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CASE REPORT

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Recurrent, atraumatic acute compartment syndrome secondary to IgA vasculitis: A case report

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Abstract

Acute compartment syndrome is a surgical emergency requiring rapid recognition in the emergency department to minimize morbidity and mortality. It is most commonly caused by traumatic extremity fractures, which account for about 75% of cases. Atraumatic acute compartment syndrome is substantially less common with current evidence mostly limited to case reports, and diagnosis is made more challenging by the absence of an obvious traumatic injury. We present the case of a young adult female patient with IgA vasculitis who developed recurrent, atraumatic acute compartment syndrome and was successfully managed with prompt fasciotomy. This is the first case of spontaneous intramuscular hemorrhage, a rare sequela of IgA vasculitis, leading to recurrent, atraumatic acute compartment syndrome. This case highlights the importance of both a thorough physical exam and maintaining a high suspicion for acute compartment syndrome in the absence of injury to ensure patients receive prompt surgical evaluation for definitive care.

KEYWORDS
acute compartment syndrome, case report, Henoch-Schönlein purpura, IgA vasculitis, intramuscular hemorrhage

1 | INTRODUCTION

IgA vasculitis, formerly known as Henoch-Schönlein purpura, is a rare systemic small-vessel vasculitis classically characterized by the triad of palpable purpura, arthritis, and abdominal pain.[1] Most commonly seen in children with a mean age of onset of 6 years, IgA vasculitis is typically a self-limited condition that infrequently follows a relapsing-remitting course.[1–3] While the pathophysiology of this vasculitis remains poorly understood, the crux of it involves the formation and deposition of immune complexes into the capillaries, venules, and arterioles of the skin, joints, gastrointestinal system, and in some cases, the kidneys, leading to IgA nephropathy.[1,4] Several criteria exist for the definition and classification of the disease, most notably the 2008 EULAR/PRINTO/PRES criteria; however, IgA vasculitis ultimately remains a clinical diagnosis.[4] Treatment in most cases is limited to supportive care, particularly analgesia.

Rarely, IgA vasculitis has been implicated in the development of acute compartment syndrome, and only two cases reports exist in the current literature: one triggered by arterial cannulation of the radial artery in a hospitalized 6-year-old male with previously diagnosed IgA vasculitis and another case of acute compartment syndrome in a 17-year-old female as an isolated, spontaneous complication of the disease.[5,6] Traumatic extremity fractures account for approximately 75% of cases, and the classical presentation of acute compartment syndrome is commonly remembered by the "6 P's": pain, paresthesias, poikilothermia, pallor, pulselessness, and paralysis.[7,8] Atraumatic

acute compartment syndrome is substantially less common with current evidence mostly limited to case reports, and diagnosis is made more challenging by the absence of obvious traumatic injury.[9] Here, we present the case of a young adult female with previously diagnosed IgA vasculitis who developed recurrent, atraumatic acute compartment syndromeACS, and was successfully managed with prompt fasciotomy.

2 | CASE

An adult female in her early 20s with IgA vasculitis and history of spontaneous intramuscular hemorrhage of the right lower leg leading to acute compartment syndrome requiring fasciotomies presented to the emergency department with a 5-day course of progressively worsening left hand and lower leg pain and swelling, left hand ecchymosis, and left foot numbness. Physical examination of the left upper extremity revealed diffuse ecchymosis extending from the dorsal and volar aspects of the hand to the proximal forearm, firm but compressible forearm compartments, limited flexion and extension of the digits secondary to pain and edema, and a 2+ radial pulse. Examination of the left lower extremity was significant for firm but compressible distal anterior foreleg compartments, diminished sensation of the foot, pain with passive dorsiflexion of the foot, and a 1+ dorsalis pedis pulse. Initial laboratory studies, which included a complete blood count, comprehensive metabolic panel, coagulation studies, and creatinine kinase level, were unrevealing.

The patient was admitted for concern for acute compartment syndrome and underwent a four-compartment fasciotomy of the left lower leg within 8 hours of her initial presentation to the emergency department, revealing for edema in three of the four compartments and intramuscular hematomas in the anterior and superficial posterior compartments. The discovery of the intramuscular hematoma in the anterior compartment was what confirmed the need for fascial release. She returned to the operating room 2 days later for washout, closure of the medial fasciotomy site wound, and partial closure of the lateral wound with placement of a wound vacuum device. Autograft to that lateral site was performed 9 days after the initial fasciotomies. Following consultation with hand surgery during the initial emergency department encounter, the patient's left hand was successfully treated with elevation and compression wrapping. She remained admitted for pain control and management of her wound vacuum device and was ultimately discharged 20 days after her initial presentation.

3 | DISCUSSION

Spontaneous intramuscular hemorrhage is a rare sequela of IgA vasculitis, and this is the first case of it leading to recurrent, atraumatic acute compartment syndrome.[10,11] The pathophysiology of IgA vasculitis and resultant spontaneous intramuscular hemorrhage is not fully understood, and like other vasculitides, an inciting event cannot always be identified. Although in IgA vasculitis, preceding respiratory infections have been associated with its onset.[12] In general, however, the current literature suggests that immune complex deposition in small blood vessels leads to endothelial damage and fibrinoid necrosis of the vessel walls.[13] This creates the opportunity for blood to extravasate into the interstitial space, which is the basis for the development of superficial purpura, and more rarely, deeper intramuscular hematomas and compartmental edema. Increased pressure in the compartments from these hematomas can ultimately result in acute compartment syndrome, as in this case.

More research is necessary to determine the minutiae of how acute compartment syndrome secondary to underlying vasculitis occurs and can be prevented. Currently, treatment of the more common, self-limited cases of IgA vasculitis is limited to analgesia with nonsteroidal anti-inflammatory drugs. For recurrent or chronic disease, as in this patient, there is a lack of randomized-controlled trials to guide management of these patients. The SHARE (Single Hub and Access point for pediatric Rheumatology in Europe) initiative has published some consensus-based recommendations for treatment of these recalcitrant cases.[14] Of note, the use of corticosteroids should be considered when the course of disease is complicated by orchitis, cerebral vasculitis, pulmonary hemorrhage, or severe gastrointestinal involvement. Cytotoxic immunosuppressants may be necessary when organ- or life-threatening involvement occurs. The question remains as to whether corticosteroid or immunosuppressive treatment could have prevented this recurrent case of spontaneous acute compartment syndrome.

As it pertains to the emergency department, physicians should maintain a high index of suspicion for acute compartment syndrome in individuals with extremity pain and swelling even when traumatic injury is absent. Practically, this means ensuring a complete physical exam of the affected extremity is performed to assess for unequal pulse strength, temperature and skin color differences, neurological deficits, range of motion, and pain out of proportion to the exam. When atraumatic acute compartment syndrome is identified, vasculitides should be considered as potential etiologies, and a respective family history of such conditions should be obtained. It is through this high index of suspicion for acute compartment syndrome that surgical evaluation for definitive treatment via fasciotomy can occur in a timely manner.

CONFLICT OF INTEREST STATEMENT

The authors declare no conflict of interest.

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REFERENCES

1. Reamy BV, Servey JT, Williams PM. Henoch-Schönlein purpura (IgA vasculitis): rapid evidence review. *Am Fam Physician*. 2020;102(4):229-233.
2. Van de Perre E, Jones RB, Jayne DRW. IgA vasculitis (Henoch-Schönlein purpura): refractory and relapsing disease course in the adult population. *Clin Kidney J*. 2021;14(8):1953-1960. doi10.1093/ckj/sfaa251
3. Alfredo CS, Nunes NA, Len CA, Barbosa CM, Terreri MT, Hilário MO. Henoch-Schönlein purpura: recurrence and chronicity. *J Pediatr (Rio J)*. 2007;83(2):177-180. doi 10.2223/JPED.1595

4. Pillebout E, Sunderkötter C, vasculitis IgA. IgA vasculitis. *Semin Immunopathol*. 2021;43(5):729-738. doi 10.1007/s00281-021-00874-9
5. Luis GE, Ng ES. Acute compartment syndrome of the hand in Henoch-Schonlein purpura. *J Med Case Rep*. 2007;1:6. doi 10.1186/1752-1947-1-6
6. Botero-Bermúdez M, Briceño-Rodríguez L, Quiñonez-Yepes T, Taborda-Aitken JC. Síndrome compartimental: una presentación atípica como causa de la púrpura de Henoch-Schönlein [Compartment syndrome: an atypical presentation as a cause of Henoch-Schönlein purpura. *Acta Ortop Mex*. doi 2022;36(5):324-328.
7. Mauser N, Gissel H, Henderson C, Hao J, Hak D, Mauffrey C. Acute lower-leg compartment syndrome. *Orthopedics*. 2013;36(8):619-624.doi 10.3928/01477447-20130724-07
8. Pechar J, Lyons MM. Acute compartment syndrome of the lower leg: a review. *J Nurse Pract*. 2016;12(4):265-270. doi 10.1016/j.nurpra.2015.10.013
9. Cara JA, Narváez A, Bertrand ML, Guerado E. Acute atraumatic compartment syndrome in the leg. *Int Orthop*. 1999;23(1):61-62. doi 10.1007/s002640050306
10. Azer S, File W, Leazer R. Intramuscular hematoma as a manifestation of IgA vasculitis. *Pediatrics*. 2020;145(5):e20191668. doi 10.1542/peds.2019-1668
11. Mahevas M, Makdassi R, Presne C, et al. Hématome intramusculaire sévère au cours du purpura rhumatoïde [Muscular haematoma in Henoch-Schönlein purpura]. *Rev Med Interne*. 2004;25(12):927-930. doi 10.1016/j.revmed.2004.08.004
12. Thomas AE, Baird SF, Anderson J. Purpuric and petechial rashes in adults and children: initial assessment. *BMJ*. doi 2016;352:i1285. 10.1136/bmj.i1285
13. Bérubé MD, Blais N, Lanthier S. Neurologic manifestations of Henoch-Schönlein purpura. *Handb Clin Neurol*. 2014;120:1101-1111. doi:10.1016/B978-0-7020-4087-0.00074-7
14. Ozen S, Marks SD, Brogan P, et al. European consensus-based recommendations for diagnosis and treatment of immunoglobulin A vasculitis-the SHARE initiative. *Rheumatology (Oxford)*. 2019;58(9):1607-1616. doi 10.1093/rheumatology/kez041

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