

Case Report

Intraneural cavernous hemangioma: a rare case of extrafascicular left ulnar nerve tumor

Hadi Ravanbod¹, Mehdi Motiffard², Mohsen Aliakbari³, Mohammad Zolfaghari⁴, Saeed Hatami³

¹Assistant Professor of Orthopaedics, Department of Orthopedic Surgery, School of Medicine, Al-Zahra Hospital, Isfahan University of Medical Sciences, Isfahan, Iran; ²Associated Professor of Orthopaedic Surgery, Department of Orthopedic Surgery, Kashani University Hospital, School of Medicine, Isfahan University of Medical Sciences, Isfahan, Iran; ³Department of Orthopedics, School of Medicine, Isfahan University of Medical Sciences, Isfahan, Iran; ⁴Shahid Sadoughi University of Medical Sciences and Health Services, Yazd, Iran

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Abstract: Hemangiomas are benign soft tissue tumors that may be found everywhere in the human body. As one of the hemangioma types, cavernous hemangioma consists of a flat endothelium along with blood-filled spaces and may be found in the central nervous system, but rarely occurs in peripheral nerves. This article pertains to the introduction of an old female patient complaining of pain and paresthesia of the ulnar side of the left forearm and hypothenar with numbness and tingling of the fourth and fifth digits and clawing. The patient was medically treated for a month but became a surgical candidate due to the poor response to medical treatment. A 1-cm lesion was observed in the surgery with compression on the ulnar nerve in the ulnar groove. Neurologic symptoms of the patient were improved after excision of the lesion, but clawing persisted.

Keywords: Ulnar nerve, hemangioma, tumor, cavernous hemangioma, peripheral nerve

Introduction

Hemangiomas are non-cancerous mesodermal tumors developed following endothelial hyperplasia and abnormal growth in blood vessels, and only include 7% of all benign soft tissue tumors [1]. Hemangiomas can originate from embryonal hemangioblasts and neoangiogenesis, but their main etiology is still unknown [2]. Cavernous hemangioma is a kind of hemangioma composed of blood-filled spaces separated by a flat endothelium. Fatty tissues may also be found between cavernous hemangiomas.

Although cavernous hemangioma is often seen in the central nervous system and even could be life-threatening, it may exist everywhere in the human body such as the liver, skin, and retina. Nonetheless, there are few reports on the existence of cavernous hemangioma in peripheral nerves [3]. Hemangioma in peripheral nerves was reported for the first time by Sato in 1913. However, there are very limited studies on the ulnar nerve hemangioma in the English literature (**Table 1**). This type of hemangioma usually appears as a mass in the cubital

tunnel and may cause symptoms such as pain, paresthesia, and numbness in the upper extremities through compression syndrome [4]. In 2008, Dođramacı and colleagues reported a case of intraneural hemangioma of the median nerve in Turkey [5]. The same case was later diagnosed and reported by Kim and others in 2011 [6]. Later in 2018, a case of intraneural hemangioma of the tibial nerve was reported in the USA [7]. It is believed that cases of intraneural hemangiomas are very rare but require precise diagnosis and treatment strategies. Surgical procedures such as dissection and excision of the lesion are used for the treatment of intraneural hemangioma. Nerve graft may be also used depending on the size of the hemangioma. Post-op recurrence rarely occurs in this case [8]. In the present study, a female patient with symptoms of pain and paresthesia of the forearm, hypothenar, and digits is reported.

Case report

A 64-year-old right-handed housewife woman visited the outpatient orthopedic clinic of Al-Zahra Hospital (Isfahan, Iran) in February

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Table 1. Reviewing of the literature on the ulnar nerve hemangioma

Authors and year	Age/Sex	Involved nerve	Side	Diameter (mm)	Treatment
Prater (2017) [9]	18 y/F	ulnar nerve (Guyon canal)	Left	10	surgical excision
Prasad (2016) [13]	13 y/M	ulnar nerve (at elbow)	Right	nm	surgical excision
	18 y/F	ulnar nerve (at elbow)	Left	nm	surgical excision
Jafari (2015) [4]	40 y/F	ulnar nerve (cubital tunnel)	Left	15×10	surgical excision
Brand (2015) [14]	15 y/F	ulnar nerve	Left	20×13×7	surgical excision
Fnini (2013) [15]	39 y/F	ulnar nerve (forearm)	Right	nm	surgical excision
	22 y/F	ulnar nerve (forearm)	Right	nm	surgical excision
Kim (2009) [16]	47 y/M	ulnar nerve (multilevel)	Right	multiple	surgical excision
Kon (1993) [17]	57 y/M	ulnar nerve (volar aspect of wrist)	Left	35×20	surgical excision
Kline (1992) [18]	63 y/M	ulnar nerve (cubital tunnel)	nm	20	surgical excision
Linde (1982) [19]	16 y/M	ulnar nerve (at elbow)	Left	80×50	surgical excision
Losli (1952) [20]	49 y/M	ulnar nerve	Left	43×33	surgical excision

2020 complaining of pain and paresthesia of the ulnar side of the left forearm and hypothenar with numbness and tingling of the fourth and fifth digits of the left hand started about 4 months ago. Within 4 months, the patient had fixed and progressive pain. The severity of pain increased with bending the elbow within the first 20 days. The patient reported more severe pain in full flexion of the elbow and also reported pain in the ulnar nerve and disability after 3-4 minutes of phone calls (a condition that her elbow was fully flexed). But she reported fixed pain in the past 3 months non-related to physical activities and hand motions. She also claimed severe issues in grasping. The patient acknowledged that the pain was increasing occasionally woken her at night. By the time of admission, there was no notable point in the past history of medical or surgical procedures, and the patient did not take medication routinely. She also mentioned no history of trauma. The neurologic examination revealed positive distal tingling to percussion (Tinel's sign) in the ulnar groove, and poor thumb adduction and abduction, and adduction of other digits. Physical examinations of the median nerve showed full flexion of first, second and third digits. Extension of fingers and sensation of the first dorsal web was performed with no problem indicating no issues in radial nerve. Intrinsic muscle atrophy of the hand and clawing were also observed in the fourth and fifth digits. The active and passive range of motion of the fourth and fifth digits were restricted but the range of motion of the elbow was normal, and there was no swelling, palpable mass and skin changes. Physical examinations related to limb pulses

showed no issues and had a normal capillary refill test (CRT) showing normal arterial conditions in the affected hand.

There was no abnormal finding in the elbow and wrist X-ray, but ulnar nerve involvement in the cubital tunnel was observed in the electrodiagnostic examination. Laboratory tests also revealed normal results.

The cubital tunnel syndrome was diagnosed according to clinical and laboratory evidence. It should also be noted that imaging studies including ultrasound and magnetic resonance imaging (MRI) were not performed because the case had typical clinical manifestations related to cubital tunnel syndrome. The patient was treated for one month with a 15 mg meloxicam tablet and 100 mg gabapentin capsule. Due to the poor response to the treatment, the patient became a surgical candidate. Open surgery was performed on the left arm of the patient with general anesthesia using a pneumatic tourniquet. The ulnar nerve was released on the medial aspect of the elbow through a 10 cm linear skin incision. A dark red tumoral oval shape tissue with an approximate size of 4×10 cm was observed with severe adhesion to the nerve (**Figure 1A**). The epineurium was cut and the tumoral tissue was removed without structurally damaging the nerve fibers using microsurgery (**Figure 1B**) and sent to the pathology laboratory. The continuity of the nerve was preserved properly in this technique. The subcutaneous transposition of the ulnar nerve was performed and a long arm splint was taken for a week. In the pathology report, large cystic dilat-

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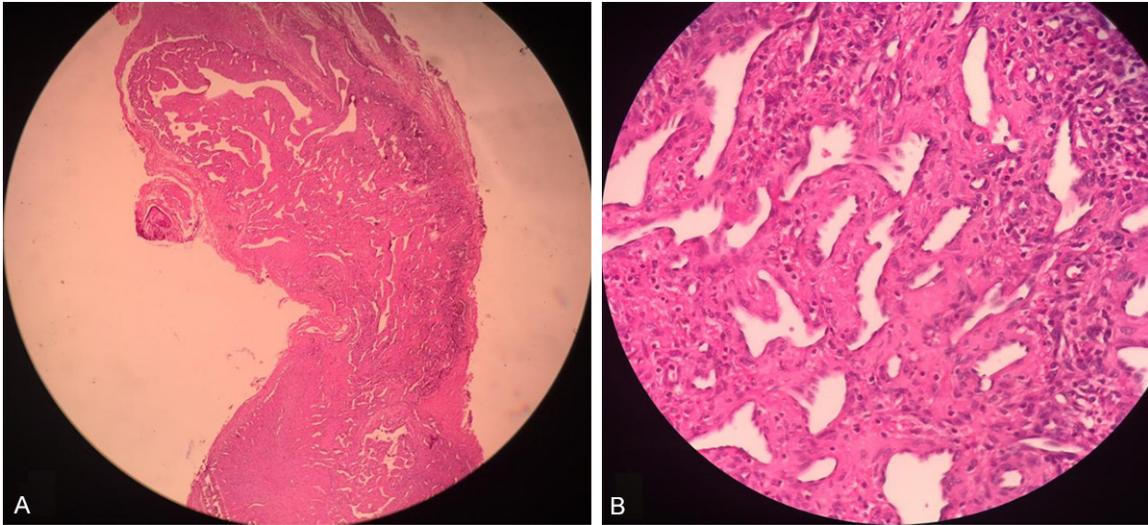


Figure 1. A. Intraneural hemangioma (white arrow) after cutting the epineurium of the ulnar nerve (green arrow). B. The tumoral tissue removed after surgical excision.

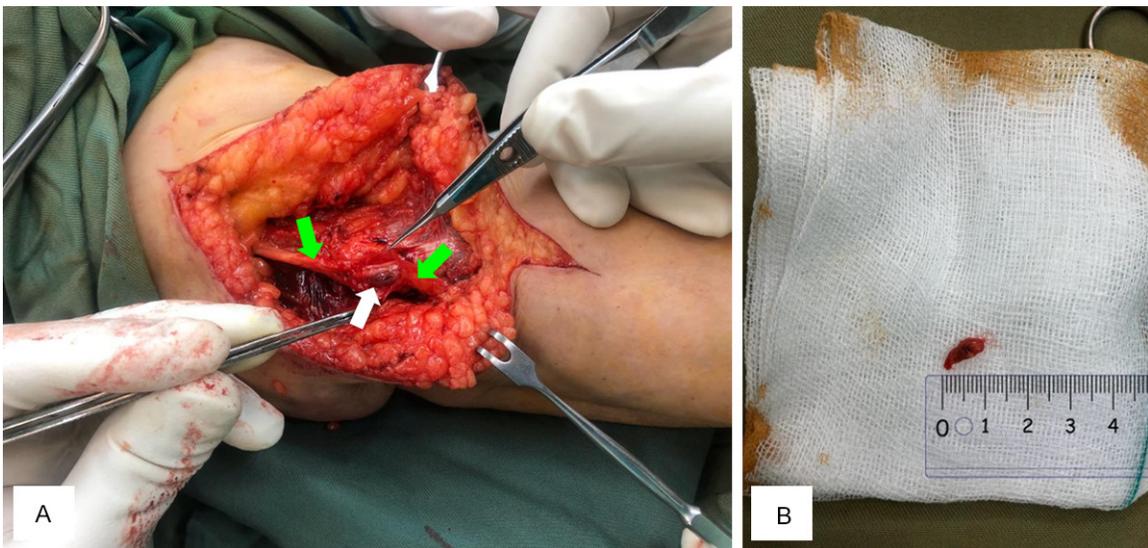


Figure 2. A. Hematoxylin and Eosin staining ($\times 100$), large cystic dilated vessels in the background of fibroconnective tissue. B. Hematoxylin and Eosin staining ($\times 200$), tortuous endothelial cells of different sizes indicative of intraneural hemangioma.

ed vessels covered by endothelial cells were seen in the background of the fibroconnective tissue without any malignancy indicative of cavernous hemangioma (**Figure 2A, 2B**). The patient was discharged within 48 hours after the surgery with a long arm splint and bandage. After 2 weeks, the splint was opened, the sutures were removed and limited passive range of motion initiated. After 1 month, physical examinations were performed showing partial improvements in sensory examinations but

limitations in hand movements and hand deformity could be observed. Within 3 months after the operation, full recovery of paresthesia and partial improvements in hand deformity were noticed but he declared limitations in grasping. After 6 months, he could hold objects with third and fourth fingers against gravity and no signs of paresthesia and positive Tinel's sign were reported. Both active and passive range of motion in the elbow were completely normal and the patient had no pain.

Discussion

Posttraumatic hematoma, glomus tumor, and angioleiomyoma are among differential diagnoses raised alongside hemangioma [16]. A hemangioma may be found anywhere in the human body. Cavernous hemangioma usually occurs in the central nervous system. Structurally, it is a single layer of endothelial cells that form dilated blood-filled vessels [9]. Cavernous hemangioma rarely occurs in peripheral nerves. Bacigaluppi and others (2018) reported 41 cases of intraneural hemangioma in peripheral nerves. The mean age of patients was 23.4±17.3 years with the most involvement in the median nerve (36.6%). The male to female ratio was 1:17, and the right side of the body was more involved than the left side (61.5%). Of 33 follow-up cases, recurrence was reported in 7 cases, and there was no report on the follow-up of patients for the other 8 cases. Among the cases, only 10 cases of cavernous hemangioma of the ulnar nerve were reported indicating the rare occurrence of intraneural hemangioma in this nerve [10]. Vascular malformations are classified into three types based on nerve involvement: Type 1: intraneural extrafascicular malformation which is highly resectable through microsurgical techniques, Type 2: intrafascicular with a risk of damage to the nerve during resection due to adhesion of neural fascicles, and Type 3: malformations with both intraneural and extraneural components [11].

Electrodiagnostic studies may play a key role in the localization of the lesion and surgical approach. The reported patient suffered from a decrease of left ulnar compound muscle action potential (CMAP) amplitude and severe axonal ulnar neuropathy, suggesting the presence of a lesion in the elbow considering the positive Tinel's sign in the ulnar groove. Given that the hemangioma growth near neural fibber causes neurological symptoms through compression syndrome, surgery is one of the most recommended therapies for such lesions. In most cases, the patient's symptoms are improved by excision of hemangioma. Pain, paresthesia, and other neurological symptoms in our case were immediately improved postoperatively, clawing of the hand however still persisted. The use of microsurgical techniques may significantly reduce nerve injury. In some cases, based on the hemangioma size, nerve segment resection may be required simultaneously with excision of the lesion, and this defect requires

interfascicular nerve grafting [8, 12]. In cases where hemangioma is not large but is anatomically resectable, the lesion can be resected with minimum nerve injury. In our case, the tumoral tissue was resected without damage to the nerve fascicles through epineurium incision, and no new nerve deficit was observed postoperatively. Despite the low postoperative recurrence rate, the patient will be followed-up for at least one year to report any probable recurrence.

In conclusion, despite the high prevalence of vascular lesions, in the case of compression syndrome on peripheral nerves along with unexplained pain and paresthesia, such lesions can be classified as differential diagnoses. A complete history and neurological and electrodiagnostic examinations may play a key role in the localization of the lesion, but the definitive diagnosis of the disease will be performed with the help of histopathology. We also suggest that imaging studies including ultrasound and MRI could be useful for better diagnosis of the pain etiology. Surgical techniques should be selected to preserve nerve fascicles with minimum damage to the nerve during excision of the lesion.

Acknowledgements

This study was approved by our institutional review board. Even though there were no identifying details about the patient in this study, informed consent was obtained for inclusion in the study.

Disclosure of conflict of interest

None.

Address correspondence to: Mehdi Motififard, Associated Professor of Orthopaedic Surgery, Department of Orthopedic Surgery, Kashani University Hospital, School of Medicine, Isfahan University of Medical Sciences, No. 22, Khorsand Street, Hezar Jarib Blv., Isfahan 6719675344, Isfahan Province, Iran. Tel: +989188400108; E-mail: pnasiri689@gmail.com

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