Acute Pediatric Leg Compartment Syndrome in Chronic Myeloid Leukemia

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Abstract

Acute compartment syndrome is an orthopedic surgical emergency and may result in devastating complications in the setting of delayed or missed diagnosis. Compartment syndrome has a variety of causes, including posttraumatic or postoperative swelling, external compression, burns, bleeding disorders, and ischemia-reperfusion injury. Rare cases of pediatric acute compartment syndrome in the setting of acute myeloid leukemia and, even less commonly, chronic myeloid leukemia have been reported. The authors report the first known case of pediatric acute compartment syndrome in a patient without a previously known diagnosis of chronic myeloid leukemia. On initial examination, an 11-year-old boy presented with a 2-week history of progressive left calf pain and swelling after playing soccer. Magnetic resonance imaging scan showed a hematoma in the left superficial posterior compartment. The patient had unrelenting pain, intermittent lateral foot paresthesias, and inability to bear weight. Subsequently, he was diagnosed with acute compartment syndrome and underwent fasciotomy and evacuation of a hematoma. Laboratory results showed an abnormal white blood cell count of 440×10⁹/L (normal, 4.4-11×10⁹) and international normalized ratio of 1.3 (normal, 0.8-1.2). Further testing included the BCR-ABL1 fusion gene located on the Philadelphia chromosome, leading to a diagnosis of chronic myeloid leukemia. Monotherapy with imatinib mesylate (Gleevec) was initiated. This report adds another unique case to the growing literature on compartment syndrome in the pediatric population and reinforces the need to consider compartment syndrome, even in unlikely clinical scenarios.
A cute compartment syndrome is an orthopedic emergency and occurs as a result of prolonged compromise of capillary perfusion within an osteofascial compartment. It has a variety of causes, including posttraumatic or postoperative swelling or hemorrhage, external compression, burns, bleeding disorders, and ischemia-reperfusion injury. Fortunately, early diagnosis and surgical intervention with fasciotomy can result in tissue salvage and maintenance of function. Rare cases of acute compartment syndrome in the setting of hyperleukocytic acute leukemia and, even less commonly, chronic myeloid leukemia have been reported. Only 1 case of acute compartment syndrome in the context of chronic myeloid leukemia has been reported in the pediatric population. The authors report the first known case of pediatric acute compartment syndrome in a patient without a previously known diagnosis of chronic myeloid leukemia.

**Case Report**

An apparently otherwise healthy 11-year-old boy presented with a 2-week history of posterior left calf pain that began after he injured his left calf while playing soccer. Symptoms were initially tolerated with a regimen of activity modification and treatment with ibuprofen and acetaminophen. After the patient ran for several hours 2 days before presentation, however, symptoms acutely worsened and were no longer relieved by over-the-counter medications. Initial outside orthopedic evaluation was obtained, and the next day, magnetic resonance imaging scan of the left leg showed a 5.6×3×8.3-cm hematoma in the left superficial posterior compartment adjacent to the gastrocnemius muscle (Figure 1). The symptoms continued to worsen over the next day, including inability to ambulate and intermittent ankle and foot numbness. Therefore, the patient was taken to the emergency room. Orthopedic consultation was immediately obtained. On physical examination, the child was white and thin and appeared to be severely anxious and in significant distress, despite administration of 4 mm of intravenous morphine. He held the left knee flexed and draped over pillows, with the foot slightly plantar flexed. The proximal left calf was markedly swollen, and given his thin habitus, noncompressible firmness in the posterior leg compartments was readily appreciable with palpation. Passive ankle dorsiflexion and plantarflexion elicited severe pain, although passive toe range of motion was painless. On neurovascular examination, the patient reported paresthesias, which had become dense by this time, in the lateral foot in the sural nerve distribution. Otherwise, sensation was normal in all other nerve distributions. Motor strength was 5/5 in all motor distributions, except 4/5 in dorsiflexion and plantarflexion secondary to severe pain.

The patient was diagnosed with acute compartment syndrome involving the superficial posterior compartment and underwent emergent left leg fasciotomy. Preoperative laboratory evaluation unexpectedly showed several abnormalities,
including a white blood cell count of 440×10^9/L (normal, 4.4-11×10^9/L), international normalized ratio of 1.3 (normal, 0.8-1.2), prothrombin time of 13.1 seconds (normal, 10.2-12), partial thromboplastin time of 42 seconds (normal, 26-37), and lactate dehydrogenase level of 556 IU/L (normal, 50-175). Hematology and oncology were immediately consulted intraoperatively, and a presumptive diagnosis of chronic myeloid leukemia was made, based on the findings of a peripheral blood smear that showed leukocytosis with increased myeloid precursors and neutrophils (Figure 2).

In the operating room, a proximal medial skin incision was made. The superficial posterior compartment fascia was incised longitudinally in an extensile manner, with care taken to preserve the saphenous neurovascular structures. Large amounts of whitish-brown fluid and clot were expressed adjacent to the gastrocnemius and proximal soleus musculature. Once evacuated, the superficial posterior and other leg compartments were palpated and were easily compressible. Therefore, no other fasciotomies were performed. The affected compartment muscle tissue was determined to be contractile and was bleeding throughout, so minimal debridement was undertaken. After the wound was thoroughly irrigated and hemostasis was achieved, the fascia was left open and the skin was approximated. The patient was placed in a posterior splint postoperatively. Multiple samples of fluid from the affected compartment were sent for microbiologic and pathologic examination.

Further evaluation and bone marrow biopsy (Figure 3) examination by hematology and oncology showed multiple markers for chronic myeloid leukemia in the myeloid cells, including the BCR-ABL1 fusion gene located on the Philadelphia chromosome t(9;22)(q34;q11). Monotherapy was initiated with imatinib mesylate (Gleevec), a tyrosine kinase inhibitor, and the patient was monitored for tumor lysis syndrome, which did not develop. He ambulated by the next day, with only mild leg pain throughout the remainder of his hospitalization. The weakness and paresthesias also improved rapidly. The patient was discharged home on postoperative day 6.

The patient continues to receive Gleevec monotherapy, with a strong white blood cell count response from 440×10^9/L to 6.3×10^9/L. He is followed closely by oncology. At surgical follow-up 7 days after fasciotomy, mild serosanguineous drainage was reported at home, and a moderate amount of brownish clot was evacuated in the office. At 2-week follow-up, the wound was healed, with no signs of infection. The patient showed no sequelae of acute compartment syndrome of the leg on follow-up. Furthermore, final results of all intraoperative cultures were negative for infection.

**DISCUSSION**

Despite devastating complications, studies indicate that delayed diagnosis of acute compartment syndrome in the pediatric population is relatively common.7 Substantial ischemia occurs once intracompartamental tissue pressure increases to within 10 to 20 mm of diastolic blood pressure. Under such conditions, paresthesias develop within hours and irreversible nerve and muscle damage begins to occur within 6 to 8 hours.2,8

The diagnosis of acute compartment syndrome in the pediatric population may
not be as straightforward as in adults. The sensitive clinical hallmarks, excessive pain and narcotic requirements, in particular, may not be present. Up to 12% of pediatric patients may not report significant pain as a chief complaint, and children may be unable to provide consequential clinical information that may lead to early diagnosis. One study found that pediatric patients most frequently presented with paresthesias (66%), whereas a substantial percentage of others had late signs of paralysis and paresthesia (36% and 18%, respectively). Other signs, including restlessness, agitation, and anxiety, are increasingly recognized as potentially important clinical indicators in the pediatric population.

A high index of suspicion for acute compartment syndrome in the pediatric population is the key to avoid delayed or missed diagnosis. Although trauma (76% of cases), external compression from casts and splints (21% of cases), and burns are the most common mechanisms, coagulopathy may predispose patients to acute compartment syndrome, even in the setting of relatively benign trauma. Although the current patient had no previous diagnosis, fortunately, he was diagnosed early and recovered without sequelae of acute compartment syndrome.

**Conclusion**

Acute compartment syndrome is an orthopedic emergency and may result in devastating complications in the setting of delayed or missed diagnosis. A high index of suspicion and careful appreciation of pediatric-specific clinical indicators in addition to the traditional signs and symptoms may help to expedite the diagnosis. Adequate fasciotomy should be performed within 6 hours whenever possible. Coagulopathy may predispose to the development of compartment syndrome, even in the setting of relatively benign trauma, when it might not otherwise be expected. In the current case, the authors salvaged the leg of an 11-year-old patient with undiagnosed chronic myeloid leukemia who presented with unremitting calf pain after minor trauma. This report adds another unique case to the literature on compartment syndrome in the pediatric population and reinforces the need to consider compartment syndrome even in unlikely clinical scenarios.

**References**