilic myopathies in patients with fever, hypereosinophilia, and proximal muscle weakness. The coexistence of eosinophilic vasculitis and myositis, in the absence of systemic involvement, is unusual and expands the phenotypic spectrum of idiopathic eosinophilic myositis. The patient's favorable response to corticosteroids and methotrexate highlights the efficacy of conventional immunosuppression and demonstrates that even in resource-limited environments, careful clinical reasoning and judicious therapy can lead to excellent outcomes.

Author declaration

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Conceptualisation and Design: TCP, BS, SM, UD; Manuscript drafting: TCP, BS, SM, UD; Study supervision: SM, UD.

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Written informed consent was obtained from the patient before they participated in the case, ensuring their understanding of the purpose, procedures, and potential implications involved.

Statement on data availability:
All data generated during this study are included in this published article.

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