Chapter 15
Unusual Presentation of Compartment Syndrome

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Background of the Problem

- Compartment syndrome is a well-described clinical entity considered to be an orthopedic emergency affecting all ages.
- Prompt recognition permits expedited treatment which is paramount for a good outcome.
- The typical scenario for acute compartment syndrome (ACS) is lower limb fracture or crush injury. However, it has been shown that this is not always the case.
- There is a big diversity of systemic diseases, which can rarely cause compartment syndrome.
- Unusual anatomical locations, rare conditions, drug interactions, and their side effects as well as surgical procedures and rare fractures can also be associated with this syndrome.
- In this chapter, we will outline and analyze the various unusual forms where compartment syndrome can be encountered.

What Is Recommended

- Physicians need to be extremely vigilant when dealing with patients who could suffer from compartment syndrome.

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- Good knowledge of its pathophysiology will help the surgeon guide his thought toward the specific diagnosis even in unusual presentations escaping from this difficult clinical setting.
- Awareness of unusual presentations, causes, or scenarios which can hide compartment syndrome should be known. The scope of this chapter is to increase the awareness of these situations.

Limitations and Pitfalls

This study is a detailed outline and analysis of a variety of rare presentations of compartment syndrome. It is impossible to include all the possible eventualities where a compartment syndrome can occur. We performed an extensive review of the available literature, but we acknowledge that there may be limited cases or conditions we may not have mentioned.

Future Directions

Further research is needed regarding diagnostic measures of compartment syndrome. More publications will give us further insight to this diverse problem.

Introduction

Compartment syndrome is characterized by an increase in pressure in a contained fibro-osseous compartment, such as the forearm or leg, resulting sequentially in decreased lymphatic and venous drainage, loss of arterial inflow, and subsequently diminishing of perfusion pressure. This leads to neuromuscular hypoxia and death of the contained structures. Established compartment syndrome if left untreated leads to contractures, sensory deficits, paralysis, permanent disability, amputation, and even death. In order to minimize morbidity and optimize treatment of a patient at risk for compartment syndrome, clinicians need a clear understanding of the pathophysiology, means (and problems) of diagnosis, and treatment of compartment syndrome. The typical scenario of compartment syndrome involves a young male with lower limb fracture or severe injury and presenting with tight leg compartments and pain out of proportion which cannot be relieved by painkillers. However, there is a great diversity of unusual conditions, common systemic diseases, medications, procedures, and atypical circumstances, which can be complicated by compartment syndrome.

Unusual Conditions

Exertional compartment syndrome is caused when strenuous exercise leads to swelling of the overexerted muscle in a closed compartment, resulting in increased tissue pressure. Wilson was the first to describe this uncommon type of compartment syndrome in 1912, and later Vogt in 1945 termed it as "march gangrene" [1]. Compartment syndrome after exercise can be divided in acute, acute-on-chronic or chronic, depending on the emergency or not of its presentation [2]. It was initially thought to affect only athletes, but Edmundsson et al. in 2007 reported that 36 out of 73 nonathletic referred patients suffering from exercise-induced pain were suffering from chronic compartment syndrome [3]. Exertional compartment syndrome has been frequently reported in people who follow a sedentary way of life and enthusiastically participate in intense sports activities.

Livingston et al. published this year a descriptive case series of seven young athletes suffering from acute exertional compartment syndrome (AECS) of lower leg [4]. In their retrospective study, they compared young athletes who suffered ACS after exertion with similar patients after a fracture. Diagnosis was set on average after 97 hours from symptom onset, whereas for fracture group this was 19 hours and only one patient required release of four compartments. Five out of seven patients had full recovery, whereas the other two needed a form of ankle orthosis. They postulated that half of those with longer than 24 hours of symptoms suffered from substantial muscle necrosis and functional deficit at final follow-up. In contrast, when the diagnosis of AECS was made in less than 24 hours of symptoms, there was no evidence of long-term sequelae. They also highlighted that patients with AECS were able to weightbear which complicates diagnosis as clinicians are trained to believe that pain from ACS precludes weightbearing. On their series, 86% exhibited neurologic deficit compared to 20% of fracture ACS, showing that neurologic damage is already present upon diagnosis. In AECS, anterior and lateral tibial compartments are mostly affected than the posterior compartments, and this phenomenon can be attributed to the fact that anterior tibialis and peroneus longus have higher percentage of fast-twitch fibers, making them prone to ischemia, whereas posterior compartment muscles have higher percentage of slow-twitch fibers, making them resistant to ischemia and suitable for endurance [5]. However, in Livingston's series, despite the high average intracompartmental pressures measured (91 mm Hg), there was no correlation between pressure and muscle damage, but a strong correlation of a time to diagnose more than 24 hours and myonecrosis is evident. This year, McKinney et al. reported a case of AECS affecting the anterior tibial compartment presenting with foot drop which was successfully treated with anterior lateral fasciotomies and rhabdomyolysis-supportive treatment, giving him full recovery apart from inability to extend of his hallux due to EHL necrosis [6]. Nicholson et al. reported AECS of the peroneal compartment on a 24-year-old healthy female after prolonged horse riding [7]. This was the first case related to a non-ground sporting activity. The patient presented with sensory deficit on the first

web space and dorsum of the foot, peroneal inability, and pain on anterolateral compartment, and this compartment was found to be necrotic during four-compartment fasciotomy. Common peroneal nerve, anterior tibialis muscle, and two other compartments were found healthy. Eventually, she made an excellent recovery, and authors postulated that this happened to her right leg due to the high boots and the leg position (knee flexed and dorsiflexed/inverted ankle) and not in the contralateral owing to a possible anatomical difference. Medial gastrocnemius tear was reported as a cause of AECS in a 55-year-old man who was running to catch a bus [8]. Four-compartment release showed the medial gastrocnemius tear 15 mm distal to the knee along with tear in peroneal artery and the resultant hematoma. Moreover, AECS can affect the upper limbs. In 2014, Bunting et al. reported bilateral supraspinatus AECS on a healthy 23-year-old male after strenuous weight lifting for an extended period [9]. Ultrasound-guided intracompartmental pressure measurements depicted pressures of 24 and 25 mmHg on the left and right trapezius, respectively, as well as 56 and 85 mmHg for left and right supraspinatus, respectively. With normal pressures considered to be from 3 to 20 mmHg, the diagnosis of bilateral supraspinatus AECS was set, and fasciotomies offered complete relief and excellent recovery.

Chronic exertional compartment syndrome (CECS) represents the result of overuse injuries affecting the extremities. The leg is the most frequent anatomical site particularly in running athletes [10]. CECS represents the second most common cause of exertional leg pain after medial tibial stress syndrome with an incidence ranging from 27% to 33% [11]. It appears equally in men and women at an average age of 20 [12]. The most commonly affected compartments are the anterior and lateral compartments (or a combination thereof) with an incidence of up to 95% of all CECS [13]. Typically, patients present with symptoms after increased intensity and duration of workouts that abate with cessation of activity. Over time, the pain that is experienced during exercise may increase, and patients may experience greater limitations during the provoking activities. Since symptoms are alleviated at rest, the ailment can frequently go undiagnosed for a period of time, increasing the severity of the condition. CECS is a clinical diagnosis; however objective measurements of intracompartmental pressures aid in confirming the diagnosis [14]. During exertion, compartment pressures increase three or four times from the baseline and return to basal levels within a few minutes in normal patients, whereas in patients with CECS, pressures increase more strikingly and take longer to return to their baseline (over 10 minutes) [15]. Pedowitz et al. in 1990 published modified criteria for the diagnosis of CECS based on intracompartmental pressures as up to then there was no consensus in the literature [16]. The criteria were based upon the intramuscular pressures recorded with the slit catheter before and after exercise in 210 muscle compartments without CECS. In the presence of appropriate clinical findings, they stated one or more of the following intramuscular pressure criteria to be diagnostic of chronic compartment syndrome of the leg: (1) a pre-exercise pressure greater than or equal to 15 mm Hg, (2) a 1-minute post-exercise pressure of greater than or equal to 30 mm Hg, or (3) a 5-minute post-exercise pressure greater than or equal to 20 mm Hg. Application of these criteria should result in a less than 5%

incidence of false-positive diagnoses. The only significant clinical difference between the group of patients suffering from CECS and non-CECS ones was muscle hernia at an incidence of 45.9% versus 12.9%, respectively. However, in 2012 Roberts et al. questioned the validity of these criteria [17]. They reviewed 38 studies from 1966 to 2010 and concluded that if clinicians carry out IMP testing, they should use a protocol with standardized catheter depth, exercise type, intensity and duration, footwear, and equipment. They argued that with the exception of relaxation pressure, the criteria set by Pedowitz for diagnosing CECS, considered to be the gold standard, overlapped the range found in normal healthy subjects. Therefore, they concluded that maximum reported upper confidence limits for pre, during, relaxation, and post 1- and 5-minute IMPs are 32 mmHg, 98 mmHg, 59 mmHg, 69 mmHg, and 48 mmHg, respectively. Pressures above these maximum values could certainly be considered abnormal under any circumstance. Although guaranteeing high specificity, the use of these values as cutoffs would likely have severe consequences on sensitivity. Therefore, they stated that mean upper confidence limits for the five time points are 14 mmHg (pre-exercise), 54 mmHg (during), 18 mmHg (relaxation), 36 mmHg (post 1 minute), and 23 mmHg (post 5 minutes). Values more than these must always be combined with clinical evaluation to safely reach a diagnosis. Nonoperative treatment of CECS includes rest, removal from inciting activity, stretching, anti-inflammatories, correction of training errors, and orthotics. However, this treatment is rarely followed due to the intensity of symptoms and also because patients cannot afford to abstain from their activities. Release of the anterior and lateral compartments has predicable success rates of roughly 80%, while deep posterior releases may yield success rates of 50% [18]. Irion et al. on their case series of 13 elite-level young athletes reported an 84.6% rate of return to their prior-activity level after an average of 10.6 weeks following surgical fasciectomy for CECS [10]. Involvement of four compartments resulted in longer return to full sporting activities after release. In a review of 100 fasciotomies for CECS, Detmer reported a recurrence rate of 3.4% [19]. CECS affecting the forearm usually involves the flexor compartment because of the higher exertion applied on these muscles during sporting activity and often occurs in rowers, climbers, and gymnasts. Open fasciotomies are considered the treatment of choice for forearm CECS, and these are the recommended ones. Nonetheless, endoscopic technique with single or multiple portals has also been described [20].

Neonatal form of compartment syndrome is rare and usually affects the forearm, wrist, and hand. The initial presentation is a superficial sentinel skin lesion or "sucking blister" at birth [21]. Several compression factors and neonatal conditions can induce neonatal compartment syndrome. Local mechanical causes include umbilical cord loops, fetal posture and oligoamnios, twin pregnancy, maternal uterine malformation, and amniotic band constriction [22]. This mechanical compression can be accentuated by maternal and neonatal conditions such as respiratory distress, vascular insufficiency, clotting disorders, and maternal diabetes [23]. It should be distinguished from gangrene of the newborn (usually involves lower limbs) [24], from necrotizing fasciitis (fulminating course of sepsis and skin lesions), and from aplasia cutis congenital (congenital absence of skin; ulcers involving symmetrical scalp,

trunk, and extremities; and heal spontaneously) [25]. The time from birth to surgery is the main prognostic factor. Misdiagnosis may lead to muscular and neuronal ischemia, with long-term devastating complications including Volkmann ischemic contracture and limb growth disturbances. Emergency surgery within hours of birth yields good results. Badawy et al. in their case report of neonatal compartment syndrome with concomitant disseminated intravascular coagulopathy advised that the decision to perform fasciotomy in a neonate with suspected compartment syndrome should be based on a clinical diagnosis rather than compartment pressures [26].

Idiopathic spontaneous is the term for compartment syndrome that developed without any identified triggering factor. Matziolis et al. in 2012 reported the case of an otherwise healthy male treated with fasciotomies of the lower leg without any identifiable causative factor or underlying health abnormality on their extensive workout [27]. A similar case affecting the tibia was also reported by Grevitt et al. in 1991, but we must highlight that this subtype remains extremely rare in literature [28].

Other rare and atypical forms of compartment syndrome can be provoked by severe infection, and a characteristic form of that is the necrotizing fasciitis. The principles of treatment in such cases are the same with the additive effect of extensive antimicrobial treatment and extensive debridement of infected tissues. Apart from that, rare infections can cause the entity. Last year, Stull et al. reported the case of a 6-year-old man treated for compartment syndrome caused by Proteus-infected hematoma of the lower leg [29].

Vascular abnormalities such as arteriovenous malformations and fistulae can be the cause for hematoma formation and recurrent compartment syndrome. Such case was reported in the thigh of a 31-year-old fit and well male from Bournemouth who suffered ten times recurrent ACS of his thigh [30]. MR angiography at the last occurrence depicted abnormal vessels arising from profunda and superficial femoral arteries which have been embolized. This vascular abnormality was considered to arise from an old femoral fixation many years before, but the authors stated that formation during the previous fasciotomy procedures could not be excluded.

Systemic Diseases

Diabetes mellitus is considered one of the diseases making patients susceptible to developing compartment syndrome [31]. Non-enzymatic glycosylation makes diabetic collagen stiff, and microvascular alterations lead to limited joint mobility, skin alterations, and cheiroarthropathy. As a result, fascias are less distensible in potential elevation of compartmental pressures. Coley et al. in 1993 reported the case of a 44-year-old insulin-dependent diabetic woman with bilateral lower leg compartment syndrome treated effectively with fasciotomies [31]. They postulated that long-term diabetes is the cause for joint stiffness and microscopic collagen alterations. Lower and Kenzora in 1994 found that diabetic feet have elevated intramuscular compartment pressures in relation to healthy controls [32]. This mechanism in

addition to diabetic collagen modulation could explain the cases of compartment syndrome reported in diabetic patients. Although it seems that long-lasting type I DM can be complicated by ACS, Flamini et al. in 2008 reported spontaneous compartment syndrome following statin administration on an asymptomatic type II diabetic patient [33]. They supported that administration of statins combined with type II DM activated a vicious circle of inflammation, edema, and necrosis.

Hypothyroidism was reported as another cause of compartment syndrome in 1993 by Thacker et al. [34]. They described bilateral lower leg ACS on a prior undiagnosed male with myxedema. In hypothyroidism, increased protein extravasation along with relatively slow lymphatic drainage leads to an increase in compartment contents [35]. On the one hand, skeletal muscle hypertrophy occurs in 1% of cases of myxoedematic myopathy [36] and is named as Hoffman's syndrome in adults and as Kocher Debre–Semelaigne syndrome in infants and children [37]. On the other hand, lack of thyroxine diminishes degradation of hyaluronate and along with TSH-derived stimulation of fibroblasts leads to increase in the connective tissues contents [38]. An increase in energy demand during mild exercise has been associated with increased risk of rhabdomyolysis in patients with uncontrolled hypothyroidism [39]. These systemic implications of hypothyroidism can explain the reported case of all-extremities compartment syndrome reported in 2016 by Musielak et al. [40]. Hypothyroidism can cause dislipidemia, which if treated with statins can both in combination cause rhabdomyolysis and concurrent ACS [41]. Therefore, in newly diagnosed dyslipidemia, screening for hypothyroidism is advocated because thyroid repletion alone can correct abnormal lipid profile and thus avoiding the risks of myopathy that statin therapy involves [42]. Primary hypothyroidism combined with adrenal insufficiency can cause rapid onset of rhabdomyolysis and myonecrosis, resulting in foot drop and poor prognosis despite treatment [43].

Hematological disorders or malignancies are known to involve or be the cause of compartment syndrome. Mostly in such cases, the underlying pathology is revealed from biopsies taken during fasciotomies for compartment syndromes of unknown origin. Non-Hodgkin lymphoma was found to infiltrate the muscles in the leg of an 80-year-old woman causing compartment syndrome, and leukemic infiltrates caused compartment syndrome in a 20-year-old man [44, 45]. In such oncological cases, aggressive tissue debridement is advised to facilitate primary closure as adjuvant chemotherapy or radiation may complicate any open wounds [46]. Myeloid sarcoma without transformation to acute myeloid leukemia (AML) has been described as a cause of ACS affecting anterior tibialis [47]. Another cause for hematological-originated ACS apart from infiltration is the excessive bleeding. Chronic phase of myeloid leukemia (CML) as a myeloproliferative disorder with excessive platelet number was found to cause ACS through excessive bleeding which persisted after fasciotomies and seized only when their number was controlled with cytosuppresive treatment [48]. However, it must be noted that in chronic CML, platelet dysfunction is not always due to their number, and urgent hematological consult should be sought [49]. Atypical ACS on grounds of chronic CML has also been described in pediatric population [50].

Clarkson reported in 1960 the first case of an otherwise fit and healthy 34-year-old Italian woman who exhibited unexplained cyclical episodes of edema and severe shock due to increase in capillary permeability which resulted in plasma shift from intravascular to the interstitial space [51]. Hemoconcentration was pronounced as red blood cells are large to be filtrated from the endothelium, and also low albumin was found. These episodes seemed to occur premenstrually, but hysterectomy and oophorectomy failed to solve the problem, and she died after a severe episode of shock. Autopsy did not shed light, and the only striking finding was monoclonal gammopathy. This rare entity was named *systemic capillary leak syndrome* (SCLS), and to date 500 cases have been described worldwide primarily in middle-aged adults [52]. It was also found to be associated with rhabdomyolysis and compartment syndrome [53]. Compartment syndrome of all four limbs has been described which was effectively treated by fasciotomies, but the syndrome itself has a poor prognosis and predisposition to multiple myeloma and leukemia [54]. SCLS presents in three phases: prodromal, extravasation, and recovery [55]. In prodromal phase, symptoms include lethargy, vomiting, abdominal pain, and generalized weakness; in extravasation phase pleuric, pericardial, epiglottic, macular, and generalized peripheral edema present along with shock; and in recovery phase, pulmonary edema can occur due to the mobilization of fluid to intravascular space. It is important not to overlook other usual cases of shock and allergies. The etiology of this syndrome remains unknown, no familiar distribution was found, and various treatments such as theophyllines, terbutaline, steroids, plasmapheresis and thalidomide have been employed with variable success, leaving a mortality rate of 25–30%. In June of this year, the first metanalysis of published SCLS in childhood depicted 24 relevant studies and showed that the syndrome also affects childhood and follows acute illness in 75% but not related to any monoclonal gammopathy [56].

Human immunodeficiency virus (HIV) infection can also rarely be complicated by compartment syndrome. The pathophysiology may vary: HIV-induced thrombocytopenia causing bleeding and myositis from antiretroviral treatment have been reported [57, 58]. A rare case of bilateral spontaneous lower leg compartment syndrome was attributed to antiretroviral-induced myositis [59].

Moreover, compartment syndrome of the hand has been reported in a case of *multiple sclerosis* possibly associated with the cutaneous changes of that syndrome [60].

Drugs

Statins (hydroxymethylglutaryl coenzyme A (HMG-CoA) reductase inhibitors are widely prescribed to treat hyperlipidemia. Myogenic damage is known to be one of their side effects. Coadministration of simvastastion with risperidone (atypical neuroleptic drug) has been reported as a cause of compartment syndrome [61]. It was postulated that risperidone may have diminished the metabolism of simvastatin via interactions with the cytochrome P450 (CYP) system, resulting in marked plasma elevation of simvastatin and consequent rhabdomyolysis and compartment syndrome.

Serotonin syndrome was also reported as a cause of compartment syndrome [62]. This syndrome involves encephalopathy, neuromuscular contractures, and clonus and autonomic hyperactivity. A 68-year-old woman was taking paroxetine (a selective serotonin reuptake inhibitor (SSRI)) and risperidone and sustained a serotonin syndrome with consequent rhabdomyolysis and compartment syndrome of both legs. As a causative factor, the serotonin syndrome was suspected, but coadministration of risperidone may have played its role. Risperidone was also found to be the cause of bilateral tibial compartment syndrome in a 31-year-old man suffering from schizophrenia after 1 and half hours of walking without any other predisposing factors [63].

Lithium has also been reported as causing an atypical form of compartment syndrome: atraumatic, painless, and affecting only one out of four tibial compartments [64]. It is considered as pain-perception altering with effect to pain receptors so patients appear obtunded.

Unusual Anatomical Locations

Gluteal compartment syndrome is rare. Gluteal compartments were described in the cadaveric study of David et al.: three compartments from lateral to medial with one enclosing tensor fascia lata, one gluteus medius plus minimus, and one containing gluteus maximus [65]. Sciatic nerve dysfunction is a common clinical finding in gluteal compartment syndrome despite the fact that sciatic nerve is enclosed within a separate compartment. Sciatic involvement is mostly attributed to external compression on its arterial supply, which most commonly arises from the medial circumflex femoral and inferior gluteal arteries. Out of the two branches of the sciatic nerve, the peroneal (fibular) is more susceptible to injury; thus, the patients may present with only isolated foot drop [66]. The nonspecific symptoms of buttock swelling and tenderness often lead to misdiagnosis such as pelvic or lower limb venous thrombosis, and the initiated antithrombotic treatment further aggravates any gluteal hematomas, resulting in even higher compartmental pressures [67]. Differentiation between gluteal compartment syndrome and thrombosis requires CPK measurement, intracompartmental pressure measurement, and imaging studies. No definition of the threshold of abnormal raised intracompartmental pressure is needed to diagnose gluteal compartment syndrome. Normal values have been reported to be 13–14 mm Hg [68]. Emergent fasciotomies are considered the treatment of choice, and even in delayed presentation after 56 hours, they provided a favorable outcome [69]. Therefore, Panagiotopoulos et al. on their case report with residual sciatic nerve palsy despite fasciotomies reported that nonoperative treatment should have limited place due to high risks and minimal benefits [66]. Kocher-Langenbeck approach is usually used for the gluteal fasciotomies. Henson et al. in 2009 published the first and only systematic review of gluteal compartment syndrome [70]. They reviewed seven papers (28 cases) which were all retrospective case reports and summarized that the causes

of compartment syndrome affecting the gluteal region can be trauma, vascular injury or surgery, intramuscular drug abuse, altered level of consciousness from alcohol intake or drug overdose, prolonged immobilization, epidural anesthesia after joint arthroplasty, and infection. On their systematic review, half of the papers had prolonged immobilization as the leading cause. Diagnosis was based solely on clinical symptoms in 53.6%, and intracompartmental pressures greater than 30 mmHg were considered as definite indication for surgical treatment. Surgical fasciotomies were the preferred method of treatment in 71.4% of the cases, and only 12 out of 25 cases recovered fully. Therefore, authors highlighted that gluteal compartment syndrome implies a big cause of patient disability and that there is a lack of an adopted system of precise indications for surgery and of functional evaluation regardless of the way of treatment. Gluteal compartment syndrome has been reported as a complication after bone marrow biopsy from iliac crest to a patient who was anticoagulated [71]. The same complication was reported after posterior iliac crest marrow biopsy of a patient suffering from non-Hodgkin lymphoma; however, on this case, platelet number and clotting times were normal [72].

Compartment syndrome in lumbar region may be a cause of severe low back pain according to Peck in 1981 [73]. Lumbar paraspinal compartment syndrome was then officially described in 1985 by Carr in a young man with severe low back pain after exertion [74]. It seems that recently weight lifting including "CrossFit," which have gained popularity, have accounted for many of such compartment cases reported. Paraspinal muscles are enclosed by the thoracolumbar fascia which behaves like a closed space with resting intracompartmental pressures varying from 3 to 7.95 mmHg, depending on the position and being elevated up to 25 mmHg during exercise [75]. Patients present typically with severe low back pain, bilateral symptoms, swollen paraspinal muscles, pain in the hip flexion but not in straight leg raise, and absent bowel sounds due to ileus. Diagnostic modalities such as MRI or CT, though not used routinely in compartment syndrome of extremities, play a significant role in diagnosis of lumbar paraspinal compartment syndrome. Paramedian Wiltse incision rather than a midline is used as it allows for delayed soft tissue closure or grafting over a viable muscle bed [76]. After the release of the fascia, the relevant compartments of longissimus, iliocostalis, spinalis, and multifidus are approached. Alexander et al. this year published the first systematic review of acute paraspinal compartment syndrome [77]. They assessed 21 retrospective case reports and found that the cause was mainly not related to direct trauma to spinal muscles but in 52 % of cases was related to weightlifting and the rest to other sport activities such as skiing or surfing and spinal surgery. Intracompartmental pressures were measured with patient prone and averaged to 73.7 mmHg, much higher than other body locations. Fasciotomies were applied in twelve of the twenty-one case reports, where nine of them received medical treatment and two hyperbaric oxygen therapy. All cases treated surgically even with delays up to 7 days had a good outcome, whereas all conservative and hyperbaric oxygen cases had ongoing symptoms or functional deficit. Therefore, they

suggested that surgical decompression on confirmed diagnosis should be the treatment of choice regardless of the delay of diagnosis. Only one case of chronic lumbar paraspinal compartment syndrome is reported after weight lifting and was treated successfully with bilateral minimally invasive fasciotomies at L3 level under local anesthesia [78]. Fascia was found thickened, and samples confirmed the hyperplastic muscle fibers. The patient after 4 weeks returned to his weight lifting training for the 2008 Olympics without any residual symptoms.

Other unusual anatomical locations which can be affected by compartment syndrome are the medial head of gastrocnemius (tennis leg) and peroneus longus [79, 80].

Procedures

Coronary artery bypass grafting can be complicated by leg compartment syndrome [81]. The exact mechanism is unknown, although prolonged bypass time or saphenous vein harvesting on patients under statins has been implicated as risk factors.

Total knee replacements in very rare occasions have been associated with compartment syndrome of either the glutei, thigh, or tibia. As regards the tibia, this situation cannot be explained as the replacement takes place in a different compartment [82]. However, tourniquet time, epidural anesthesia, continuous passive movement device, thromboprophylaxis, and aggressive physiotherapy have all been thought as contributing factors without a clear correlation being identified with the exception of the tourniquet time. Functional outcomes after fasciotomies remain moderate mainly due to periprosthetic infections.

Postprocedural compartment syndrome resulting from placement of neuromonitoring needles in forearm has also been described [83]. Patients undergoing such endovascular procedures are on antiplatelet treatment, and therefore at an increased risk of bleeding and elevated intracompartmental pressures. Prevention can be achieved by extra vigilance to avoid superficial veins and vertical insertion to the skin.

Unusual Fractures

Innocuous distal radius in elderly and low demand patients have been reported to be complicated by compartment syndrome. This was an extremely rare manifestation unable to be explained by fracture displacement, mechanism, and severity of injury or any other factors. In one case, amputation of digits was performed despite initial fasciotomy [84]. Chloros et al. in their paper highlighted the significance of the pronator quadratus space and its potential role [85].

Take-Home Message

- Compartment syndrome can present atypically and in variable context. Raised awareness on a suspected case can prove limb/lifesaving.
- Systemic diseases, drugs, rare body locations, unusual conditions and fractures, and common surgical procedures can all be associated with this syndrome.
- Symptoms do not differ from a usual acute compartment syndrome.
- Measurement of intracompartmental pressures should not delay treatment in highly suspicious cases with clear clinical symptoms.
- Irrespective of the cause, condition, or location, surgical fasciotomies of involved compartments remain the treatment of choice as per acute compartment syndrome (ACS).
- Due to their atypical manifestation, delays in treatment can commonly appear unfavoring its final prognosis.

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