Case Report / *Olgu Sunumu*

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The Treatment of Advanced Cubital Tunnel Syndrome in a Patient with Hemophilia and Its Postoperative Results

Bir Hemofili Hastasında İleri Evre Kübital Tünel Sendromunun Tedavisi ve Postop Sonuçları

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ABSTRACT

We would like to inform and guide other hand surgery physicians by sharing the results of a patient with ulnar nerve compression due to hemosiderin accumulation due to hemophilia. We operated a hemophilia patient who applied late and had ulnar nerve compression and we achieved a successful result. In this publication, the patient we operated on was presented as a case report and discussed in the light of literature.

Keywords: Ulnar nerve entrapment, hemophilia, nerve entrapment, claw hand, neuropathy

ÖZ

Kas-iskelet sistemi hemorajileri homofili hastalarında sık görülür. Hemofili hastalarında ulnar nöropati ile ilgili son yıllarda literatürdeki yayın sayısı çok sınırlıdır. Hemofilide periferik nöropatilerin en sık femoral sinirde görüldüğü belirtilmiş olsa da diğer periferik sinirlerde de görülebilmektedir.

Anahtar Kelimeler: Ulnar sinir sıkışması, hemofili, sinir sıkışması, pençe el, nöropati

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INTRODUCTION

Musculoskeletal hemorrhages are common in patients with hemophilia. In this publication, we will describe the pre- and post-operative clinical condition of a patient with severe ulnar nerve compression around the elbow region due to hemophilia-related accumulation of hemosiderin and significant atrophy in areas innervated by the ulnar nerve.

PATIENT

A 25-year-old right-handed 55 kg 172 cm male patient presented with complaints of numbness, tingling, numbness and loss of strength in his left hand. He had claw hand deformity in the 4th and 5th fingers. There was no abduction-adduction in the fingers and there was atrophy of the interosseous and hypothenar muscles innervated by the ulnar nerve. Tinel, Wartenberg, Jeanne and Froment sign findings were positive. Two point discrimination measurements increased in the ulnar region, especially on the 5th finger and it was measured as 9mm.

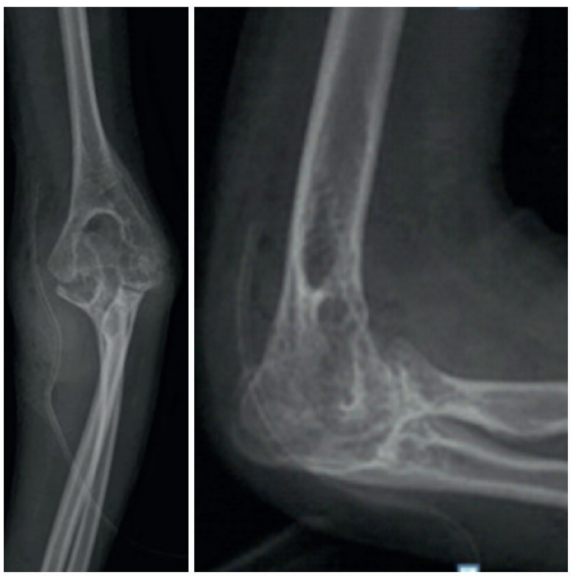

Figure 1

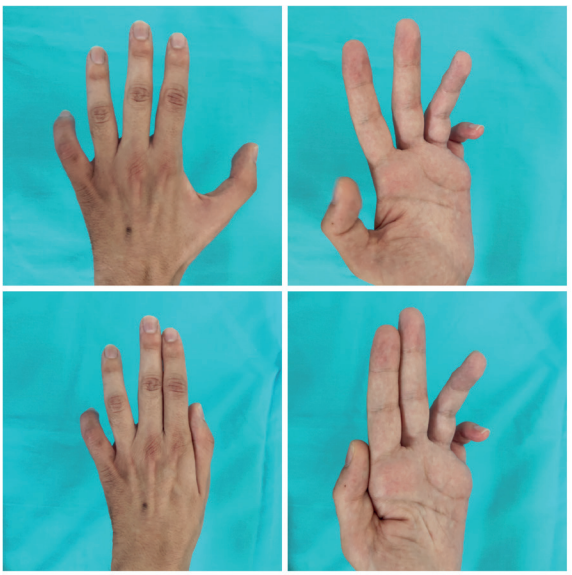

Figure 2

While MR and USG images showed a mass lesion formation due to accumulation in the ulnar nerve region at the level of the medial epicondyle, on USG it was found that the diameter of the ulnar nerve increased compared to the measurements of the healthy side, the mass was outside the nerve at the beginning, but the nerve could not be observed due to the compression of the mass after it passed through the medial epicondyle.

In EMG, ulnar motor conduction block was observed in the elbow segment. In addition to the ulnar neuropathy findings, there was a 50-degree limitation of extension in the elbow due to hemophilic arthropathy, and the flexion was 135 degrees. The patient with hemophilia A had factor VIII deficiency. Early surgery decision was made owing to the advanced complaints and findings. Factor VIII was given at 2500-unit doses before the operation. The patient underwent ulnar nerve neurolysis, anterior transposition and excision of the mass. The operation was performed under tourniquet control. He continued to take factor VII for 3 days after the operation. There was no abnormal bleeding in the dressing or

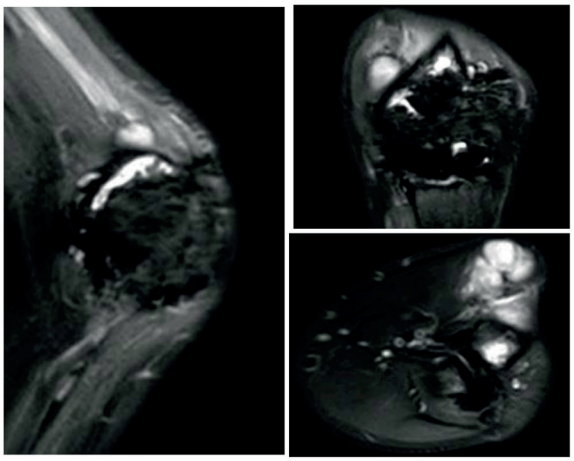

Figure 3

hematoma in the wound area. It was determined that ulnar neuropathy findings regressed on the 2nd postoperative day. In the follow-ups, the numbness disappeared, the loss of sensation and the claw hand deformity regressed. In the third month follow-up, the patient's complaints regressed significantly. VAS pain score decreased from 7/10 to 1/10. The atrophies were still partially ongoing, but the Wartenberg and Froment findings were negative. The two-point separation was 4mm. While Gabel and Amadio's Ulnar Nerve Rating Scale preoperatively was 2/9, on the second day post-operatively it was 3/9 and on the 3rd month it was 7/9.

METHODS

In tourniquet hemostasis, a longitudinal incision extending 8 cm proximal and 4 cm distal to the medial epicondyle was entered, and the subcutaneous mass was reached immediately. The ulnar nerve was separated from the mass and freed, its motor branches were exposed, and the mass was excised while preserving them. A part of the medial intermuscular septum was excised, and the ulnar nerve was transposed anteriorly to the subcutaneous tissue. In order to keep the ulnar nerve at the transposed site, a pocket was made from the surrounding soft tissue and covered loosely. No compression or folding was observed along the liberated nerve trace.

DISCUSSION

In recent years, the number of publications in the literature about ulnar neuropathy in patients with hemophilia is very limited. Therefore, we think that our case will contribute to the current literature. Although it has been stated that peripheral neuropathies are most commonly seen in the femoral nerve in hemophilia, they can also be seen in other peripheral nerves. In the study of Ehrmann L. et al., the number of patients with ulnar nerve lesion was shown as 4 among 36 hemophilia patients with peripheral nerve lesions [(1)]. Besides, cases of carpal tunnel syndrome due to compression in the median nerve in patients with hemophilia have also been shown [(2)].

In peripheral nerve compression due to acute bleeding, if the symptoms are mild, coagulation factor supplementation and immobilization of the joint can be tried first to ensure bleeding control. In patients with severe symptoms, surgical decompression is recommended. Debkowska MP et al. stated that in this case, there was no response to medical treatment alone. In such cases, they recommended the prevention of hematoma with minimal dissection to provide decompression of the nerve [(3)].

Cases caused by chronic compression are well treated with surgical treatment. Peripheral nerve compressions are usually seen secondary to intramuscular hematoma or hemarthrosis, but intraneural hemorrhages have also been reported [(4)]. Although we saw neural edema in our case, we did not observe intraneural hemorrhage. According to a study by Mortazavi SM et al. in 2010, a high rate of HIV and HCV positivity was

shown depending on the possible frequency of transfusion. In this publication, 6/6 patients were positive for HCV and 4/6 patients were HIV positive [5]. In our case, HIV and HCV serology was negative, but we think that surgeons planning such operations should pay maximum attention to viral serology results.

Arthropathy is also a very important problem in hemophilic patients. We think that advanced arthropathy due to recurrent intra-articular hemarthrosis, in addition to restriction of movement in the joint, compresses the nerve. Hogh J. et al. found in their study that 87% of hemophilic patients had radiographic evidence of elbow arthropathy [6]. The pathological mechanism underlying the development of hemophilic arthropathy is quite complex and is not yet fully understood. The two main processes that play a role in the pathogenesis of arthropathy are inflammation of the synovial membrane and cartilage degeneration [7,8]. The presence of blood in the joint induces chondrocyte apoptosis and has a direct corrosive effect. As a result of recurrent bleeding in the same joint, erythrocyte-derived hemosiderin accumulates in synovial macrophages and triggers synovial inflammation, causing the synovium to become hypertrophied [9]. The joint is deformed and range of motion is limited with a tendency to flexion contracture. These changes lead to joint destruction known as "chronic hemophilic arthropathy". In our case, flexion contracture continues even though the patient's ulnar neuropathy findings regressed.

CONCLUSION

We would also like to point out that although the complaints and findings of this patient were obvious, the patient visited the clinic very late so that clinical findings like nerve compression was in advanced stage. This patient's complaints had regressed significantly after the operation, if he had visited or been noticed earlier, he could have been treated before findings such as atrophy occurred. It carries an important role for physicians who frequently follow hemophilia patients, carefully examine neuropathy findings and be alert, in order to diagnose and intervene in such patients earlier. With early intervention, the results will be better.

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