

## Case Report

# Multifocal recurrent atraumatic acute compartment syndrome in an adolescent

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## ABSTRACT

Recurrent, atraumatic acute compartment syndrome (ACS) is an uncommon and poorly understood phenomenon. We report a female in her teens who experienced two distinct episodes of atraumatic ACS. The first episode involved the right lower extremity, and the second, the right upper extremity. Each case required emergent fasciotomy and involved postoperative complications, requiring repeat surgical intervention and vacuum-assisted closure. Extensive multidisciplinary evaluation, including rheumatologic, hematologic, and genetic testing, failed to identify an etiology. Given the recurrence and localization to separate limbs, a focal vascular abnormality is suspected, supported by comparison with a similar reported case successfully treated with angiographic embolization. This case underscores the importance of prompt surgical intervention and multidisciplinary evaluation and highlights the potential role of vascular malformations in cases of unexplained compartment syndrome.

**Keywords:** Compartment syndrome, Atraumatic, Recurrent, Fasciotomy, Vascular malformation, Adolescent

## INTRODUCTION

Compartment syndrome is a condition defined by increased pressure within a closed osteofascial compartment to a level that compromises the microcirculation of the tissues contained within that space.<sup>1</sup> It results from either a reduction in compartment volume or an increase in the volume of its contents. Both mechanisms lead to arteriolar compression and subsequent impairment of tissue perfusion. Sustained hypoperfusion can rapidly result in cellular death within the affected compartment, with skeletal muscle being particularly susceptible to ischemic injury.<sup>2</sup> If left untreated, compartment syndrome may produce significant morbidity or mortality. The diagnosis is primarily based on the clinical symptoms and the physical examination findings.<sup>3</sup>

The incidence of ACS is estimated at 7.3 per 100,000 in males and 0.7 per 100,000 in females.<sup>4</sup> Most cases occur secondary to traumatic injury, including high-energy mechanisms, fractures, or constrictive forces. Atraumatic compartment syndrome is rare, with existing evidence largely limited to case reports.<sup>5</sup> Recurrent atraumatic compartment syndrome has been described previously, both secondary to systemic vasculitis and as an idiopathic process confined to a single limb.<sup>12,13</sup> To our knowledge, however, this represents first reported case of recurrent, atraumatic compartment syndrome affecting anatomically distinct regions in separate extremities in a patient with a comprehensively negative systemic evaluation.

## CASE REPORT

A case of an otherwise healthy adolescent female who experienced two anatomically distinct episodes of

atraumatic was reported. ACS. The patient had no significant past medical history and was physically active, participating in competitive athletics and resistance training. She denied tobacco, alcohol, or illicit substance use. Family history was notable for Legg-Calvé-Perthes disease in first-degree relatives, scoliosis, epilepsy, and aortic valve insufficiency.

The initial episode occurred nocturnally while the patient was supine at rest. She described a progressive sensation of deep, expanding pressure localized to the right lower extremity. At onset, she attributed the symptoms to delayed-onset muscle soreness following exercise. Over several hours, the pressure evolved into severe pain disproportionate to activity and progressively intensified. Over the subsequent 48 hours, the affected extremity developed increasing swelling and escalating pain. By day four, the patient was unable to ambulate secondary to pain and reported new-onset paresthesias (Table 1).

**Table 1: Clinical timeline.**

| Timepoints                      | Clinical Course                                                        |
|---------------------------------|------------------------------------------------------------------------|
| <b>Year 0 (Initial episode)</b> | Progressive atraumatic right lower extremity pain and swelling at rest |
| <b>Day 4</b>                    | Inability to ambulate; MRI confirmed ACS                               |
| <b>Same admission</b>           | Urgent fasciotomy performed                                            |
| <b>POD 1</b>                    | Postoperative edema and clot formation → repeat fasciotomy + NPWT      |
| <b>Months later</b>             | Completed rehabilitation; full functional recovery                     |
| <b>Year 4</b>                   | Recurrent atraumatic ACS involving right forearm during air travel     |
| <b>Same day</b>                 | Emergent fasciotomy                                                    |
| <b>Postoperative course</b>     | Wound dehiscence → repeat fasciotomy + NPWT                            |
| <b>Subsequent surgery</b>       | Third intervention with incision extension + additional NPWT           |
| <b>4 months later</b>           | Completed occupational therapy; full recovery                          |
| <b>Post-recovery</b>            | Extensive systemic and genetic workup negative                         |
| <b>2024</b>                     | Angiography recommended; not performed                                 |

Initial evaluation at an urgent care center included plain radiographs and ultrasonography. She was referred to the emergency department for further evaluation. Magnetic resonance imaging (MRI) demonstrated findings consistent with ACS of the right lower extremity. The patient underwent urgent fasciotomy. On postoperative day one, she developed significant postoperative edema and clot formation at the fasciotomy site and underwent repeat fasciotomy with placement of a negative-pressure wound therapy (NPWT) device. Following several months of structured physical therapy, the patient regained full

strength and function in the right lower extremity, with no residual neurologic deficit.

Four years following the index event, the patient experienced a second episode while traveling on an aircraft. During flight, she noted progressive pressure along the lateral aspect of the right forearm. Within hours, visible swelling developed, accompanied by rapidly escalating pain. Upon arrival in Europe, she was transported directly to the emergency department, where she was again diagnosed with ACS of the right forearm. She underwent emergent fasciotomy. Her postoperative course was complicated by marked soft-tissue swelling, resulting in mechanical failure of staple closure and wound dehiscence. She subsequently underwent repeat fasciotomy with placement of NPWT. A third operative intervention was later required, during which the prior fasciotomy incision was extended, and an additional wound VAC was applied. After four months of intensive occupational therapy, she achieved full recovery of upper extremity function.

Following recovery from the second episode, an extensive workup was undertaken to evaluate for underlying etiologies of recurrent atraumatic ACS. Comprehensive hematologic, metabolic, and connective tissue evaluations revealed no abnormalities. Targeted exome sequencing of genes associated with metabolic myopathies, structural myopathies, and connective tissue disorders-including GYG1, PYGM, RYR1, COL1A1, COL1A2, COL3A1, COL5A1, and LMNA-demonstrated no pathogenic variants. Whole-genome sequencing of a 5.29 Mb region of homozygosity on chromosome 11 similarly revealed no clinically significant abnormalities.

In 2024, the patient was evaluated by an orthopaedic surgeon, who reported a separate case of recurrent, large, spontaneous hematomas ascribed to a small bleeding arterial branch arising from the lateral femoral circumflex artery. In that case, angiographic identification and embolization of the bleeding vessel resulted in definitive resolution. Given the clinical parallels and absence of alternative etiologies, diagnostic angiography was recommended for our patient to evaluate for an occult vascular malformation or an anomalous arterial branch. However, this study was not conducted due to insurance-related barriers.

**Differential diagnosis**

This patient was diagnosed with atraumatic ACS on two separate occasions. The underlying etiology remains under investigation. During the initial episode, the patient’s symptoms were initially attributed to an acute musculotendinous strain of the calf. Classically, gastrocnemius strain presents with acute onset pain following forceful eccentric contraction, frequently accompanied by a perceptible defect, ecchymosis, and a reported “popping” sensation.<sup>6</sup> In this case, the absence of a precipitating eccentric load, the insidious progression of

pain, and the degree of progressive swelling and escalating pain rendered this diagnosis unlikely.

Deep vein thrombosis (DVT) was also considered, given calf pain and swelling. The patient's Wells score was 3, denoting a low clinical probability. Duplex venous ultrasonography demonstrated no evidence of thrombus. Upon presentation to the emergency department, the patient exhibited four of the six classic clinical findings associated with ACS—pain out of proportion to examination, pain with passive stretch, paresthesias, and tense compartments—consequently elevating compartment syndrome to the leading diagnosis. MRI demonstrated findings consistent with compartmental edema and elevated intracompartmental pressure, confirming the diagnosis.

Following the second atraumatic episode, the investigation centered on possible systemic or structural contributors to recurrent ACS. A bleeding diathesis, including haemophilia, was considered despite the absence of haemarthrosis, spontaneous ecchymosis, or prior bleeding history. Comprehensive hematologic evaluation, including coagulation studies, was unremarkable, effectively excluding an underlying coagulopathy. A heritable connective tissue disorder was also suspected because of the family history of Legg-Calvé-Perthes disease, scoliosis, and aortic valve insufficiency. However, rheumatologic and genetic evaluation revealed no pathogenic variants or clinically significant abnormalities, arguing against a connective tissue or rheumatologic etiology.

Finally, IgA vasculitis was considered in light of a previously reported case of recurrent atraumatic compartment syndrome associated with small vessel vasculitis.<sup>7,12</sup> IgA vasculitis characteristically presents with evident purpura, arthralgias, gastrointestinal involvement, and renal manifestations. This patient demonstrated none of these features, and hematologic evaluation was unremarkable, rendering this diagnosis unlikely.

### **Outcome and follow-up**

Following surgical management and completion of an extensive course of structured physical therapy, the patient achieved full functional recovery and returned to her baseline. She has successfully resumed participation in athletic activities and continues to maintain a physically active lifestyle without limitation. Since the second episode, there have been no further recurrences of ACS. The patient remains otherwise healthy, with no residual deficits or interval medical concerns.

## **DISCUSSION**

ACS usually results from trauma. In contrast, atraumatic ACS is rare and mostly described in isolated case reports. This case describes a healthy adolescent female with

recurrent, anatomically separate atraumatic ACS. Hematologic, rheumatologic, and genomic evaluations were unrevealing. The recurrence, affecting different extremities over 4 years, differs from previous single-limb cases and prompts key orthopaedic considerations.

The key pathophysiology in ACS is a rise in intracompartmental pressure (ICP). This pressure impairs the microvascular gradient required for capillary perfusion.<sup>1</sup> As tissue pressure nears diastolic pressure, capillary blood flow depends on pressure, leading to reduced arteriovenous pressure gradient, endothelial dysfunction and increased capillary permeability, progressive interstitial edema, and further ICP elevation within a noncompliant fascial envelope. This positive feedback loop leads to local ischemia, myonecrosis, and neuropraxia.<sup>2</sup>

In atraumatic ACS, the absence of obvious injury makes it less clear what begins the expansion. Proposed mechanisms include spontaneous intramuscular hemorrhage, small-or medium-vessel vasculitis, exertional muscle edema in metabolic or structural myopathies, and focal arterial branch hemorrhage or vascular malformation. In this patient, recurrence in separate compartments—the lateral leg followed years later by the forearm—makes a localized fascial compliance problem unlikely. Postoperative findings during the first episode included significant edema and clot formation, raising suspicion for a haemorrhagic component not due to systemic coagulopathy. Standard coagulation panels and genetic analysis did not reveal a cause, making focal vascular pathology a plausible yet untested explanation.

Current orthopaedic guidelines state that ACS is primarily a clinical diagnosis. The most sensitive early finding remains pain out of proportion to examination and pain with passive stretch. The remaining “5 P’s” represent late manifestations and should not be waited for. Measuring pressure inside the muscle is helpful if the diagnosis is unclear. In this case, diagnosis of the first episode relied on clinical findings and MRI evidence of compartmental edema. MRI can highlight diffuse T2 hyperintensity in affected compartments, but its role is auxiliary and should not delay urgent surgery in clear cases.

For recurrent atraumatic ACS, diagnosis should go beyond routine musculoskeletal evaluation. After emergency fasciotomy and recovery, clinicians should systematically rule out underlying systemic causes, including coagulation abnormalities, vasculitis, and metabolic or structural myopathies. In cases of recurrence or unexplained etiology, CT angiography or catheter-based angiography should be strongly considered, as diagnostic angiography may have both diagnostic and therapeutic value if a vascular source is identified.

Both episodes were managed with emergent fasciotomy. The need for repeat surgical intervention and NPWT in both episodes illustrates the magnitude of postoperative

edema. In spite of these complications, the patient ultimately achieved full functional recovery following structured rehabilitation in both extremities. This favorable outcome reinforces the notion that timely decompression remains limb-salvaging, even in atypical presentations.

### ***Comparison with previously reported cases***

Placing this case alongside the published literature clarifies both what it shares with earlier reports and where it departs from them. In a systematic review of nontraumatic ACS of the upper extremity, Ogrodnik et al. found that infection, systemic anticoagulation, and inherited bleeding disorders accounted for the majority of reported cases, with a smaller group remaining unexplained after complete evaluation.<sup>11</sup> Our patient belongs to that smaller group: there was no infection, anticoagulation was never administered, and coagulation testing was normal on two separate occasions.

The closest published parallels are few. Ebeling and Hudson described recurrent atraumatic ACS in a young woman with IgA vasculitis, in whom spontaneous intramuscular haemorrhage was the presumed mechanism and the underlying vasculitis supplied a unifying explanation for both episodes.<sup>12</sup> Shiraz et al reported a 19-year-old woman with repeated episodes of forearm ACS over several years in whom extensive investigation, including CT angiography of the upper limb, identified no cause.<sup>13</sup> Dalton et al described acute bilateral spontaneous forearm compartment syndrome occurring without antecedent injury.<sup>14</sup> The present case differs from each of these. Unlike the patient described by Ebeling and Hudson, no systemic inflammatory disorder was identified here despite directed testing. Unlike the patient reported by Shiraz et al., in whom recurrence was confined to a single limb, our patient's episodes involved anatomically separate compartments in different extremities and were separated by four years of complete, symptom-free function. Widely spaced recurrence across distinct anatomic regions in a fully investigated patient does not correspond to any report we were able to identify.

### ***Interpretation and the vascular hypothesis***

Any proposed mechanism must account for two features of this case: the anatomic separation of the two episodes, and the intraoperative finding of clot within the decompressed compartment during the first event. A diffuse systemic process, such as a vasculitis or a coagulopathy, would explain multifocality but is difficult to reconcile with an entirely negative rheumatologic and hematologic evaluation and with the absence of any systemic manifestation over more than four years of observation. Conversely, a purely local abnormality, such as reduced fascial compliance, would explain focality but not the later involvement of a distant compartment in a different limb.

A multifocal vascular abnormality accommodates both features. Vascular anomalies of the extremities are frequently multiple, may remain clinically silent until a bleeding event occurs, and are characteristically invisible on the non-contrast imaging and laboratory studies that constitute a standard ACS evaluation. Compartment syndrome arising from an occult arterial branch lesion is well documented: Arbeloa-Gutierrez et al described a false aneurysm of the lateral circumflex femoral artery producing thigh compartment syndrome that was identified only on angiography and treated definitively by embolization.<sup>15</sup> This closely mirrors the comparison case described to our patient in 2024, in which recurrent spontaneous hematomas arising from a small branch of the lateral femoral circumflex artery resolved after angiographic embolization. Taken together, these reports indicate that such lesions are treatable, but that they are detectable only by contrast vascular imaging. In our patient the hypothesis remains untested, as angiography was deferred for reasons unrelated to clinical judgement.

Exertional physiology merits separate consideration given the patient's athletic history and resistance training. Livingston et al described acute exertional compartment syndrome in young athletes, a population our patient resembles, and observed that a proportion present without an identifiable acute injury.<sup>16</sup> Acute exertional compartment syndrome, however, characteristically follows an identifiable bout of intense activity. Our patient's first episode began at rest during sleep and the second during passive sitting on an aircraft, so neither presentation conforms to an exertional pattern.

### ***Diagnostic considerations in atraumatic presentations***

These comparisons bear directly on how such patients should be assessed. The epidemiologic series of McQueen et al from which commonly quoted incidence figures derive, described 164 patients in whom an associated fracture was present in 69% and in whom young men predominated.<sup>8</sup> Our patient matches neither the mechanism nor the demographic profile of that cohort, which is precisely why an atraumatic presentation is vulnerable to delayed recognition. Duckworth and McQueen have emphasized that no single physical finding is sufficiently sensitive in isolation, so that serial examination by the same observer carries greater diagnostic weight than any one assessment.<sup>9</sup> Where the clinical picture is equivocal or the patient cannot be examined reliably, intracompartmental pressure measurement is indicated; McQueen and Court-Brown demonstrated prospectively that a differential pressure of less than 30 mmHg below diastolic blood pressure identified all patients requiring decompression without missed cases, and this threshold remains the most widely adopted.<sup>10</sup> Because these thresholds derive from fracture-associated cohorts, their extension to atraumatic presentations has not been formally validated, and the present case illustrates that evidence gap.

Two limitations temper these conclusions. First, the vascular hypothesis advanced here rests on exclusion and on analogy with published reports rather than on direct imaging confirmation, and it must therefore be regarded as unproven. Second, intracompartmental pressures were not recorded during either episode, so the diagnosis rests on clinical findings supported by MRI. Neither limitation altered management, since decompression was indicated on clinical grounds in both instances, but both constrain the mechanistic inference that can be drawn.

## CONCLUSION

This case demonstrates recurrent, anatomically distinct atraumatic ACS in a healthy adolescent with negative systemic evaluation. The clinical course and absence of identifiable systemic pathology suggest that occult focal vascular abnormality represents plausible and under-recognized mechanism. For orthopaedic subspecialist, recurrent atraumatic ACS should prompt consideration of vascular imaging after standard etiologic exclusion. Early recognition and decisive surgical decompression remain the basis of a favourable functional outcome.

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