An unusual presentation of aplastic anaemia: compartment syndrome

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This is a case of a previously fit and healthy 17-year-old man presenting with atraumatic forearm compartment syndrome. On his initial blood film, he was found to have a pancytopenia and was subsequently diagnosed with aplastic anaemia. His compartment syndrome was treated with urgent fasciotomy after correction of his pancytopenia. He made a good functional recovery with only a very minor deficit, and no recurrence of his compartment syndrome. He has undergone a successful bone marrow transplant for treatment of his aplastic anaemia.

INTRODUCTION

Upper-limb compartment syndrome is an uncommon diagnosis [1, 2]. We present a case of a previously fit male presenting with a spontaneous upper-limb compartment syndrome and subsequent diagnosis of aplastic anaemia.

CASE REPORT

A 17-year-old man presented with pain and swelling of his left forearm with his wrist and fingers held in a flexed position (Fig. 1). Prior to this, there was a 3-day history of progressive pain in the forearm, but no history of trauma. The patient had no previous medical history, was on no regular medications and had no history of previous infections. His observations showed pyrexia of 39°C, but all others were within normal limits. A clinical diagnosis of compartment syndrome was made and the patient was planned for urgent fasciotomies. Routine blood examination demonstrated pancytopenia with a haemoglobin level of 3.9 g/dl, white cell count of 0.9 × 109/l and platelet count of 4 × 109/l. In addition, an urgent ultrasound scan was performed to rule out haematoma and possible tamponade of an arterial bleed. The scan demonstrated a collection within the ulna volar forearm. After transfusion of three units of cross-matched blood, two units of fresh frozen plasma and two units of platelets, an urgent fasciotomy and drainage of the collection under general anaesthesia was performed with a concurrent iliac crest bone marrow biopsy.

A modified Henry’s approach and carpal tunnel decompression was performed and revealed a collection of pus was superficial and ulna to flexor digitorum superficialis (FDS) muscle. The distal muscle of FDS was ischaemic and swollen preventing finger extension due to the tight wrist and carpal tunnel fascias. The ischaemic muscle was debrided and a dusky flexor carpi ulnaris (FCU) was decompressed and left in situ for a review at second look surgery (Fig. 2).

Deep tissue and pus samples grew gram-positive cocci—Staphylococcus aureus, sensitive to flucloxacillin, on which he was commenced after samples were taken. No further focus of infection was identified. In the following 12 days, the patient underwent four further surgical debridements, topical negative pressure wound therapy and delayed primary closure of the wound. Only partial resection of FDS and FCU was necessary; therefore, all functional units were maintained (Fig. 2).

The bone marrow biopsy confirmed a diagnosis of aplastic anaemia. An immediate family member was found to be a match for a bone marrow transplant which has been carried out successfully.

At latest review, 6 months following initial presentation, the patient had regained full extension of wrist and digits, full wrist flexion and full finger flexion apart from a mildly limited...
middle finger which comes within 1 cm of his palm. His median and ulna nerve sensation has resolved after being slightly reduced compared with the opposite side in the initial postoperative period.

DISCUSSION

This is the first reported case in the literature of a compartment syndrome being the first presentation of aplastic anaemia. Aplastic anaemia is a rare haemopoietic stem-cell disorder that is defined as pancytopenia with a hypocellular bone marrow. It has a variety of presenting symptoms, including anaemia, bleeding disturbances and a wide range of infections [3, 4]. This case demonstrates the susceptibility of these pancytopenic patients to unusual presentations of infections. The diagnosis of aplastic anaemia was made following the routine blood results and subsequent bone marrow biopsy.

Compartment syndrome occurs when the pressure in an osteofacial envelope increases and prevents tissue perfusion, resulting in ischaemia and subsequent tissue necrosis. Early diagnosis is vital to prevent permanent tissue necrosis and the formation of fibrotic tissue which leads to permanent disability.

Upper-limb compartment syndrome is a rare diagnosis and is most commonly caused by trauma resulting in forearm fractures, but haemorrhage, iatrogenic injuries and many atraumatic causes are reported [1, 2]. These include post-thrombolysis, secondary to acquired Factor VIII Inhibitor, following anticoagulation and secondary to insect or snake bites [1, 2, 5–8]. These were all unusual presentations, and therefore the clinician must have the diagnosis of compartment syndrome in their mind for unexplained pain in the forearm.

The management of compartment syndrome is through fasciotomy which provides release of the high compartment pressures and allows tissue perfusion to be restored. There are numerous different reported techniques for the incision for a forearm fasciotomy [1, 9]. Patients with upper limb compartment syndrome have a varied outcome with up to a third suffering some long-term complications, and a fifth suffering some neurological deficit [1, 9].

In conclusion, it is vital that the attending clinician considers the potential causes of atraumatic compartment syndrome and a multidisciplinary approach with early intervention can improve both short- and long-term outcomes.

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