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Case Report

An unusual cause of posterior interosseous nerve palsy and contribution of ultrasonography to electromyography in a patient with neurofibroma

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ABSTRACT

Neurofibroma, a benign peripheral nerve sheath tumor, represents a rare cause of posterior interosseous nerve syndrome. Electrodiagnostic studies may not identify the exact site of nerve compression, a possible lesion that compresses the nerve and do not provide information about the morphological changes. Ultrasound is a cost-effective, practical modality that provides the opportunity for dynamic tracking in the peripheral nerves, and it is widely considered as the initial imaging modality for peripheral nerves. Herein, we report a case of posterior interosseous nerve palsy in a 13-year-old boy with neurofibroma of posterior interosseous nerve diagnosed with ultrasound. The benefit of ultrasound in localizing and determining the etiology of the posterior interosseous nerve palsy is emphasized in this case report. A meticulous ultrasound examination is recommended in suspected peripheral nerve lesions, regardless of the results of electrophysiological and imaging modalities.

Keywords: Neurofibroma, peripheral nerve sheath tumors, posterior interosseous nerve syndrome, ultrasonography.

Posterior interosseous nerve (PIN) palsy, characterized by weakness of the wrist and digital and thumb extensor muscles, can be caused by traumatic and atraumatic pathologies. Among the atraumatic causes, repetitive overuse, external compression, and spontaneous entrapment neuropathy are reported.[1] Neurofibroma, a benign peripheral nerve sheath tumor (PNST), represents a rare cause of PIN syndrome.[2] Electrodiagnostic studies may not identify the exact site of nerve compression, a possible lesion that compresses the nerve, and does not provide information about the morphological changes. In recent years, ultrasonography (USG) has been accepted to be a useful method for the visualization of multiple morphological abnormalities of even small nerves such as the PIN.[3-5] Herein, we report an exceedingly rare case of PIN neurofibroma causing PIN palsy, demonstrating the importance of sonographic evaluation and its contribution to electroneuromyography in peripheral nerve pathologies.

CASE REPORT

A 13-year-old male presented with a seven-month history of extension weakness in the right-hand fingers. There was no complaint of sensory disturbance or pain. The patient denied any previous traumatic history. The patient had previously visited an orthopedic surgeon and was referred to the electrophysiology department with the prediagnoses of cervical radiculopathy and radial nerve palsy. The cervical magnetic resonance imaging (MRI) results were normal.

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In electrophysiological evaluation, subacute partial axonal injury of the PIN was diagnosed. The nerve conduction studies are summarized in Table 1. Needle electromyographic examination revealed abnormal spontaneous activity in the form of positive sharp waves and fibrillation potentials with normal motor unit configuration and reduced recruitment in the right extensor digitorum communis. The right triceps, deltoid, and brachioradialis muscles were normal.

The patient was then referred to our physical medicine and rehabilitation outpatient clinic for ultrasonographic visualization of the radial nerve and the rehabilitation program. Physical examination revealed extension weakness in the patient's right second through fourth digits (2/5) and the fifth digit (3/5) at the metacarpophalangeal joints, and extension and abduction weakness was observed in the right thumb (3/5) with no other neurological deficits. The sensory examination was normal. There was no tenderness at the cervical spine, and Spurling's maneuver was negative. Tenderness of the right elbow, just distal to the right lateral epicondyle, was also detected.

In USG, a well-defined, hypoechoic, fusiform swelling of the PIN with a diameter of 2.4×7.0×3.2 mm just before its entrance into the supinator muscle was present in both short and long-axis views (Figure 1). There was no vascularity on color Doppler. The patient has primarily been diagnosed with the PNST of the PIN, most likely neurofibroma, and MRI was planned for further diagnostic evaluation. In the elbow MRI focusing on the lesion site, a fusiform swelling of the PIN was also detected. Surgery was planned with the

TABLE 1
Nerve conduction studies

Nerve stimulated	Amplitude Motor: mV Sensory: μV	Latency (ms)	Conduction velocity (m/sn)
Right median nerve (M)			67
Wrist (APB)	9.4	2.6	
Elbow (APB)	8.8	6.0	
Left median nerve (M)			64
Wrist (APB)	12.8	2.6	
Elbow (APB)	11.8	6.0	
Right ulnar nerve (M)			60
Wrist (ADM)	9.3	2.0	
Elbow (ADM)	8.3	5.8	
Left ulnar nerve (M)			60
Wrist (ADM)	10.3	2.2	
Elbow (ADM)	8.9	6.0	
Right radial nerve (M)			
Forearm (EIP)	0.5	3.8	
Left radial nerve (M)			
Forearm (EIP)	8	2.7	
Right median nerve (S)			
Wrist (Index finger)	63	3.08	
Left median nerve (S)			
Wrist (Index finger)	64	2.84	
Right ulnar nerve (S)			
Wrist (Little finger)	13	2.68	
Left ulnar nerve (S)			
Wrist (Little finger)	12	3.2	
Right radial nerve (S)			
Forearm (Snuffbox)	30	2.3	
Left radial nerve (S)			
Forearm (Snuffbox)	30	2.0	

M: Motor; S: Sensory; APB: Abductor pollicis brevis; ADM: Abductor digiti minimi; EIP: Extensor indicis proprius.

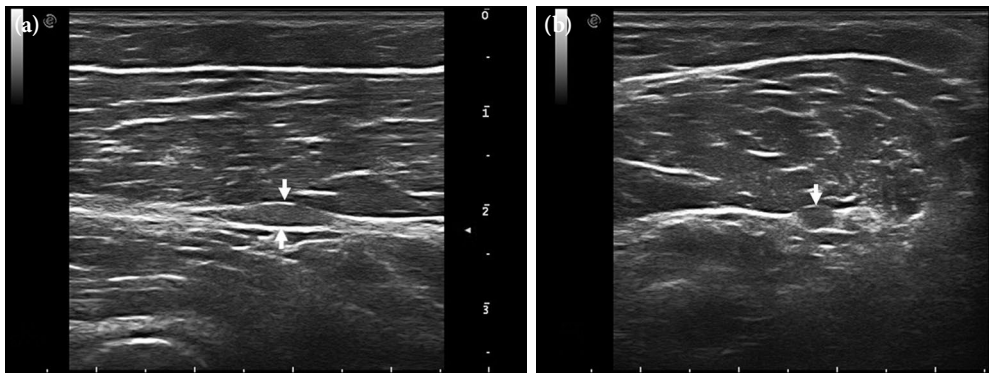

Figure 1. Longitudinal **(a)** and transvers **(b)** views of the PIN. A well-defined, hypoechoic swelling just before its entrance into the supinator muscle is shown with arrows.

PIN: Posterior interosseous nerve.

prediagnosis of neurofibroma of the PIN. In addition, ocular, dermatologic, and cranial evaluations were performed for further investigation, and no pathology was detected.

In the surgery, a dorsoradial incision at the proximal forearm was performed to explore the PIN at the supinator muscle entrance. Fusiform enlargement of the nerve was detected without any response to nerve stimulation. Extensor muscles of the fingers were pale, and atrophy was evident. The fusiform mass was resected, and no nerve repair was done since there was apparent muscle atrophy of the extensor compartment innervated by the PIN (Figure 2). Alternatively, the tendon transfer of flexor carpi radialis to the extensor digitorum communis and palmaris longus to the extensor pollicis longus was performed. The histopathological study of the resected mass confirmed the diagnosis of neurofibroma. A hand rehabilitation program was then started in the postoperative period.

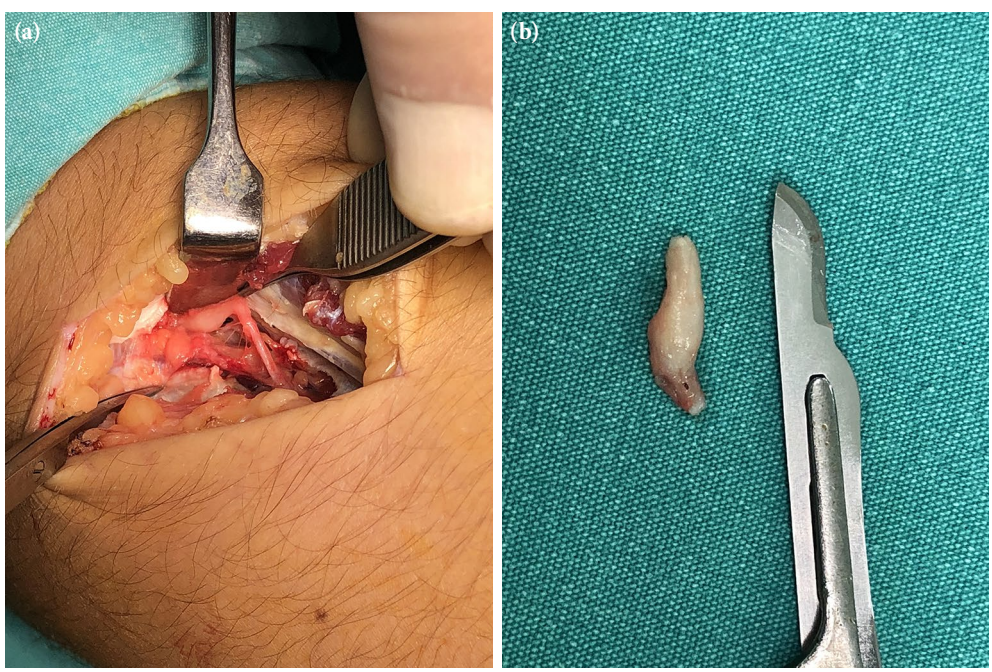

Figure 2. **(a)** Intraoperative photograph demonstrating the mass in the PIN. **(b)** The resected fusiform mass.

PIN: Posterior interosseous nerve.

DISCUSSION

Posterior interosseous nerve compression neuropathy may occur at multiple sites along its course, most frequently under the arcade of Frohse.[6] In addition to anatomical structures, mass lesions can also cause compression of the PIN. Inflammatory or neuralgic amyotrophy and spontaneous fascicular hourglass constriction of the nerve are the other reported causes of atraumatic PIN palsy.[7] Neoplasms of the PIN may also cause this rare clinical entity.

Nerve entrapment syndromes are usually diagnosed with electrodiagnostic testing. Although these analyses have the advantage of localizing the point of the lesion and assessing its severity, imaging studies are superior in identifying the cause of the entrapment.[3] In comparison with MRI, USG may better detect small peripheral nerve pathologies, which are often fusiform in shape and can extend along the length of the nerve without greatly altering its cross-sectional area. Moreover, USG is a cost-effective, practical modality that allows the dynamic tracking of the peripheral nerves, and it is widely considered as the initial imaging modality for peripheral nerves.[8] In this case, USG showed that it was not only an entrapment but also a peripheral nerve sheath tumor was present. Therefore, we referred the patient to a surgeon instead of an unnecessary rehabilitation program.

Differential diagnosis of atraumatic PIN palsies includes entrapment neuropathy, Parsonage-Turner syndrome, spontaneous hourglass constriction, and PNST.[1] Ultrasonographic diagnosis of a PNST is based on the presence of a solid hypoechoic mass in direct continuity with the nerve at its proximal and distal poles.[9] Most of the PNSTs are reported as hypoechoic, display posterior acoustic enhancement, and demonstrate intrinsic blood flow on color Doppler.[10] While USG allows for an accurate assessment of the presence of PNSTs, differentiation between schwannoma and neurofibroma can be challenging. Central location within the nerve and lacking a capsule help to differentiate neurofibromas from schwannomas, which tends to be located eccentrically.[10] Neurofibromas are also less hypervascularized on color Doppler than schwannomas.[9] Based on these significantly different findings, the sonographic distinction of these pathologies is possible.[11]

Schwannomas and neurofibromas are benign PNSTs, which are derived from Schwann cells.[12] Neurofibromas, which are the most common PNSTs, may present as solitary lesions, classified as localized neurofibroma.[1] They may also present as multiple serpentine-like masses found along the tract of a nerve and classified as plexiform neurofibroma.[13] Localized intraneural neurofibromas are the most common form and occur mostly sporadic; however, plexiform neurofibromas are often associated with neurofibromatosis. To the best of our knowledge, there has been only one report of neurofibroma causing PIN palsy.[2]

In conclusion, the localization and severity of the nerve lesion were detected by electroneuromyography; nevertheless, it was insufficient to identify the etiology. Both USG and MRI revealed that the tumor originated from the PIN and was most likely a neurofibroma. This rare case of PIN neurofibroma is a prime example of the contribution of USG to electroneuromyography.

Patient Consent for Publication: A written informed consent was obtained from the parent of patient.

Data Sharing Statement: The data that support the findings of this study are available from the corresponding author upon reasonable request.

Author Contributions: DP, KA, MKE and AA carried out the experiment; DP, TOM wrote the manuscript with support from RT; KA and AA helped supervise the project.

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