Eosinophilic Polymyositis: A Rare Cause of Atraumatic Compartment Syndrome of the Forearm

Michael K. Boyajian^{1,2} Lauren O. Roussel^{1,2} Galam Khan^{1,3} Edward Stopa^{1,3} Reena Bhatt^{1,2}

¹Warren Alpert Medical School of Brown University, Providence, Rhode Island, United States

²Division of Plastic and Reconstructive Surgery, Brown University, Providence, Rhode Island, United States

³Division of Neuropathology, Rhode Island Hospital, Brown University, Providence, Rhode Island, United States

Address for correspondence Michael K. Boyajian, BA, The Warren Alpert Medical School of Brown University, 222 Richmond Street, Providence, RI 02903, United States (e-mail: michael_boyajian@brown.edu).

J Hand Microsurg:2020;12(suppl S1):S67–S69

Abstract

Background Eosinophilic polymyositis is a rare disorder in which eosinophils infiltrate muscle and supporting connective tissue structures, resembling autoimmune or immunologically mediated disease. This disorder can be associated with muscle inflammation and death, and can be a cause of atraumatic compartment syndrome.

Methods This is a retrospective chart review of a case report as well as review of pertinent literature.

Results This report presents a rare case of atraumatic compartment syndrome of the forearm caused by eosinophilic polymyositis. It provides a case summary and histological examination of this patient.

Conclusion This is an important case to report because it highlights eosinophilic polymyositis as a unique etiology of compartment syndrome. In appropriate clinical situations where patients do not improve despite standard interventions, one should consider the rare and unusual etiology of compartment syndrome due to eosinophilic polymyositis. Furthermore, primary surgical intervention should not be delayed while waiting to ascertain a definitive diagnosis.

Keywords

- hand surgery
- compartment syndrome
- myositis

Introduction

Acute compartment syndrome occurs when intrafascial pressures within a closed osteofascial compartment exceed perfusion pressures.¹ Without decompression, there is a high risk for ischemia and necrosis of the affected tissues.² Cases of compartment syndrome associated with eosinophilic polymyositis are extremely rare, and there are few literature sources that report the surgical management of this disease.

Case Report

We describe the case of a 62-year-old man who presented with a 1-week history of left forearm pain and swelling. The patient denied any antecedent trauma. History was notable for recently worsening myalgia in his bilateral

lower extremities and back, which precipitated his visit to the emergency department. Medical history is remarkable for cutaneous T cell lymphoma and a remote history of juvenile idiopathic arthritis. There was nontender, focal swelling and erythema of his left volar forearm, minimal pain with passive and active movement in the upper extremities, and intact neurovascular exam. Lower extremity exam was notable for mild tenderness along anterior thighs bilaterally with the remainder of his clinical exam being unremarkable. Laboratory tests at the time of presentation were significant for creatine kinase (CK) level elevation (7,886 U/L), white blood cell (WBC) count within the normal range at 6.4 with an abnormal predominance of eosinophils (19.3%), erythrocyte sedimentation rate elevation (32 mm/h), and C-reactive protein elevation (114.1 mg/L). The patient was admitted for serial monitoring and treatment of presumed cellulitis

to the medicine service. The forearm swelling was initially managed conservatively with a Carter pillow for elevation and gentle ace wrap compression. The patient's CK progressively increased to a maximum level of 10,177 U/L. Given the persistent increase in CK level and physical exam notable only for forearm swelling and erythema, intracompartmental pressures were obtained. This was done using a handheld manometer. Resting direct intracompartment pressures were 33 mm Hg of the volar mid-forearm and 17 mm Hg of the dorsal compartment, with a diastolic pressure of 70 mm Hg.

The patient was brought to the operating room for forearm exploration and fasciotomies given increased intracompartmental pressures in the setting of persistently elevating CK levels and swelling that did not improve with intravenous (IV) antibiotics, extremity elevation, or gentle compression. Right volar and dorsal forearm fasciotomies were performed. Pale, grayish appearing muscle fibers with associated bulging of muscles were noted in the superficial flexor compartment, which were debrided and sent for culture. The deep volar and dorsal compartments appeared grossly normal. Histologic analysis of the muscle biopsy performed intraoperatively revealed evidence of acute muscle necrosis accompanied by marked inflammation, with infiltration of numerous eosinophils, polymorphonuclear leukocytes, lymphocytes, and plasma cells consistent with eosinophilic polymyositis as the underlying pathology (►Fig. 1A, B). In addition, C5b-9 attack complex immunohistochemical stain was performed on sections obtained from the formalin-fixed paraffin-embedded tissue block after specimen processing. This stain is routinely used to demonstrate evidence of complement activation in association with the inflammatory reaction. This C5b-9 attack complex staining reveals robustly positive fibers (►Fig. 2). Postoperatively, the patient's CK levels and eosinophilia were normalized.

Discussion

Acute compartment syndrome occurs when the pressure in a closed fascial space rises to a point where perfusion to that compartment is compromised.^{1,3} This is a surgical emergency,

and it is most frequently caused by long bone fractures, crush injuries, or severe trauma.^{2,3} Atraumatic compartment syndrome is extremely rare. It can occur in the setting of IV drug use, anticoagulation, animal bites, or venous cannulation.³ In rare cases, it can also be caused by eosinophilic myositis.

Eosinophilic myositis consists of three clinically distinctive and pathologically heterogeneous diseases that include focal eosinophilic myositis, eosinophilic perimyositis, and eosinophilic polymyositis.⁴ All are characterized by eosinophilic infiltration of skeletal muscles or, in cases where eosinophils are absent, by major basic protein released by eosinophils.⁴ Laboratory findings typically show eosinophilia and elevated serum levels of CK. Focal eosinophilic myositis commonly presents with focal and circumscribed involvement of the lower legs, while eosinophilic perimyositis more commonly presents with myalgia and some skin abnormalities.⁴ Diagnostic criteria for eosinophilic polymyositis include proximal weakness affecting limb-girdle muscles, myonecrosis, and deep infiltration of eosinophils into muscle.⁴ Additional diagnoses to be considered include muscle dystrophies, systemic autoimmune diseases, infectious etiologies, morphea profunda, flexor tenosynovitis, and necrotizing fasciitis.⁴⁻⁶ Distinction

Fig. 2 C5b-9 attack complex staining reveals robustly positive fibers (arrows).

Fig. 1 (A, B) Hematoxylin and eosin section shows widespread evidence of acute muscle fiber necrosis with a marked inflammatory infiltrate composed of numerous eosinophils (arrows), polymorphonuclear leukocytes, plasma cells, and lymphocytes.

from other diseases is critical because eosinophilic polymyositis, while an exceptionally rare disorder, is potentially lethal as it can be accompanied by systemic organ involvement.⁷

As eosinophils degranulate, histologic examination of a patient with possible eosinophilic polymyositis should ideally include immunostaining for extracellular eosinophil major basic protein, whose diffusion is considered a hallmark for eosinophilic cytotoxicity.⁸ Eosinophilic infiltration of muscle and supporting connective tissue structures can resemble autoimmune or immunologically mediated disease. Activation of eosinophils is followed by the release of cytotoxic major basic protein, which can, in turn, further exacerbate local muscle injury.

It should also be noted that during the forearm fasciotomies, the surgeon noted pale, grayish appearing fibers overlaying the muscles in the superficial flexor compartment. Similar fibrinous exudates have been reported in the few published cases where eosinophilic infiltration progressed to compartment syndrome and required operative intervention.^{1,2} Thus, in future cases, where patients present with increased intracompartmental pressures, elevated CK, and eosinophilia, the intraoperative finding of superficial myonecrosis may serve as a diagnostic clue for eosinophilic polymyositis. Interestingly, our patient described mild but worsening pain but never described the severe discomfort associated with classic compartment syndrome.

Overall, this is an important case to report because it highlights eosinophilic polymyositis as a unique etiology of compartment syndrome. In appropriate clinical situations where patients do not improve despite standard interventions, one should consider the rare and unusual etiology of compartment syndrome due to eosinophilic polymyositis. Furthermore, primary surgical intervention should not be delayed while waiting to ascertain a definitive diagnosis. Although rare, surgeons should be aware of the potential for this uncommon cause for a common problem, and the known potential for associated multiorgan involvement

should also be considered. Inclusion of full-thickness biopsy during the fasciotomy procedure is critical to achieving an accurate diagnosis of eosinophilic polymyositis. As histologic eosinophilic infiltration is not present in all cases as eosinophilic degranulation may have already occurred, histologic examination should ideally include immunophenotypic staining for extracellular major basic protein to help confirm the diagnosis.

Conflict of Interest

None declared.

References

- 1 Nassar M, Jalil W, Isur A. Unilateral compartment syndrome secondary to eosinophilic fasciitis. *Plast Surg Case Studies* 2016;2(2):37–39
- 2 Shivakumar CN, Mani NJ, Vinod V. Atraumatic acute compartment syndrome of forearm due to eosinophilic myositis. *Kerala J Orthop* 2012;25:96–99
- 3 Smith K, Wolford RW. Acute idiopathic compartment syndrome of the forearm in an adolescent. *West. J Emerg Med* 2015;16(1):158–160
- 4 Selva-O'Callaghan A, Trallero-Araguás E, Grau JM. Eosinophilic myositis: an updated review. *Autoimmun Rev* 2014;13(4–5):375–378
- 5 Weinstock A, Green C, Cohen BH, Prayson RA. Becker muscular dystrophy presenting as eosinophilic inflammatory myopathy in an infant. *J Child Neurol* 1997;12(2):146–147
- 6 Onajin O, Wieland CN, Peters MS, Lohse CM, Lehman JS. Clinicopathologic and immunophenotypic features of eosinophilic fasciitis and morphea profunda: a comparative study of 27 cases. *J Am Acad Dermatol* 2018;78(1):121–128
- 7 Ishizawa K, Adachi D, Kuboi K, et al. Multiple organ involvement in eosinophilic polymyositis: an autopsy report. *Hum Pathol* 2006;37(2):231–235
- 8 Cantarini L, Volpi N, Carbotti P, et al. Eosinophilia-associated muscle disorders: an immunohistological study with tissue localisation of major basic protein in distinct clinicopathological forms. *J Clin Pathol* 2009;62(5):442–447