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Acute compartment syndrome of forearm in a haemophiliac patient

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Abstract
Introduction: Haemophilia A and B are X-linked bleedingdisorders included among the rare diseases and causedby mutations in the factor VIII (FVIII) and factor IX(FIX) genes. Acute compartment syndrome (ACS) of the limb refersto a constellation of symptoms, which occur following a risein the pressure inside a limb muscle compartment. Here we are reporting a rare case of Haemophilia A patient presenting with Acute compartment syndrome and the sequential steps in the management which include emergency fasciotomy, debridement and flap cover.
Conclusion: Haemophilia presenting as ACS is rare and a high index of suspicion is needed to diagnose it. The results are favourable when the diagnosis is made early.

Keywords: Compartment syndrome, forearm, haemophiliac patient

Introduction
Haemophilia A and B are X-linked bleeding disorders included among the rare diseases and causedby mutations in the factor VIII (FVIII) and factor IX(FIX) genes [1]. Although the disease has been mentioned in ancient times John Conrad Otto, a physician from Philadelphia in 1803 published "An account of ahaemorrhagic disposition existing in certain families [2]" where he noted the characteristic feature of haemophilia, i.e., an inherited tendency of males to bleed. The first use of the word "haemophilia" was written in 1828 by Hopff from the University of Zurich [1].
Acute compartment syndrome (ACS) of the limb refersto a constellation of symptoms, which occur following a risein the pressure inside a limb muscle compartment. ACS has been recognised since 1881 when Volkmann first described the contracture of the hand caused by compartment syndrome and hence the name Volkmann's Ischemic Contracture (VIC) [3].
Haemophiliacs are predisposed to bleeding and can develop intra-articular, muscular, or neural haemorrhage [4]. A scenario with a bleed into the muscular compartment, either spontaneous or following trivial trauma could lead to compartment syndrome. There have been very few case reports of such cases and we would like to report a similar case.

Case report
A 9 year old boy was brought to the outpatient department with complaints of pain and swelling of the left upper limb for two days. He was unable to move his fingers due to pain in the forearm and had paraesthesia of his fingers. The symptoms were constant with no relief in pain and progression of paraesthesia. He had allegedly sustained a fall two days earlier following which he was treated elsewhere and a splint had been applied. He had not undergone any surgery earlier and there was no relevant past or family history suggesting prolonged bleeding. He is the first born male child of a consanguineous marriage.
The splint was removed in the emergency room and a diffuse swelling of the forearm was noted with the skin being stretched and shiny (Fig 1). The extremity was cold to touch with tenderness at the proximal third of forearm. Active movements of the hand and wrist were absent. Stretch pain of the fingers was present. Abnormal mobility/crepitus was absent. There was no sensation below the elbow. Radial pulse was not palpable but capillary refill was present. A doppler study did not show any flow in the radial or ulnar arteries.

Radiographic study was normal and the patient was prepped for emergency fasciotomy. Coagulation profile revealed aPTT values were elevated and on further assessment, factor VIII value was found to be reduced. A diagnosis of acute compartment syndrome of forearm in an undiagnosed haemophiliac patient was established.

He was operated with precautions for excessive bleed under the cover of Factor VIII, fresh frozen plasma, cryoprecipitate and packed cells. An emergency fasciotomy was done with an incision over the middle of the anterior aspect of the arm, curved across the elbow along the crease and the extending along the length of the forearm and wrist (Fig 2). The superficial and deep compartments of the forearm was exposed. The arm and forearm muscles were contused and flexor digitorum superficialis & profundus (FDS &FDP) were found to be necrotic and debrided (Fig 3). A large hematoma, (Fig 4) probably from the trivial injury and a cause for the increased compartmental pressure, was evacuated from the deep compartment. The carpal tunnel was released freeing the median nerve and radial artery.

A second debridement done two days later showed a completely devitalised FDS and FDP which was hence removed (Fig. 6). An Abdominal flap cover was given on the sixth day to the exposed forearm musculature. It was divided at the end of three weeks (Fig. 7 &8). Patient had serial PTT analysis and transfusion support as required under the supervision of the Haematologist.

Satisfactory recovery in sensation and functions of the limb was noted at 18 months' follow up.

Discussion

The prevalence of haemophilia A is 1 in 5000 male live births, and that of haemophilia is 1 in 30,000 [1]. The increased propensity to bleed could result in haemorrhage into a closed muscular compartment such as the forearm resulting in acute compartment syndrome which if not attended to can lead to permanent damage and may end in amputation. Diagnosis and management in these scenarios especially in children remains a challenge [5].

A spontaneous bleed into a closed muscle compartment could cause a "major bleed", which is a critical event in haemophilia that could cause a drop of haemoglobin up to 2g/dl necessitating a transfusion [6]. Such a bleed is an emergency in itself, but the ensuing compartment syndrome is equally dangerous. In haemophilia, bleeding into the compartment causes increased pressure in the compartment causing muscle ischemia which causes tissue membrane damage leading to leakage of fluid which further increases the hydrostatic pressure in the compartment. This sets up a vicious cycle that causes capillary reperfusion to fall below the necessary level for tissue viability [7, 8].

The diagnosis of haemophilia in a patient who presents as ACS could be challenging due to the very scarce literature and awareness [9]. A high degree of suspicion is warranted in young males born out of a consanguineous marriage to develop a spontaneous ACS as in our patient. The first case was published by Hey-Groves [10] in 1906 and 71 years later, 4 similar cases were described by Lancourt *et al*. [11]

Pain on passive stretch, palor, pulselessness, paraesthesia and paralysis are the "5P's" and the diagnostic symptoms of ACS [12]. However there was a high chance of false positive rates when clinical features alone were considered and hence other diagnostic methods are recommended to be considered [12].

The role of Intra compartmental Pressure (ICP) measurement is still controversial and not standardised regarding the cut off critical pressure. The most reliable method as per Whitesides *et al*., is a difference between the diastolic pressure and the ICP with the cut off at 30 mm of Hg [13].

Near infrared spectroscopy (NIRS) is an expensive, non-invasive, continuous method for monitoring the compartment pressures suited in children. It measures tissueoxygenation or hypoxia by measuring the muscle oxy-haemoglobin which reflects compartment pressure, perfusion pressure and loss of myoneural function [14, 15].

X-rays are done as a routine and ultrsonography & doppler studies can be done to check for blood flow and collections.

Creatine Phosphokinase levels could indicate muscle breakdown. These aforementioned methods hold more theoretical value as the decision for fasciotomy is taken primarily by clinical signs rather than investigations [16].

The time interval between the onset of symptoms and tissue necrosis is very short and hence early intervention could be limb saving and even life saving. Due to the tamponade effect, a significant drop in the haemoglobin would not be noted hence adequate preparation is essential. The ongoing bleed, the underlying pathology needs to be controlled and hence transfusion of factors could arrest the haemorrhage and result in spontaneous resolution with rest, splinting and limb elevation limited to the early period [17, 18].

The rise in ICP beyond 30 mm of Hg is a definitive indication for surgery in the form of emergency fasciotomy. Complete decompression of the forearm compartments including the neurovascular structures is of utmost importance. Tissue with doubtful viability is to be left in place due to better healing potential in children. Skin is to be secondarily closed or split skin graft or abdominal flap can be done at an appropriate date [19]. Naranja *et al* advised a 50-100% factor level to be maintained pre and post operatively.

Complications are more common in delayed fasciotomies and include infection of soft tissue or bone; permanent nerve damage; development of Volkmann's Ischemic Contracture; amputation. Systemic complications can occur due to rhabdomyolysis and myoglobinuria leading to renal insufficiency and death. Total permanent complication in one study stood at 4.2%.

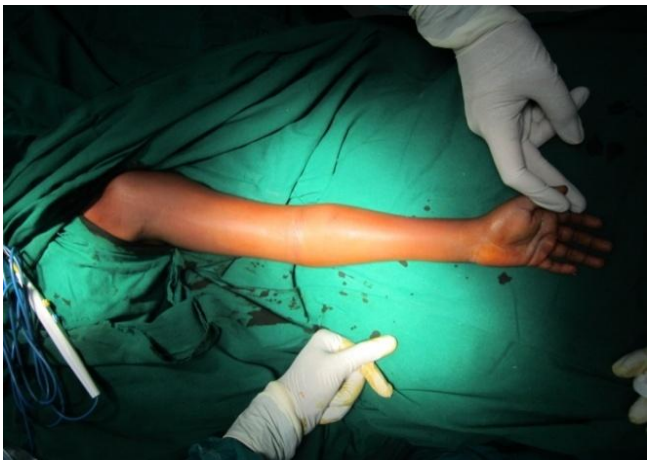

Fig 1: Swollen forearm with stretched and shiny skin

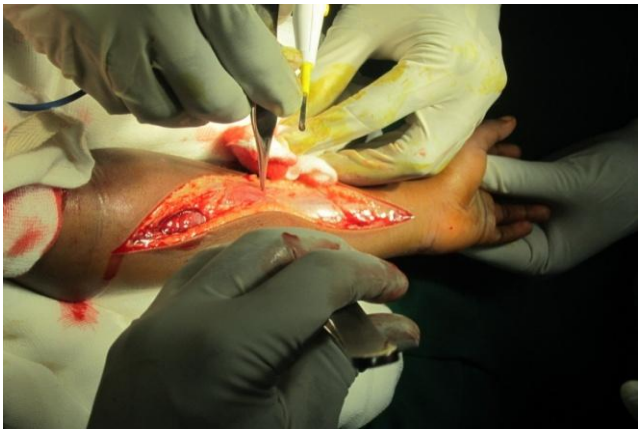

Fig 2: Incision along the middle of arm extended, curved along the elbow crease and down the middle of forearm.

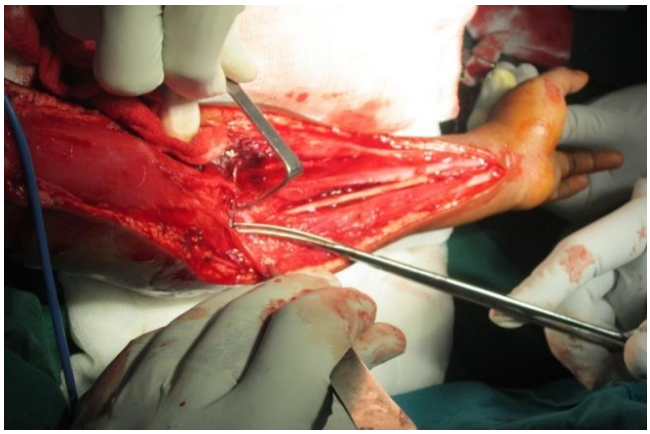

Fig 3: Contused FDS and FDP

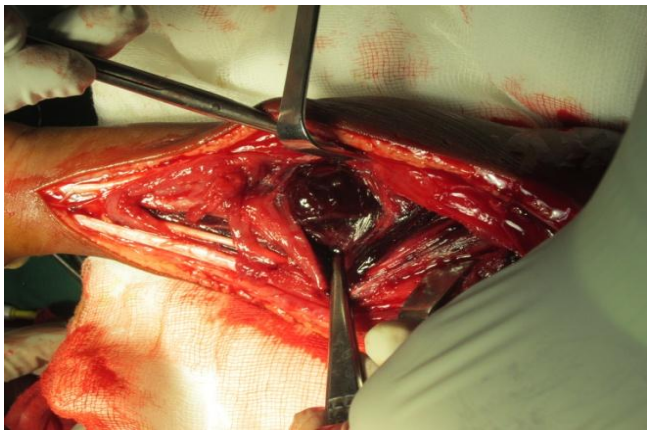

Fig 4: Deep compartment showing hematoma

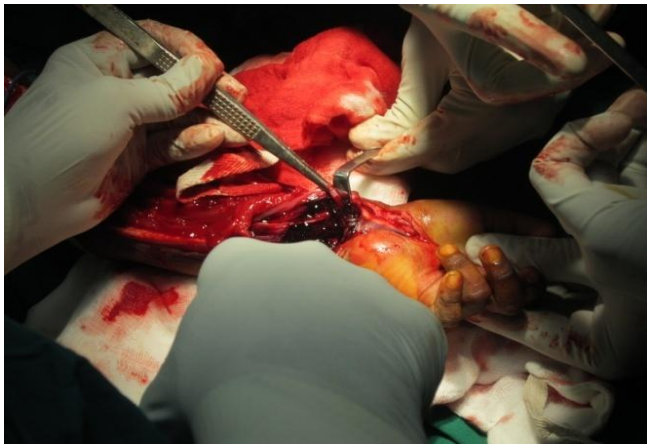

Fig 5: Extension of hematoma upto the hand

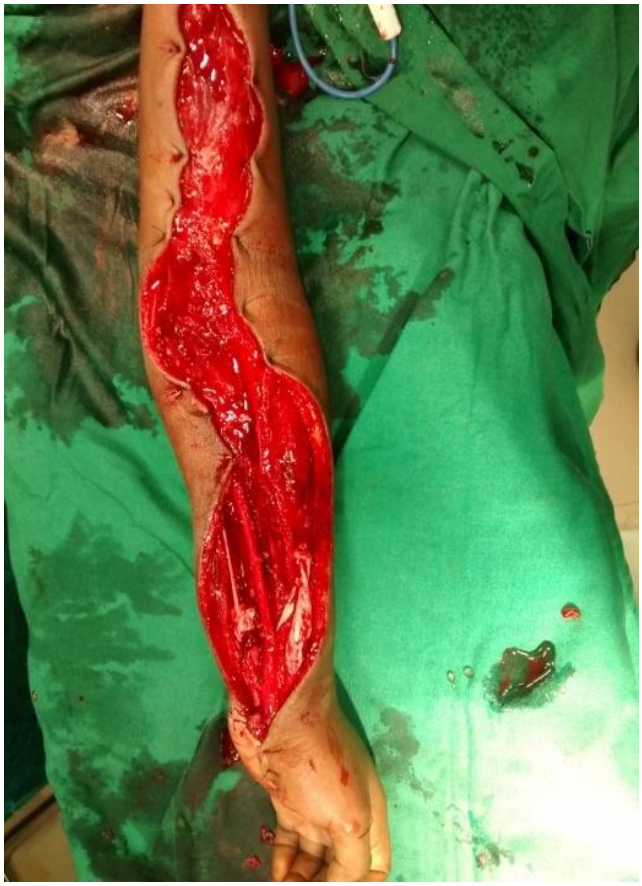

Fig 6: Second debridement and removal of FDS and FDP. Median nerve is exposed.

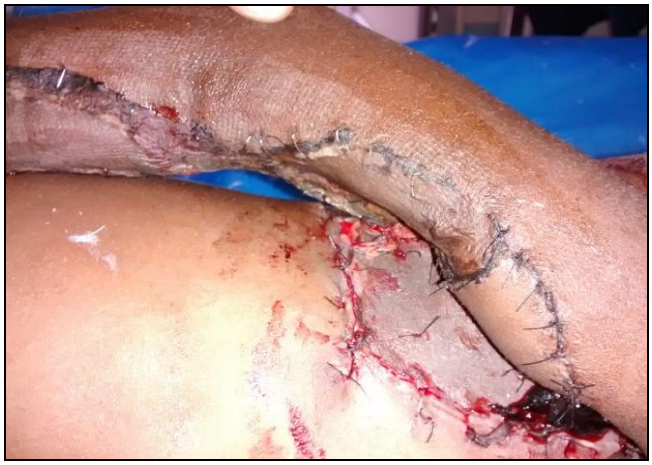

Fig 7: Abdominal Flap Cover

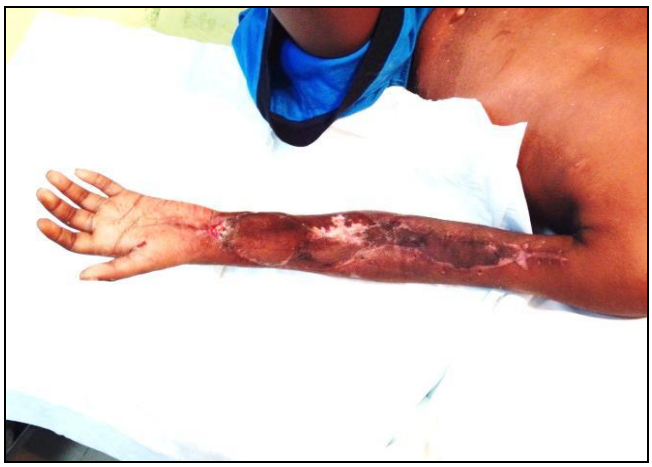

Fig 8: After flap division. SSG taken up completely

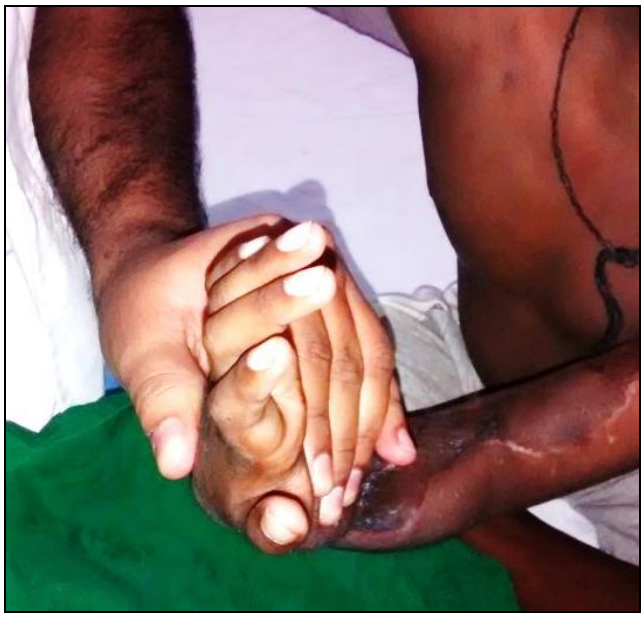

Fig 9: Recovered wrist flexion

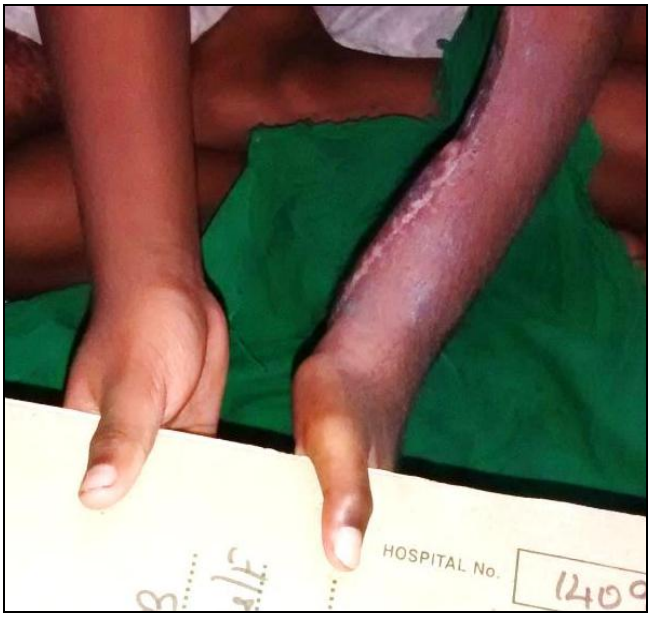

Fig 10: Recovering thumb adduction

Conclusion

Haemophilia presenting as ACS is rare and a high index of suspicion is needed to diagnose it. Routine blood studies may reveal the diagnosis. The treatment of haemophilia is simple in comparison to the treatment of acute compartment syndrome of forearm in a patient with haemophilia. Treatment includes an emergency fasciotomy and thorough debridement of necrotic tissues under the cover of factor VIII and blood transfusions as required. A second stage plastic surgery is planned. This is followed by rehabilitation and additional surgeries to regain the lost function. The results are favourable when the diagnosis is made early.

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