

Rare disease

Acute carpal tunnel syndrome in a patient with haemophilia

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Summary

Acute carpal tunnel syndrome (CTS) is a rare surgical condition usually resulting from wrist trauma. We present the case of a young haemophilic man who developed acute CTS following trivial injury. The patient was initially managed conservatively but symptom progression resulted in carpal tunnel decompression. A literature review and management approach are presented. This is an important complication of haemophilia to be aware of as prompt conservative management can obviate the need for surgery. This case is useful in (a) highlighting the importance of considering a patient's medical history when formulating differential diagnoses and (b) outlining a management approach to this condition.

BACKGROUND

Acute carpal tunnel syndrome (CTS) occurring as a consequence of haemophilia is rare, with only a handful of case reports in the literature. This patient was initially discharged from an emergency department following normal radiographs, before symptom deterioration resulted in re-presentation to a different emergency department hours later. This case therefore highlights the need to consider a patient's medical history when considering differential diagnoses. Various management approaches have been suggested in the literature—we present our experience based on the literature and this case.

CASE PRESENTATION

A 23-year-old man with haemophilia type A presented to the emergency department with a painful left hand following a fall on an outstretched hand at work. The patient had suffered bleeds in his groin and foot in the previous year. Radiographs revealed no fracture and he was discharged with simple analgesics. Pain progressed significantly, resulting in the patient re-presenting to a different hospital 8 h later with severe pain in his left hand which was not relieved by 30 mg intravenous morphine. On examination, the left hand was not particularly swollen but there was bruising over the volar aspect of the wrist and mild tenderness in this region. There was reduced sensation in the distribution of the median nerve, with normal motor function. There was no pain on passive stretching of the fingers.

Investigations revealed an activated partial thromboplastin time of 46.3 s (normal range 26–35 s) and factor VIIIc levels of 2.5%. The patient was transferred to our tertiary trauma institution and administration of recombinant factor VIII (Helixate) was started as a haematoma in the carpal tunnel was suspected.

The patient's symptoms deteriorated over the following days, with increasing signs of median nerve dysfunction. The patient underwent surgical decompression of the carpal tunnel, which revealed a large liquid haematoma

within the carpal tunnel, extending proximally and distally within the tendon synovial sheath.

OUTCOME AND FOLLOW-UP

The symptoms improved significantly postoperatively and are expected to completely resolve.

DISCUSSION

Acute CTS is much less common than chronic CTS and occurs when a rapid rise in pressure within the carpal tunnel results in median nerve symptoms developing over hours, rather than weeks to months as with chronic CTS. Acute CTS is usually the result of trauma to the hand and wrist, with atraumatic causes, including infection, rheumatological, haematological and vascular disorders being rare.¹

Haemophilia can cause haemarthroses and haematomas, with the latter potentially resulting in neurological complications and compartment syndrome.²

In haemophilic patients, symptoms of acute CTS often begin following trivial upper limb trauma, with severe pain and median nerve symptoms predominating. A thorough examination using the 'look, feel, move' approach is essential to help differentiate causes and guide investigations. On examination, patients may be tender over the volar aspect of the wrist, may have a positive Phalen's or Tinel's test, and will have evidence of median nerve dysfunction. The diagnosis requires careful clinical evaluation as initial clinical features may be subtle.

Differentials to be considered in haemophilic patients presenting with wrist pain include fractures, dislocations, haematoma causing compartment syndrome, haemarthroses and, as this case highlights, acute carpal tunnel syndrome.

Radiographs are useful in excluding a co-existing fracture, but do not show the presence or absence of a haematoma in the carpal tunnel. If a diagnosis of acute CTS is suspected, MRI of the wrist is advised as the primary investigation as, in addition to imaging the carpal tunnel,

Table 1 Case reports of acute carpal tunnel syndrome due to haemophilia

Authors	Reference number	Number of patients	Patient details, management and outcomes
Molitor and Wimperis	3	1	13-year-old boy, treated with human factor VIII, symptoms resolved and full recovery within 1 week
Rahimtoola and Van Baal	4	2	36-year-old man, presented with pain in fingers and wrist following trivial trauma Given Factor VIII and surgical decompression on day of admission. Complete recovery was made within 12 h 24-year-old man, pain and paraesthesia in hand following fall on outstretched hand. Surgical decompression on day of admission. Signs and symptoms resolved over 2 days
Adamson <i>et al</i>	5	1	16-year-old man, blow to wrist and developed pain and paraesthesia over hours Administration of fresh frozen plasma and surgical decompression, does not state when
Case	6	1	Initial conservative management with delayed surgical decompression, full recovery at 12 weeks
Ehrmann <i>et al</i>	7	2	Conservative management, both made full recovery
Lancourt <i>et al</i>	8	2	Case 1: treated with Factor VIII replacement for 5 days with a full recovery within 2 weeks Case 2: treated with immobilisation of the wrist for 2 weeks, no Factor VIII replacement due to previous anaphylactic reaction. 80% recovery by 3 months, with persistent mild atrophy and sensory deficit
Khunadorn <i>et al</i>	9	1	15-year-old man with severe haemophilia awoke with pain in left wrist and swelling on volar surface, no prior injury. Symptoms progressed, and median nerve symptoms developed. Treated conservatively with splint and cryoprecipitate. Pain quickly disappeared, swelling resolved over 2 weeks but median nerve did not have full recovery for 8 months.
Monheim and Gribble	10	2	Case 1: treated with factor VIII replacement therapy and full recovery made Case 2: initial management with Factor VIII replacement therapy. Surgical decompression and internal neurolysis (intra-neural bleeding) after failure of conservative management

this will allow diagnosis of intra-articular pathology and other problems. Alternatively, ultrasound scan, widely available in many centres, may be used. In haemophilic patients, it is important to check serum clotting factor levels for which they are deficient (ie, Factor VIII: C for haemophilia A and Factor IX for haemophilia B) along with the activated partial thromboplastin time.

Acute CTS occurring due to haemophilia is extremely rare and optimal management is debated. A literature search to establish optimal management of this condition was performed using PubMed. The case reports identified are summarised in table 1.

From the available literature and our own experience, we suggest that the initial management of acute CTS in patients with haemophilia should be conservative. Patients require early administration of recombinant factor VIII for 3–5 days in conjunction with elevation of the limb and immobilisation. Close clinical observation is required as if symptoms fail to improve or deteriorate, prompt surgical decompression of the carpal tunnel is required. A haematologist must be involved in the patient’s care as haematological advice is critical both during the non-operative period and peri-operatively (if surgical management is undertaken). In patients who undergo operative decompression, it is important that haematology remain involved in the patient management for many days after surgery to ensure no re-bleed.

It is important that both patients and doctors are aware of the need to promptly start factor VIII replacement therapy to prevent symptom progression and reduce the need for surgery.

Learning points

- ▶ Patients’ medical history must be considered when formulating differential diagnoses.
- ▶ Acute carpal tunnel syndrome should be considered in patients with haemophilia presenting with pain and/or neurological symptoms in the hand, even following trivial trauma.
- ▶ Early identification and initiation of recombinant clotting factors can avoid the need for surgery.

Competing interests None.

Patient consent Not obtained.

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Please cite this article as follows (you will need to access the article online to obtain the date of publication).

Mayne AIW, Howard A, Kent M, Banks J. Acute carpal tunnel syndrome in a patient with haemophilia. *BMJ Case Reports* 2012;10.1136/bcr-03-2012-6152, Published XXX

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