Compartment syndrome: a unique presentation

Zain Khalpeya, Christopher Grossa, Tec Chonga and Jonathan Gatesb

aDepartment of Surgery, Brigham and Women's Hospital, Harvard Medical School, Boston, MA, USA; bDepartment of Trauma, Burns, and Critical Care, Brigham and Women's Hospital, Harvard Medical School, Boston, MA, USA

Corresponding address: Zain Khalpey, Brigham and Women's Hospital (PB-B-4), 75 Francis Street, Boston, MA 02115, USA.
E-mail: zkhalpey@partners.org

Date accepted for publication 14 August 2008

Abstract

Compartment syndrome is a potentially limb- and life-threatening clinical entity resulting from elevated intra-compartmental pressures. A high clinical suspicion is paramount in diagnosis since full recovery is time-sensitive. We present a unique case of chronic myelomonocytic leukemia-induced (CMML) compartment syndrome which illustrates the importance of quick diagnosis and treatment.

Keywords

Compartment syndrome; chronic myelomonocytic leukemia (CMML); leukemia; unusual causes of compartment syndrome.

Case report

A 76-year-old male presented to his oncologist for a routine appointment with asymmetry of his left thigh with anterolateral thigh pain of less than 24 h. Concerned for a deep vein thrombosis, chloroma, or spontaneous hematoma his hematologist ordered a lower extremity ultrasound which was normal. A day later, he presented to the local emergency room (ED) with a sharp increase in thigh pain, specifically with flexion and extension at the knee joint. His family noted that he experienced chills and increasing confusion.

His past medical history was significant for chronic myelomonocytic leukemia (CMML), a myelodysplastic and myeloproliferative overlap syndrome manifesting as anemia and thrombocytopenia. His anemia was managed with Aranesp. The patient also had chronic renal insufficiency (eGFR of 29.9 ml/min per 1.73 m) but never required dialysis. Given his profession, he was exposed to and treated for Lyme disease twice. He frequently traveled to the developing world, his last trip being over one year ago.

On examination, he was alert, oriented, frail, and in no acute distress. He denied recent trauma and animal/insect bites. His initial temperature of 37.7°C (99.9°F) rose to 38.9°C (102.0°F). Other vital signs were within normal limits. His pertinent musculoskeletal exam findings were localized.
swelling and tenderness of the left upper thigh exacerbated by a motion at the left hip and knee joints. There was no evidence of joint effusion, tenderness or erythema in the left thigh or knee. His distal lower extremities had 2+ dorsalis pedal pulses and were warm and non-tender. No compartment pressures were measured. He was neurologically intact with good sensory and motor function, with the exception of limited leg motion due to pain. He did not note paresthesias.

His laboratory findings were significant for an elevated white cell count of 24.6 K/ml, a hematocrit of 24.7%, and a platelet count of 51,000/ml. His creatinine level was elevated from a baseline of 2.5 mg/dl to 4.3 mg/dl; his CK was normal at 209 U/l.

A plain X-ray of his left femur was unremarkable. A computer tomography scan revealed extensive subcutaneous soft tissue edema in the quadriceps muscle group, particularly the vastus lateralis muscle (Figs. 1 and 2). There was no evidence of fracture, air, interruption of the facial planes, or focal fluid collection to suggest other infective or inflammatory process. However, soft tissue stranding was present over the area of maximal tenderness.

Given the constellation of a chronic immunosuppressed state, fever, leukocytosis, and thigh tenderness, a diagnosis of necrotizing fasciitis was entertained. He was started on an antibiotic regimen of vancomycin, levaquin, and flagyl. The ED surgery team assessed the patient and promptly brought him to the operating room (OR) for urgent exploration of his thigh.

**Fig. 1.** Axial CT of mid-thigh. Diffuse swelling located near the vastus lateralis. Several non-specific findings are noted, including: some fat stranding, fascial plane effacement, and fluid. No evidence of gas or fracture.
In the OR, fasciotomy of the left lateral aspect of the left thigh was performed. The underlying muscle was edematous but bled appropriately on biopsy. There were patches of pale, necrotic tissue, without evidence of infection, upon opening the fascia. The vastus lateralis responded to electrical stimulation 30 min after decompression. The wound was left open with a VAC in place. Frozen section revealed a massive infiltration of mononuclear cells. Cultures taken at the incision did not grow any microorganisms. Final pathology correlated with the frozen specimen and showed extensive, diffuse infiltration of immature and mature monocytes consistent with the patient’s CMML.

On post-operative day 19, his fasciotomy site was closed and he was discharged three days later. A bone marrow biopsy performed two months afterwards revealed a greatly increased expansion of immature myeloid and myelomonocytic forms in hypercellular marrow.

**Discussion**

Compartment syndrome is a potentially life- and limb-threatening entity caused by increased intracompartmental pressure (normal values 0–10mmHg) usually secondary to ischemia and reperfusion injury. Not only is the literature scant in thigh compartment syndrome, but compartment syndrome secondary to CMML has not been described.

The causes for thigh compartment syndrome are speculative. The rich collateral flow of the lateral femoral circumflex artery and the superficial femoral arterial branches protects the thigh from a compromising vascular event. Since the thigh fascia fuses with the surrounding gluteal fascia, the compartment pressure may decompress into the hip compartment\(^{[1]}\). Additionally, the large volume of the thigh compartment raises the threshold pressure which could compromise the blood supply\(^{[1]}\).

---

*Fig. 2. Muscle biopsy, left quadriceps, thigh fascia (20x). Skeletal muscle, dense fibrous tissue, and adipose tissue show marked edema with a prominent mononuclear cell infiltrate. This infiltrate is comprised of intermediate-sized cells with irregularly folded nuclei, coarse to somewhat fine chromatin, prominent nucleoli, and a moderate amount of cytoplasm which is consistent with immature monocytic forms, including monoblasts, promonocytes, and mature monocytes.*
The pathophysiology of leukemia-induced compartment syndrome is multifactorial. The volume of the monocytic infiltrate cannot fully account for an acute increase in pressure. Disruption of the normal dynamics in interstitial fluid movement with muscle contraction by leukemic infiltrates may play a role. Furthermore, the addition of the leukemic infiltration may represent a tipping-point in the already-compromised compartment secondary to trauma, infection and/or venous thrombosis\(^\text{[2]}\).

Research has suggested that muscle injury and tissue necrosis occur at an interstitial pressure above 30 mmHg, although capillary perfusion is jeopardized at pressures greater than 20 mmHg\(^\text{[3]}\). Nerves are most sensitive to pressure, followed by muscle. Emergent surgery is usually reserved for pressures above 30 mmHg.

Irreversible tissue damage is inevitable after 12 h of clinical symptoms\(^\text{[3]}\). Although time-to-treatment is controversial, the golden period for complete recovery via decompression is less than 6 h. In our patient, the diagnosis of compartment syndrome was made in the OR. Fortuitously, there was no irreparable damage to the compartment since the ED team called for the appropriate surgical consultation resulting in a prompt trip to the OR. Appropriate treatment in previous leukemia-related case reports was uncommon because of delay in diagnosis; unfortunately, insult to the compartment was already permanent.

Clinical suspicion must be sensitive to: pain out of proportion to exam, paresthesia, paralysis, pain on passive motion, and pallor. Our patient had pain disproportionate to exam and pain on passive range of motion. Evidence shows that these clinical findings are not sensitive, but rather specific in diagnosing acute compartment syndrome, at 13–19% and 97–98%, respectively\(^\text{[4]}\). Therefore, the absence of the findings is more useful in ruling out the diagnosis. If the ED team were to suspect compartment syndrome, a quick way to measure the pressure would be a Stryker needle.

Chemotherapy and radiotherapy are contraindicated until fasciotomy site closure since underlying tissue edema could be exacerbated\(^\text{[2]}\). Unfortunately, chemotherapy is not an option in CMML since there have been no proven effective treatments to date. Closure of the fasciotomy site is performed after resolution of swelling and stabilization of the patient.

**Teaching point**

In conclusion, this case serves to highlight the existence of this rare occurrence of CMML-induced acute compartment syndrome. The gravity and acuity of the patient’s limb was quickly identified by the ED team, surgery was consulted, and the patient underwent an emergency fasciotomy. Despite the lack of history of trauma or other ischemic injury, it highlights the importance of having a strong clinical suspicion so that the neuromuscular compartments can be spared from permanent injury.

**Acknowledgements**

We would like to thank Dr Maria Alejandra Duran-Mendicuti for review of the radiographic findings and Dr Dick Hwang for his help in procuring the pathology slide.

**References**