



An Unusual Presentation of Acute Compartment Syndrome (ACS) in Adult and Pediatric patients: A Case Series and literature review

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Abstract

Acute compartment syndrome (ACS) is a serious condition that requires prompt diagnosis and immediate surgical intervention to prevent tissue necrosis, nerve damage, soft tissue and bone infections, and possible amputation. Typically, ACS presents following trauma with fractures of long bones. However, it can present uniquely without any cardinal symptoms of ACS, making the diagnosis of ACS challenging and increasing the risk for complications. According to the literature, intercompartmental pressure (IPC) and serum creatine phosphokinase can be helpful in the diagnosis of ACS. Fasciotomy should be performed as soon as possible to prevent unwanted complications. In this paper, we will present three unique cases of ACS in adult, adolescent, and young child with a review of current literature.

Keywords: Compartment Syndrome; Fasciotomy; Intramuscular Pressure; Perfusion; Pressure; Trauma

Abbreviations

Acute Compartment Syndrome (ACS), Emergency Room (ER), Physical Examination (PE), Creatine Phosphokinase (CPK), Intercompartmental Pressure (ICP), Open Reduction and Internal Fixation (ORIF), Closed Reduction and Internal Fixation (CRIF), Titanium Elastic Nails (TEN)

Background

Acute compartment syndrome (ACS) has been recognized since 1881 when Volkmann first described the contracture of the hand caused by compartment syndrome.[1] The first reported treatment of acute limb compartment syndrome was by Petersen in 1888.[2] The diagnosis and management of ACS in the adult population is well known, but only a few authors have reported on the paediatric population.[3]

ACS is a sometimes-diagnostic dilemma in patients who present with low energy trauma or no trauma at all (e.g., strenuous exercises, I.V drug use) or may not have the cognitive and verbal ability to provide clinical information, particularly in

an extraordinary situation (drug abuse), head trauma resulting in delays in diagnosis and adequate therapy. However, early diagnosis is pivotal for the outcome of ACS and is determined by the duration of ischemia and pressure in the osteofascial compartment. Immediate and adequate treatment can lead to healing with good functional and cosmetic results. If operative treatment is delayed or inadequate, excessive tissue necrosis may lead to severe local and systemic complications, such as reversible or irreversible muscle and neurovascular damage with functional loss of the extremity or myoglobinuria with subsequent acute renal failure.

The aim of the study was to analyze the diagnostic procedure for compartment syndrome in adult and pediatric patients and present the 3 cases of various clinical conditions that may lead to ACS diagnosis.

Case Presentation 1

A 15-year-old girl presented to the ER with a 3-hour history of left knee pain following a twisting injury to her knee while skiing, as her leg hit a stone during a fall. Following the fall, her left knee became erythematous, swollen, and painful, limiting her range of motion and ability to walk. Physical examination of the left knee confirmed this, as she was unable to bear weight on the left lower limb. Neurovascularly, the patient's left lower limb was intact, sensation present, and no signs of an open fracture were noted as the overlying skin was not disrupted. The patient's history is unremarkable. She was diagnosed with a left tibial tuberosity fracture type 2 that was confirmed on X-ray and CT-scan (Figure 1a-c).

An open reduction and internal fixation (ORIF) with a single screw was performed (Figure 1d,e). Once intraoperative radiographs were satisfactory, she was immobilized in a long, full cast. Patient was discharged home the next day and was

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Figure 1a Lateral X ray of 15 y/o with tibial tuberosity fracture (Ogden III).



Figure 1b CT in sagittal plane of the tibial tuberosity fracture (Ogden III).



Figure 1c CT in frontal plane of the tibial tuberosity fracture (Ogden III).

scheduled for follow up within 5-days or immediately in case of any complications (e.g., pain swelling, numbness, weakness).

Patient appeared in the clinic 1-week later. The cast was removed, and the surgeon noticed that the leg was pale, cold and the pulses were absent. The ultrasound (US) Doppler confirmed lack of blood flow. She was immediately scheduled for the surgery that confirmed total muscle necrosis and absence of blood flow. Unfortunately, the leg had to be amputated below the knee level.

Further studies and evaluation showed her tibial nerve was

damaged due to her primary injury and her pain sensitivity became decreased, thus not allowing her to feel symptoms of ACS. The compartment syndrome had developed due to damage of the anterior tibial recurrent artery.

Case Presentation 2

A 4-year-old boy was brought to the ER by his parents after falling from a swing an hour before. The parents noticed he started to cry immediately and stopped moving his right hand which had become swollen. He was healthy otherwise with an unremarkable history.

On physical examination, the patient was holding his left arm in flexion, supported by his right hand. Ulnar and radial pulses were present. The patient did not cooperate with sensory



Figure 1d A-P and lateral X ray of 15 y/o with amputation below the knee secondary to compartment syndrome.



Figure 1e A-P and lateral X ray of 15 y/o with amputation below the knee secondary to compartment syndrome.



evaluation. The patient was scheduled for x-ray of the left forearm, which revealed ulnar and radial shaft fractures at the same level (Figure 2a,b).

The patient was scheduled for CRIF with Titanium Elastic Nails (TEN) which was performed the next day with no complications. The forearm back slap was applied. Post-op x-ray demonstrated anatomical alignment of fractures and proper positioning of elastic nails (Figure 2c,d).

While being prepped for discharge that afternoon, the patient began feeling forearm pain that was unresponsive to medication. The hand was swollen with the restriction of finger movement. Pulses were present, but slightly diminished compared to the opposite upper limb. Sensation was difficult to evaluate due to lack of cooperation.

The intra-compartmental pressure was measured using



Figure 2a AP X-ray presenting the right forearm radial shaft fracture and plastic deformation of ulnar shaft.



Figure 2b Lateral X-ray presenting the right radial shaft fracture and plastic deformation of ulnar shaft.



Figure 2c Lateral X-ray of the right forearm after CRIF with TEN.



Figure 2d AP X-ray of the right forearm after CRIF with TEN.

Stryker device and under sedation, demonstrated elevation in dorsal and superficial Volar compartments 39° and 45°, respectively. The patient was taken immediately to OR and all compartments were decompressed and left open. Edema subsided after 3 consecutive days of repeated washouts, allowing wound closure.

The child was followed-up in the clinic and showed a well-healed fracture with no sensory or motor impairment after a 3-month post-physiotherapy period.



Case Presentation 3

A 43-year-old patient came to emergency room (ER) after hitting his right forearm over the stove while working in the restaurant kitchen. Initially he did not seek medical attention, but he woke up next day with pain, swelling and decreased sensation in his fingers. His medical history is significant for uncontrolled and untreated high blood pressure that was diagnosed 5 years ago, smoking of 1 pack a day for past 20 years and alcohol abuse (2-3 beers every night for past couple years).

On physical examination there is swelling in his right forearm, wrist and fingers, active movement of fingers was decreased because of pain and weakness. Pulses on the radial and ulnar arteries are diminished comparing the opposite limb. Sensation markedly decreased in the area of median and ulnar nerve distribution and slightly decreased in the area of radial nerve distribution. X ray shows no fracture in the forearm (Figure 3a,b).

Since the pain was increasing and patient was not responding well to the pain medications the pressure in the compartments was measured and was elevated up to 54 mmHg in superficial volar compartment and 34mmhg in the dorsal compartment.

The patient's management included emergent fluid resuscitation, fasciotomies, and debridement of necrotic muscle from his anterior compartment, and delayed primary closure.

After six months of intensive outpatient physical therapy he still presented with inability to abduct the thumb and weakness

in flexion of all fingers. Sensation was decreased in the area of ulnar nerve distribution.

Discussion

The most common cause of ACS is trauma, usually after fractures.[4] Following trauma, bleeding or edema in a closed osteofascial compartment causes an increased intercompartmental pressure (ICP) with ischemia.[5] Ischemia results in tissue membrane damage and leakage of fluid through capillary and muscle membranes. With arterial reperfusion, the damaged membranes continue to leak, and the rise in the hydrostatic force results in further increase of ICP, thus developing a vicious cycle.[6] The capillary perfusion decreases below a level necessary for tissue viability compromising the circulation and function of the tissues leading to muscle and nerve ischemia with muscle infarction and nerve damage.[7] In the early stage of ACS, Hoffmeyer et al., found marked perifascicular and intrafascicular edema with dissociation of the muscle fibres and necrosis in the tissue sections.[8] In the late stages, atrophy and hypertrophy of muscle fibres with lipid globules appear in the tissue examined.

Diagnosis on clinical grounds is not easy. In our case series, we present three patients (the adult, the adolescent and the young child) with atypical presentations of ACS, with one case resulting in amputation. ACS may present insidiously and affect patients with low energy trauma or nerve injury which alters the pain and sensory perception. Thus, clinical presentation, history, geographic location, and season is important when evaluating extremity swelling. This is highlighted in the case reports presented by Tadeka[34] and Maniar et al.[35]. Tadeka et al.[34] described two unique cases of ACS, hypothesized to be secondary to venomous bites from Pit Viper, mamushi (*Gloydius blomhoffii*). The presentation did not follow the typical high velocity injuries with subsequent fractures and/or soft tissue injuries radiographically and intraoperatively. Maniar et al.[35] presented a case of ACS in a 49-year-old-male with widespread prominent blisters and erythema that resembled necrotizing fasciitis, bullous pemphigoid, and/or infection/cellulitis. Unusual presentations, such as these, can lead to delayed diagnosis – therefore, increasing the patient's risk of developing further complications. In addition to helping diagnose ACS, clinical presentation is also key for fasciotomy indication, but must be confirmed with other diagnostic devices, since Ulmer reported a high false positive to true positive ratio when using the cardinal signs and symptoms of ACS for diagnosis.[12] Relevant trauma, tight extremities, excessive pain and pain on passive stretching, along with sensory loss requiring analgesia should raise suspicions of ACS. Checking levels of serum creatine phosphokinase (CPK) may also be helpful in ACS diagnosis, although not used in presented cases.

Young children are also challenging to evaluate in terms of history (obtained from parents) and PE, which increases the difficulty in early diagnosing ability. Pulselessness is usually only noted in the late stages,[11] thus making the classic cardinal ACS symptoms unreliable in diagnosing.[10] Special attention needs to be paid to skeletally immature patients presenting with tibial tuberosity fracture (Figure 1a,b,c). In this case, the anterior



Figure 3a A-P and lateral X ray of the forearm of 43-year-old male with blunt injury- swelling of the soft tissue is very well marked.



Figure 3b A-P and lateral X ray of the forearm of 43-year-old male with blunt injury- swelling of the soft tissue is very well marked.



recurrent tibial artery can be damaged and blood accumulation in tight leg compartments can lead to pressure elevation.

Utilizing (ICP) with a presumptive critical cut-off pressure in the diagnosis of ACS is still an ongoing discussion amongst authors. Fasciotomy indications vary: Mubarak et al., states an ICP of more than 30mmHg is indicative;[13] Whitesides et al., considered differences in diastolic pressure and ICP for indication. [14] Others evaluated the mean arterial pressure (MAP) and ICP difference as being indicative.[15] Along with Whitesides et al., Schmidt stated perfusion pressure, ($P = RR_{dia} - ICP$), as being the safest diagnosing method.[16] Using this definition, fasciotomy should be considered whenever pressure is more than 30 mmHg.

Due to increasing muscle growth pressing upon surrounding fascia,[18] healthy children present with higher compartment pressures (13-16mmHg, lower limb), than those of adults (0-10mmHg, lower limb).[17] Therefore, if clinical signs are noted in children, unconscious patients, or patients with regional nerve blocks, immediate ICP measurement is recommended. Some case report authors do warn against epidural anesthetic use, as it may mask symptoms of ACS in patients at risk,[19, 20] though some reviewed pediatric cases by Johnson and Chalkiadis found no clear evidence of delayed diagnosis with epidural anesthesia in children.[21]

Compartment pressure measurement can be made for example with the wick catheter technique modified by Mubarak et al.,[22] the simple needle manometry by Whitesides et al.,[23] infusion technique by Matsen et al.,[24] slit catheter technique modified by Barnes et al.,[25] side ported needles, etc.[26] An elevated pressure in one compartment is sufficient to establish the definite diagnosis.[18]

Overall, the diagnosis of ACS in children and adults can be challenging. The classic symptoms of compartment syndrome may be unobtainable. Adjunctive diagnostic tests such as invasive ICP measurement must be performed to guide treatment. In absence of high energy trauma, the compartment syndrome is also a possible diagnosis.

Learning Points:

- Low energy trauma without fractures can also lead to development of compartment syndrome.
- Coexisting nerve damage can decrease pain sensation and delay the diagnosis of compartment syndrome.
- Patients presenting with tibial tuberosity fracture with possible damage of the anterior recurrent tibial artery should be monitored closely and full casts should be avoided.
- In pediatric patients, the sensory evaluation can be challenging so intercompartmental pressure evaluation should be done under sedation to make the right diagnosis.

Declarations

Ethics Approval and Consent to Participate

Patients were lost to follow-up. All attempts to reach the

patients' or family members were unsuccessful. Therefore, the paper has been sufficiently anonymized to keep patient confidentiality and not cause harm to the patient or patient's family.

Consent for Publication

All authors, including me, Nicole Handloser, give consent to publish our manuscript in the Journal, BMC Musculoskeletal Disorder.

Competing Interest

All of the above authors declare that they have no conflict of interest.

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Authors' Contribution

MK, study concept and design, manuscript draft, review, and approval of final version. NH, study design, manuscript draft, review, and approval of final version. KC, study design, manuscript draft, review, and approval of final version.

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