DOI: 10.7860/IJARS/2024/68585.2984

Case Report

Radiology Section

Ulnar Neuropathy caused by Repeated Thrombophlebitis of the Slow-flow Venous Malformation in the Cubital Tunnel: A Case Report

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ABSTRACT

Cubital tunnel syndrome is the most common entrapment neuropathy of the ulnar nerve at the elbow. Although there are many primary and secondary causes of cubital tunnel syndrome, ulnar neuropathy secondary to recurrent thrombophlebitis of the slow-flow Venous Malformation (VM) in the vicinity of the cubital tunnel has not been reported. Hereby, authors discuss a case of 22-year-old female with slow-flow VM in the vicinity of the cubital tunnel causing symptoms of transient ulnar neuropathy, progressing to ulnar neuritis resulting in persistent ulnar neuropathy. This was successfully treated by direct Stick Sclerotherapy of the VM, leading to complete relief of the symptoms. Follow-up Magnetic Resonance Imaging (MRI) revealed near-total resolution of the VM with complete resolution of the ulnar neuritis. This was an atypical presentation of ulnar neuropathy secondary to recurrent thrombophlebitis of a slow-flow VM in the cubital tunnel, which was successfully managed by treating the malformation.

Keywords: Sclerosant, Sclerotherapy, Ulnar nerve, Venogram

CASE REPORT

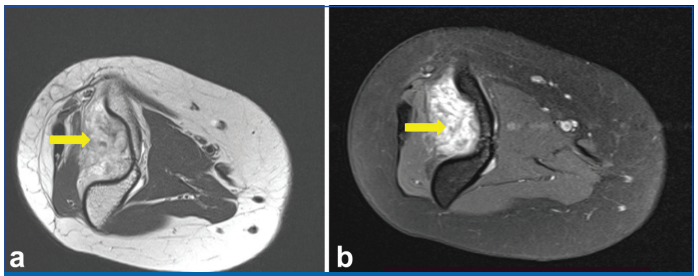
A 22-year-old female was referred to the Interventional Radiology Outpatient Department (OPD) from the Department of Orthopaedics for symptoms of repeated attacks of pain and paraesthesia in the medial aspect of the right elbow, forearm, and the ring and little fingers for the past month, with associated swelling in the posteromedial aspect of the elbow extending above to the distal one-third of the arm. These symptoms were recurrent and self-limiting over the past three weeks and had become persistent in the past week before the presentation. No aggravating or relieving factors were noted.

The patient had a history of similar complaints over the past year, during which the patient was unable to move the elbow joint and experienced pain along the medial aspect of the forearm with a Visual Analogue Scale (VAS) of 8/10. The symptoms would resolve spontaneously within a week. There were no significant co-morbidities, no family history of similar issues, and no history of trauma.

On examination, swelling measuring approximately 5×4 cm was noted in the posteromedial aspect of the right elbow, with associated tenderness in the region of the cubital tunnel. Power in the right forearm was 5/5 with palpable peripheral pulses and a normal range of movements.

The patient underwent an ultrasound elsewhere a few months ago, which raised suspicion of a slow-flow VM at the cubital tunnel. The scanned copies and images of these reports are unavailable. The patient was conservatively managed with analgesics. The swelling at the elbow resolved, but the symptoms of pain and paraesthesia in the medial aspect of the elbow, forearm, and the ring and little fingers (ulnar neuropathy) persisted with two to three attacks per month. An MRI of the elbow was recommended to assess the ulnar nerve and evaluate the VM.

The MRI was performed using a 3T Siemens scanner, and fast SE T1WI, SE T2WI, SE T1WI + FS, SE T2WI + FS, and Gradient-recalled-echo (GRE) T2WI sequences were obtained. The MRI revealed a T1 mildly hyperintense, T2 and Short Tau Inversion Recovery (STIR) hyperintense lesion measuring 3.7×3.1×1.5 cm in the posterior aspect of the right elbow, between the triceps insertion and the posterior cortex of the humerus, extending to the cubital fossa and abutting the ulnar nerve in the cubital fossa. Dynamic contrast enhancement was subsequently performed using 3D fast GRE technique, in which the lesion demonstrated progressive and delayed intense heterogeneous enhancement consistent with slow-flow VM [Table/Fig-1a,b].

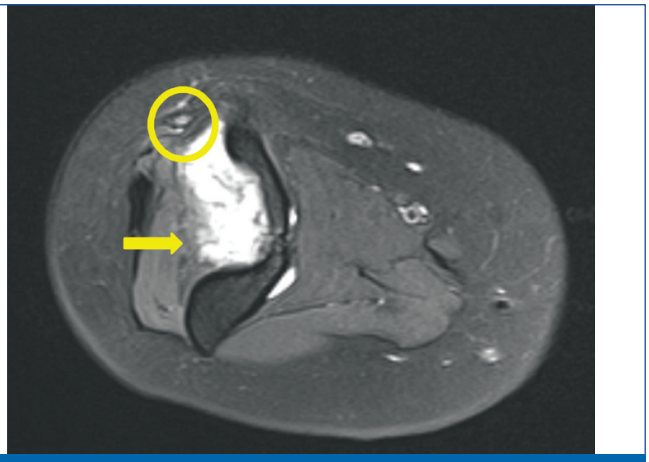


[Table/Fig-1]: a) Axial T2W image at the level of elbow demonstrating well-defined heterogeneous hyperintense lesion within the triceps muscle, just anterior to the triceps tendon (arrow); b) Axial postcontrast T1FS image demonstrating intense heterogeneous enhancement of the lesion (arrow).

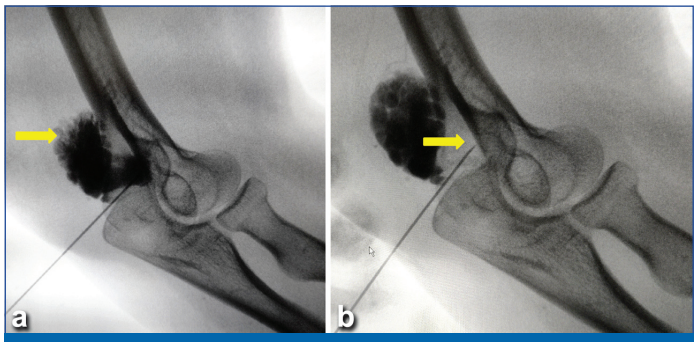
The ulnar nerve was thickened and demonstrated T2 hyperintensity, suggesting ulnar neuritis [Table/Fig-2], secondary to repeated compression of the ulnar nerve during episodes of recurrent thrombophlebitis of the VM.

Direct stick sclerotherapy was planned for the VM. Under ultrasound guidance, using a 22 G Lumbar Puncture (LP) needle, the VM was punctured, and 50% diluted iodinated contrast media was injected (direct stick venogram), revealing Puig and Dubois Type I VM with a total volume of 7 mL [Table/Fig-3a] [1].

Under aseptic technique, local anaesthesia and ultrasound-guided sclerotherapy were performed using an intralesional injection of 6 mL of 3% Sodium Tetradecyl Sulphate (STS) (by preparing 6 mL of foam Tessari in a 1:3 ratio of STS: air) [Table/Fig-3b]. Postsclerotherapy, the patient experienced transient symptoms of ulnar neuritis for six hours, apart from local pain and swelling, which were conservatively managed.

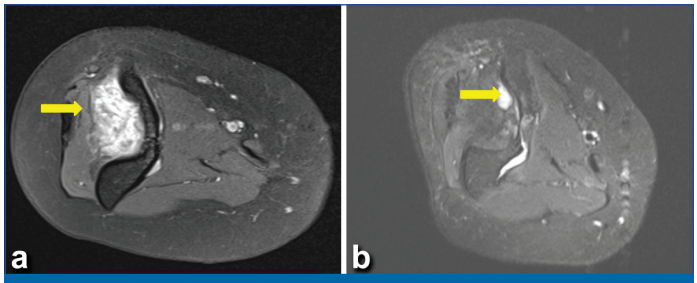


[Table/Fig-2]: Axial Proton Density Fat Saturated (PDFS) image at the level of elbow demonstrating well-defined heterogeneous hyperintense lesion within the triceps muscle, just anterior to the triceps tendon, consistent with Venous Malformation (VM) (arrow). There is PDFS hyperintensity within the ulnar nerve, which is in close association with the posterior aspect of the lesion (circle).

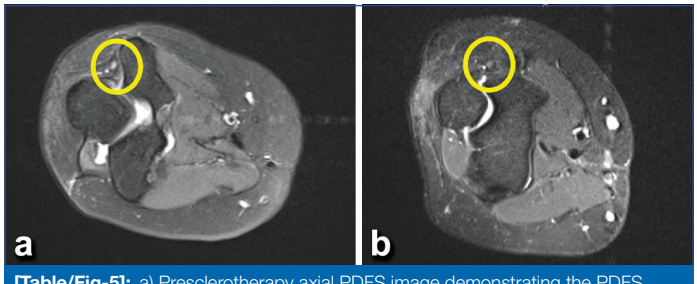


[Table/Fig-3]: a) Flouroscopy demonstrating direct stick venogram using 22G LP needle demonstrating intense contrast blush and stasis filling the Venous Malformation (VM) without any significant draining veins (arrow); b) Flouro-guided sclerotherapy with STS foam which can be seen as filling defect within the contrast stasis (arrow).

In the three-month follow-up, the patient was completely stable without any episodes of ulnar neuropathy. In the follow-up MRI, there was near-total regression in size (1.5×1×1 cm) and extent of slow-flow VM in the posterior fossa and cubital tunnel [Table/Fig-4a,b]. Furthermore, the MRI also revealed complete resolution in the abnormal signal intensity of the ulnar nerve at the cubital fossa, secondary to relief in the compression caused by the VM [Table/Fig-5a,b].



[Table/Fig-4]: a) Presclerotherapy axial PDFS image demonstrating the Venous Malformation (VM) (arrow); b) Postsclerotherapy axial PDFS image demonstrating near total resolution in the Venous Malformation (VM) with minimal residual lesion (arrow).



[Table/Fig-5]: a) Presclerotherapy axial PDFS image demonstrating the PDFS hyperintensity of the ulnar nerve at the cubital tunnel, suggesting ulnar neuritis (circle); b) Postsclerotherapy axial PDFS image demonstrating complete resolution in the abnormal signal intensity within the ulnar nerve (circle).

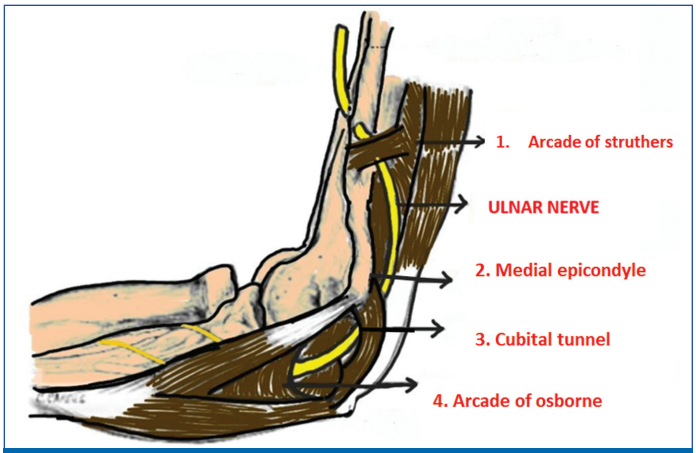
DISCUSSION

The VMs are slow-flow, congenital vascular anomalies secondary to a congenital error in vascular embryogenesis, resulting in the formation of abnormal endothelium and thin-walled, non functional, and ectatic veins/venous spaces [2,3]. Most of them result from sporadic mutations, with the majority of phenotypes arising from endothelial dysgenesis through the TIE2–PI3K (phosphoinositol-3-kinase)–AKT–mTOR (mammalian target of rapamycin) pathway [4].

The most common symptom is pain, which can occur due to various reasons, including local compression of adjacent structures such as muscles, joints, and nerves; congestion and thrombosis due to venous stasis within the malformation, suggestive of thrombophlebitis, sometimes leading to phleboliths (small calcific thrombi); and bleeding into surrounding tissues and joints. VMs may enlarge and become symptomatic during puberty, pregnancy, and trauma [5]. Without treatment, VMs generally slowly increase in size and are the most common symptomatic vascular anomalies referred for intervention, usually due to pain caused by these complications. When located in the extremities, these symptoms may worsen with activity. Rarely, there may be pulmonary embolism in the context of thrombophlebitis of the VM if there is communication with the deep venous system [5,6].

Cubital tunnel syndrome is the most common entrapment neuropathy of the ulnar nerve at the elbow level [7]. Patients typically complain of numbness and pain along the 5th and ulnar half of the 4th digit. Compression of the ulnar nerve at the elbow also leads to sensory symptoms on the dorsum of the ulnar side of the hand in the distribution of the dorsal cutaneous branch of the ulnar nerve and on the ulnar side of the palm in the distribution of the palmar cutaneous branch of the ulnar nerve. Patients may also report pain in the elbow and forearm. As the condition progresses, patients may experience weakness and clumsiness of the hand due to malin nervation/denervation of the intrinsic lumbricals of the hand [8]. In advanced stages, atrophy of the interosseous muscles and the adductor pollicis muscle, as well as claw hand deformity, may occur.

The cubital tunnel is located posterior to the medial condyle of the humerus along the posteromedial aspect of the elbow and comprises three parts: the retrocondylar groove, the humero-ulnar arcade, and the deep flexor aponeurosis. The retrocondylar groove is partially enclosed by the humeroulnar arcade. The humeroulnar arcade, also known as the Osborne ligament, passes into the aponeurosis between the two heads of the flexor carpi ulnaris muscles and is also referred to as the Cubital Tunnel Retinaculum (CTR). The deep flexor aponeurosis, or flexor-pronator aponeurosis with multiple variable bands, constitutes the third component [Table/Fig-6] [9].



[Table/Fig-6]: A schematic diagram representing the possible anatomical locations of ulnar nerve compression at the elbow. These are (from proximal to distal): 1) Arcade of Struthers; 2) Medial epicondyle; 3) Cubital tunnel; 4) Arcade of osborne.

Risk factors for cubital tunnel syndrome include subluxation of the nerve, accessory muscles like anconeus epitrochlearis, and hypertrophy of the medial head of the triceps muscle. The secondary

form can result from a distal humerus fracture, osteoarthritis at the elbow with osteophytes in the cubital tunnel, or inflammatory arthritis of the elbow where proliferative synovium causes symptoms. Other causes include soft-tissue lesions such as lipoma, Intra-articular/ para-articular ganglion cysts, inflammatory processes in the cubital tunnel or adjacent areas, and nerve sheath tumors [10].

Due to its high resolution for soft tissues, MRI remains the gold standard for VM imaging, while diagnostic ultrasonography and venography can provide useful information about the blood flow distribution and path in VMs. MRI can also be used to monitor the treatment effect of VMs [11].

Sclerotherapy is now the first-line treatment for slow-flow VMs under image guidance, either using Ultrasound (USG) or phlebography. Commonly used sclerosants include dry ethanol, polidocanol, ethanolamine oleate, Sodium Tetradecyl Sulphate (STS), sodium morrhuate, Picibanil (OK-432), bleomycin, and doxycycline prepared using Tessari's technique. Multiple sessions may be required for satisfactory results, and lesions may recur, but overall symptom reduction rate, quality of life, and patient satisfaction are significantly improved [12].

Muchemwa FC et al., reported an unusual case of intramuscular VM with calcifications that presented with features of involvement of the ulnar nerve [13]. A similar study by Kelley N et al., revealed two case studies of vascular anomalies in the cubital tunnel as causes of ulnar nerve compression and concluded that vascular anomalies are a rare but potential cause of ulnar nerve compression and impairment [14].

A case report by Magnussen JS et al., described an atypical variant of Reflex Sympathetic Dystrophy (RSD) in a 45-year-old female with a vascular malformation of the right arm and chest wall. The mechanism was thought to be secondary to compression of the brachial plexus by the malformations [15].

Altenbernd J et al., concluded that sclerotherapy is a promising way to treat slow-flow malformations, with patients reporting a reduction in symptoms including pain (57.7%), swelling (65.4%), functional impairment (60%), and cosmetic complaints (44.4%) [16].

A retrospective study by Kim H et al., concluded that VMs of the extremities affect patients' quality of life due to their appearance and pain. Sclerotherapy was performed in 16.7% of cases, and clinical outcomes have been excellent for localised VMs [17].

Pain, swelling, and ulcerations are common local complications. However, pulmonary embolism, stroke, vision loss, compartment syndrome, tissue necrosis, haemolysis, anaphylaxis, nerve palsy, and pulmonary complications have been known to occur [12].

CONCLUSION(S)

The present case is a rare instance of Cubital tunnel syndrome caused by repeated thrombophlebitis of the slow-flow VM in the vicinity of the cubital tunnel. The episodic ulnar neuropathy symptoms, which appeared during the episodes of thrombophlebitis, gradually progressed to continuous symptoms with changes of ulnar neuritis on imaging. There was complete relief of the ulnar neuropathy after three weeks of single-session direct stick sclerotherapy, with near-total regression of the lesion as well as complete resolution of the signal changes of the ulnar nerve at the cubital tunnel in the 3-month follow-up MRI.

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PLAGIARISM CHECKING METHODS: [Jain H et al.]
- Plagiarism X-checker: Nov 23, 2023
- Manual Googling: Jan 08, 2024
- iThenticate Software: Feb 15, 2024 (12%)

ETYMOLOGY: Author Origin

EMENDATIONS: 6

AUTHOR DECLARATION:
- Financial or Other Competing Interests: None
- Was Ethics Committee Approval Obtained for this study? Yes
- Was informed consent obtained from the subjects involved in the study? Yes
- For any images presented appropriate consent has been obtained from the subjects. Yes

Date of Submission: **Nov 11, 2023**
Date of Peer Review: **Jan 08, 2024**
Date of Acceptance: **Feb 16, 2024**
Date of Publishing: **May 01, 2024**