Ulnar tunnel syndrome caused by neurofibroma in the elbow: A case report

Abstract

Purpose:compression of the ulnar nerve at the elbow is the second nerve entrapment syndrome of the upper limb, often idiopathic, neurofibroma of the ulnar nerve is an exceptional cause of this syndrome.

Case Report: A 55-year-old man presented with progressive numbness and weakness of the rightring finger and the right little finger for one year. A provisional diagnosis of cubital tunnel syndrome was made according to physical examination and electrophysiological studies. The magnetic resonance imaging showed a nodular lesions over ulnar aspect of the right elbow, which was demonstrated to be a neurofibroma by histopathology. The patient had moderate improvement after surgical decompression of the right cubital tunnel.

Conclusion: This case illustrates the heterogeneous group of pathologies causing peripheral neuropathy. The diagnosis of ulnar nerve neurofibroma with ulnartunnel syndromewas made carefully according to clinical manifestations as well as a series of electrophysiological, imaging, and pathological studies.

Key Words: ulnar-tunnel syndrom, ulnar nerve, nerve compression, neurofibroma.

1. INTRODUCTION :

The syndrome involving compression of the ulnar nerve at the elbow represents the second most common nerve entrapment syndrome of the upper limb. Although history and physical examination are essential parts of the diagnosis, nerve entrapment syndrome is primarily diagnosed through electrophysiological examination. It is often idiopathic, with tumors being a rare cause, predominantly synovial cysts (1, 2, 3).

We report a case of ulnar nerve syndrome at the elbow secondary to a neurofibroma of the ulnar nerve, which showed significant improvement following surgical neurolysis.

2. CASE PRESENTATION

Mr.AS, 55 years old, civil servant, with no notable pathological history. Presented with paresthesia of the 4th and 5th right fingers evolving for a year, initially nocturnal then becoming diurnal.

The clinical examination found a positive Tinel sign at the level of the epitrochleo-olecranon groove, a Phalen sign at the elbow and an ulnar nerve with a parasitized superficial cutaneous sensitivity (S2) and a preserved motor function (M5).

Furthermore, the morphological appearance of the elbow is normal with normal mobility. No clinical signs of Van Recklinghausen disease. The rest of the upper limb examination is normal. Faced with this clinical picture, a syndrome of the ulnar nerve at the elbow is suggested.

The elbow's x-ray shows no osteoarticular lesions or architectural narrowing of the elbow.

The electrophysiological examination of the ulnar nerve revealed severe damage to the ulnar nerve in the right elbow (conduction velocity=23.6 m/s and latency=8.7ms)

A secondary etiology has been suggested for this syndrome given the absence of predisposing elements. So we deepened our paraclinical examination.

Ultrasound showed a swollen appearance of the right ulnar nerve which is compressed by two oblong cystic formations at the entrance and exit of the epitrochleo-olecranon groove(Fig.1).

The MRI found a swollen right ulnar nerve, it is in T2 hyper signal and covered by two cystic lesions(Fig.2).

Faced with these different aspects, an intrinsic tumoral cause of compression of the ulnar nerve was suggested, requiring exploration and surgical neurolysis.

The patientwas operated on conventionally. The exploration found, after opening the epitrochleo-olecranon gutter, the Struthers arcade and theOsborne arcade, a swollen ulnar nerve, with a tortuous appearance, the site of multiple cystic formations.

Exoneurolysis was carried out with biopsy of the cystic lesions and the perinervus (Fig.3,4). The histopathological examination was in favor of a myxoid neurofibroma (Fig.5).The outcome after surgical treatment was good with disappearance of paresthesia.

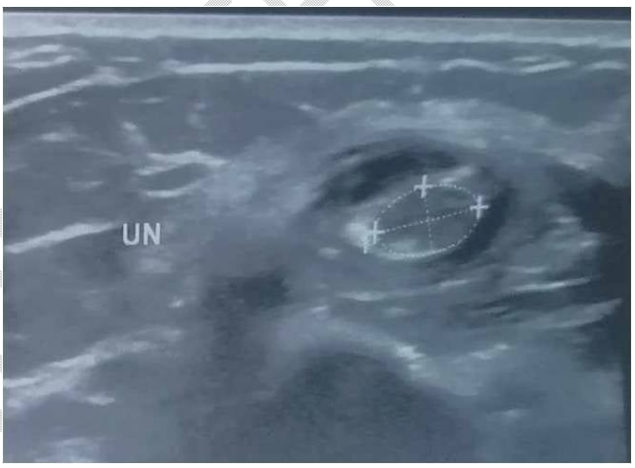

Fig1. Ultrasound appearance

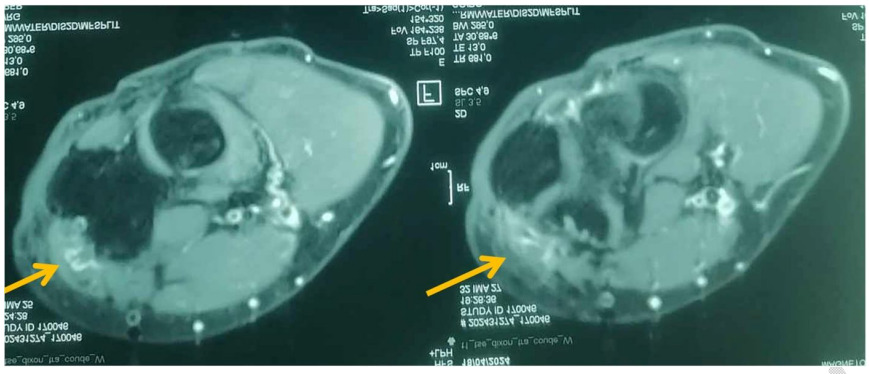

Fig2. MRI appearance

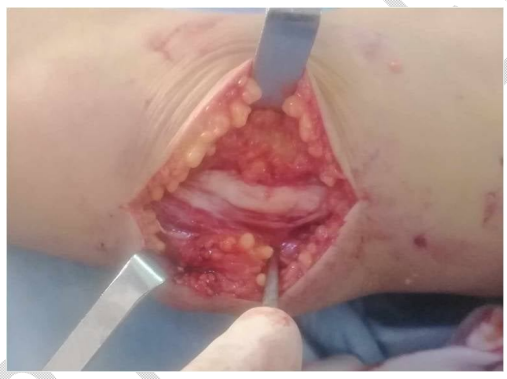

Fig 3. Intraoperative view of the ulnar nerve at the level of the epitrochleoolecranon groove

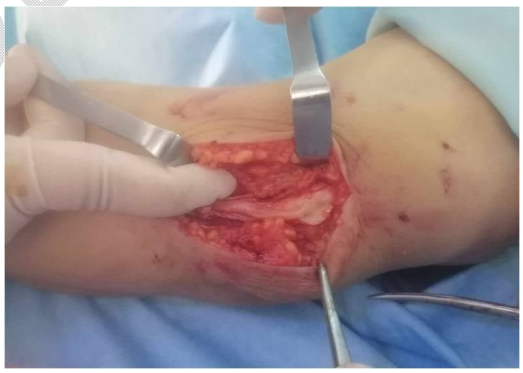

Fig 4. Appearance of the nerve after neurolysis and biopsy

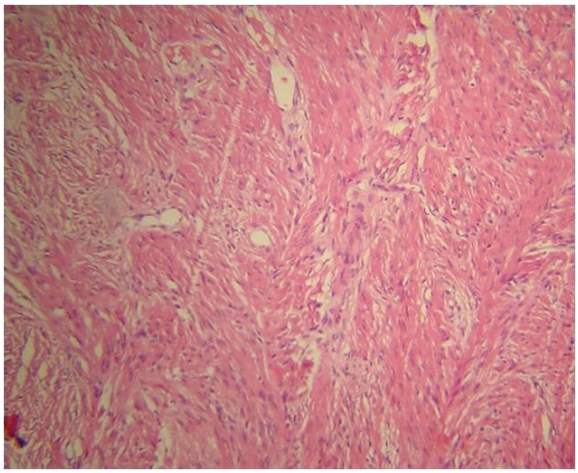

Fig 5. Image showing the Histopathological appearance of neurofibroma :Microscopy (H&E Stain, ×100), indicating a spindle cell tumor with a fibromyxoid background of neurofibroma.

3. DISCUSSION :

"The secondary Cubital tunnel syndrome will, in many cases, occur as tardy ulnar nerve palsy, following a previous injury of the elbow joint. Especially a malunited distal humerus fracture is leading to deformities such as cubital valgus or varus, osteoarthrotic changes with exostoses. In addition, secondary CuTS occurs in an unstable elbow joint as seen in rheumatoid arthritis, often with proliferative synovium and in heterotopic ossification seen with old trauma". [12]"Other less common causes include osteochondromatosis, aneurysmatic bony cysts or osteoarthrotic changes of Paget's disease. Finally, primary soft tissue lesions such as lipoma, ganglia, thickened veins or venous plexus, cysts, and inflammatory processes in the cubital tunnel or its vicinity causeCuTS. Space occupying processes like neurilemmomas or neurofibromas within the cubital tunnel occur very rarely. Peripheral polyneuropathy, which may be related to age, osteoarthritis, or diabetes, may predispose some patients to CuTS" (4,5).

Schwannomas and neurofibromas of the ulnar nerve are benign encapsulated tumors. With a peak incidence between 3-5th decade, without gender predilection, often located at the level of the head and neck, while the peripheral location is unusual representing 10% of cases, which attests to the rarity of our case. These tumors occur in the context of Von Recklinghausen neurofibromatosis. Levi et al. reported that 12 of 34 patients with neurofibroma had no association of neurofibromatosis [6], making our case of interest which illustrates a neurofibroma apart from neurofibromatosis. The complementary examination of choice for this type of tumor is MRI to locate and determine the size of the tumor and its extent of nerve infiltration; which is crucial for good preoperative planning. Management of these tumors consists of excisional biopsy and neurolysis. Often by conventional route, however it can be done endoscopically. With a good result after excision of any bulky lesion the epitrochleo-olecranialgottiere (7).

Plexiform neurofibroma is a benign tumor of the perineurium of the peripheral nerves. It is made up of a proliferation of Schwann cells arranged in a myxoid tissue of variable abundance including numerous fibroblasts and collagen fibers with a so-called "grated carrot" appearance [8]. The presence of antineurofilament antibodies within the proliferation can help in the diagnosis. The myxoid background is of variable abundance. The cellular shape can mimic a schwannoma but in immunohistochemistry, the S100 protein is expressed to a lesser extent than in schwannomas. A more myxoid form can resemble a myxoma. Plexiform neurofibroma is strongly associated with neurofibromatoses [2], in particular with Von Recklinghausen disease (NF1) where it occurs in 24 to 32% of affected patients [8,9]. The imagery is not very specific. Ultrasound still makes it possible to eliminate a tumor or a rapidly circulating vascular malformation. MRI has the classic appearance of peripheral nerve tumors (low to moderately intense T1 signal, T2 hypersignal often with a

heterogeneous appearance). The areas of hyperintensity correspond to areas of myxoid or cystic degeneration tissue. The nodular areas of hyposignal correspond to collagenous and fibrous tissue and can be enhanced with gadolinium. The risk of sarcomatous degeneration justifies, whenever technically possible, excision of the lesion as completely as possible. Finally, it is appropriate to carry out, at best by a multidisciplinary team, clinical and radiological monitoring of these patients, at least annually until the age of ten, then regularly, in order to assess a possible recurrence or malignant transformation (10, 11).

4. CONCLUSION :

When faced with ulnar nerve syndrome at the elbow, we must always think of a secondary etiology, even if it is rare; radiological investigations allow the diagnosis and conventional surgical treatment gives better results.

Consent

As per international standards or university standards, patient(s) written consent has been collected and preserved by the author(s).

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REFERENCES :

1. Meng-Ta Liu , Jiunn-Tay Lee , Chih-Hsin Wang , Yu-Chieh Lin, Chung-Hsing Chou. Cubital Tunnel Syndrome Caused by Ulnar Nerve Schwannoma in a Patient with Diabetic Sensorimotor Polyneuropathy Acta Neurol Taiwan 2016;25:60-64.

2.Leila Hamadou, Sofien Benzarti, Mohamed Ghorbel, Amine Badr, Mohamed Amine Triki, Thabet Mouelhi, Mohamed Laaziz Ben Ayache. Résultats fonctionnels du traitement chirurgical dans le syndrome canalaire du nerf cubital au coude. J.Neurol.2022.02.131

3. M. Allagui, M.F. Hamdi, A. Fekih, M. Koubaa, I. Aloui, A. Abid. Le syndrome canalaire du nerf ulnaire au coude d'origine idiopathique : à propos de 20 cas.Chirurgie de la Main 2023 ; Volume 32, Issue 2 : 74-79.

4. Forthman CL, Blazar PE. Nerve tumors of the hand and upper extremity. Hand Clin 2004;20(3):233-242.

5. Woojin Shin, Taebyeong Kang, Jeongwoon Han. Cubital tunnel syndrome associated with previous ganglion cyst excision in the elbow: a case report. Clin Shoulder Elbow 2024;27(1):131-135.

6.Levi A, Ross AL, Cuartas E, Qadir R, Temple HT. The surgical management of symptomatic peripheral nerve sheath tumors. Neurosurgery. 2010 Apr;66(4): 833-40.

7. H. Assmus, G. Antoniadis, C. Bischoff, R. Hoff mann, A. -K. Martini, P. Prei ß ler, K. Scheglmann, K. Schwerdtfeger, K. D. Wessels, M. W ü stner-Hofmann. Cubital Tunnel Syndrome –A Review and Management Guidelines. Cen Eur Neurosurg 2011; 72: 90 – 98.

8. Ali Tabrizi,Ahmadreza Afshar,Iraj Mohebbi,Masoumeh Pourjabali and Hassan Taleb. Schwannoma and Neurofibroma, Originating from the Ulnar Nerve in Neurofibromatosis: A Case Report and Review of the Literature. Surg J (N Y). 2020 Jul; 6(3): e139–e144.

9. Trăistaru R, Rogoveanu O, Popescu R, Enăchescu V, Ghiluşi M. Periarticular diffuse neurofibroma of the upperlimb. Rom J MorpholEmbryol. 2011;52(04):1377–1383.

10. Gonzalez-Darder JM, Pesudo-Martínez JV. Surgical treatment of multiple neurofibromas of the ulnar nerve in segmental neurofibromatosis. J Neurosurg.1999.90.6.1137.

11. X. Paredes-Carnero and J. A. Álvarez-Jorge. An Ulnar Neurofibroma: Management with Sural Graft Reconstruction. J Hand Microsurg. 2015 Dec; 7(2): 332–334.

12. Assmus H, Antoniadis G, Bischoff C, Hoffmann R, Martini AK, Preissler P, Scheglmann K, Schwerdtfeger K, Wessels KD, Wüstner-Hofmann M. Cubital tunnel syndrome–a review and management guidelines. Central European Neurosurgery-ZentralblattfürNeurochirurgie. 2011 May;72(02):90-8.

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