Case report: Acute cubital tunnel syndrome in a hemophiliac patient

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Abstract
A 24-year-old right-hand dominant male with severe Hemophilia A presented with acute elbow pain, associated paresthesias, and weakness in the ulnar nerve distribution after upper body weight lifting. In the week prior, he missed three doses of Factor VIII replacement. After no improvement with conservative measures, he was taken to the operating room urgently for decompression and was noted to have a perineural hematoma in the cubital tunnel. At final follow-up, the patient reported complete resolution of symptoms. Acute cubital tunnel syndrome in hemophiliac patients that does not respond to medical treatment is best treated with surgical decompression using minimal dissection to prevent hematoma formation, preserve perineural membranous tissue, and avoid destabilizing the nerve.

Keywords
Acute cubital tunnel syndrome, hemophilia, in situ decompression, perineural hematoma

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Introduction
Compression neuropathy of the ulnar nerve, commonly referred to as cubital tunnel syndrome, is the second most common compressive neuropathy in the upper extremity after carpal tunnel syndrome.1 Typically, clinical symptoms are chronic and slowly progressive. Acute compression of the ulnar nerve at the cubital tunnel is rare and infrequently reported in literature. Hemophiliac patients are prone to acute peripheral nerve entrapment from intramuscular, intraarticular, or intraneural hemorrhage with an incidence of 4%–19%.2–5 These acute nerve compression syndromes occur spontaneously or following minimal trauma and most commonly involve the femoral nerve.2–6 Involvement of the ulnar nerve occurs less frequently, with no true incidence reported in literature.4 Due to the infrequent presentation, optimal treatment has been debated.5,7,8

While there are several reports debating conservative versus surgical decompression for acute carpal tunnel syndrome in the hemophilic patient, there have only been two other case reports of acute cubital tunnel syndrome in hemophilic pediatric patients and only one other small case series of six patients with sub-acute to chronic cubital tunnel syndrome in hemophilic patients.2,4,6 All previous studies report surgical decompression and anterior ulnar nerve transposition when surgery was performed.4,6 We are reporting a rare case of acute cubital tunnel syndrome in a 24-year-old male hemophilic due to external compression from a perineural membrane hematoma treated with in situ surgical decompression without transposition.

Case
A 24-year-old right-hand dominant male with severe Hemophilia A presented to the hospital with a 3-day history of left medial elbow pain and swelling after lifting weights. He also described pain and paresthesias in the ring and small fingers. He was receiving Factor VIII replacement three times a week but reported missing the last three doses. He was receiving Factor VIII replacement three times a week but reported missing the last three doses.

On exam, the patient had moderate swelling and tenderness to palpation over the medial epicondyle with decreased sensation over the small and ring fingers in the ulnar nerve distribution. Intrinsic strength was decreased with 4/5 on the Medical Research Council (MRC) grading scale. There was a positive Tinel’s sign at the elbow. The compartments of the forearm were soft and compressible with no evidence of compartment syndrome. Factor VIII was administered to bring the patient’s levels over 100%. He was admitted for observation. His left arm was elevated in a stockinet. However, the

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following day, the patient’s symptoms continued to worsen, and he was taken to the operating room for surgical decompression.

The procedure was performed under a supraclavicular block with an additional field block using lidocaine with epinephrine to minimize bleeding. All sites of ulnar nerve compression around the elbow were decompressed. On exploration of the cubital tunnel, the ulnar nerve was noted to be compressed underneath the Osborne’s ligament due to a hematoma that had formed in the mesoneurium (perineural membrane) presumably originating from the medial triceps muscle (Figure 1). The internal structure of the nerve was not violated or involved. There was no neuroma noted or obvious constriction deformity as seen with chronic compression. There was no appreciable hemarthrosis.

Postoperatively, he was followed by hematology for maintenance of his Factor VIII levels. At the 6-week follow-up, the patient subjectively reported doing well without activity restrictions. His paresthesias had completely resolved, and grip strength returned to full 5/5. Written informed consent was obtained from the patient for which identifying information is included in this article.

Discussion

As classified by Wadsworth, external compression of the ulnar nerve at the cubital tunnel may be acute, subacute, or chronic. It may also lead to varying degrees of neuropathy from solely diminished sensation to both sensory and motor deficits. There are several well-documented sites of ulnar nerve compression that are released during surgical decompression. However, spontaneous hematoma or hemarthrosis formation increases the potential for nerve compression in the hemophiliac patient. In addition, the elbow is the second most common site for hemophilic arthropathy, and medial joint involvement is thought to put patients at increased risk for chronic ulnar neuropathy.

Although patients are at a greater risk for cubital tunnel external compression syndrome with hemophilic arthropathy, only a limited number of case reports have been described. Dumontier et al. evaluated peripheral nerve palsies in the upper limb of 17 patients with hemophilia, citing only three ulnar nerve palsies at the wrist and only one at the elbow. No specific cause or acuity was further described. Ehrmann et al. evaluated all peripheral nerve lesions in 234 patients with hemophilia citing four cases in which the ulnar nerve was affected, two at the elbow, one in the forearm, and one at the wrist. The causes included direct trauma through a glass door and multiple-elbow hemarthrosis over a period of 4 days, and the rest were unknown.

Only two case reports have described acute cubital tunnel syndrome in patients with hemophilia, both in children. In the first case report, the patient developed worsening sensory and motor dysfunction following a bump to his elbow while playing. He continued to worsen even after receiving Factor replacement and was taken for acute cubital tunnel release, noting to have an intra-neural hemorrhage. After surgery, the patient demonstrated both sensory and motor improvement. In the second case, Renwick and Moneim described a patient who developed an elbow hemarthrosis after a simple fall with 1 week of paresthesias and weakness. After failing to improve with Factor replenishment, he was taken to surgery and was found to have a hemorrhagic cyst compressing the nerve with a stalk communicating with the hemarthrosis, which was excised. The ulnar nerve was then decompressed and anteriorly transposed. After 3 months, his intrinsic strength and sensation improved and regained full strength and sensation at the 7-year follow-up.

While accurately diagnosing acute cubital tunnel syndrome or other acute compressive neuropathies in the hemophiliac patient may seem clinically apparent, determining the optimal treatment requires incorporating multiple variables and is not always straightforward. Historically, treatment has been largely conservative: administration of coagulation factors, limb elevation, and joint immobilization. Patients refractory to conservative management eventually have undergone surgical decompression with nerve transposition, ultimately yielding clinical improvement, but not always with complete resolution of their symptoms.

Mortazavi et al. reported a small case series of six patients with sub-acute (<6 months) to chronic (>6–12 months) cubital tunnel syndrome. Five patients underwent surgical decompression with subcutaneous anterior transposition. The other patient refused surgery and was treated conservatively. Due to the improved outcomes in the surgically treated patients, the authors recommended early surgical decompression.

To date, no case reports within the literature of adult acute cubital tunnel syndrome due to external compression to our knowledge have been described. Furthermore, the optimal surgical technique for adequate decompression of the ulnar
nerve has been debated, and recently, there has been a trend favoring in situ decompression over transposition.\textsuperscript{1} Most previously reported cases of hemophiliac patients with cubital tunnel syndrome underwent surgical decompression with anterior transposition, making our current case report unique.\textsuperscript{4,9}

In the current literature, there is no recommended standard treatment algorithm for acute cubital tunnel syndrome. While the initial repletion of Factor is undoubtedly necessary, the allotted time for observation as the initial treatment is debatable given the potential for irreversible or permanent nerve damage as the ulnar nerve has a poor long-term prognosis. In cases that do not improve promptly, urgent surgical decompression should be considered as recent evidence supports superior outcomes with less extensive exposures, decreasing the risk for adverse events.\textsuperscript{1} In general, in situ ulnar nerve decompression at the elbow is commonly performed by hand and upper extremity surgeons and considered a relatively low-risk procedure. Therefore, we believe a surgical release to be a low-risk treatment option in acute cubital tunnel syndrome. We also recommend simple in situ decompression as the surgical method of choice to preserve perineural membranous tissue and avoid destabilizing the nerve or creating potential hematoma formation.

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