ESETISMERTETÉS
CASE REPORT

Patient with recurrent neuralgic amyotrophy; right brachial plexitis and left posterior interosseous neuropathy

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Neuralgic amyotrophy (NA), also known as Parsonage-Turner syndrome or idiopathic brachial plexopathy, is a multifocal inflammatory neuropathy that usually affects the upper limbs. The classic picture is a patient with acute onset of asymmetric upper extremity symptoms, excruciating pain, rapid onset of multifocal paresis often involving winged scapula, and a monophasic course of the disease.
We present an unusual case of recurrent NA characterized first by right brachial plexitis and then isolated left posterior interosseous nerve palsy.

Keywords: recurrent neuralgic amyotrophy, brachial plexitis, posterior interosseous nerve (PIN)

Visszatérő neuralgikus amyotrophiás beteg; jobb oldali brachialis plexitis és bal oldali hátsó interossealis neuropathia

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A neuralgikus amyotrophia (NA), más néven Parsonage–Turner-szindróma vagy idiopathiás brachialis plexopathia egy multifokális gyulladásos neuropathia, ami általában a felső végtagokat érinti. A klasszikus képet a betegnél akutan kezdődő aszimmetrikus felső végtagi tünetek, kínzó fájdalom, gyorsan kialakuló, gyakran elálló, „szárnyas" lapockát eredményező, multifokális paresis és monofázisos lefolyás jellemzi.
Bemutatunk egy szokatlan esetet, amikor a visszatérő NA-t először jobb oldali brachialis plexitis, majd izolált bal oldali nervus interosseus posterior bénulás jellemezte.

Kulcsszavak: rekurrens neuralgikus amyotrophia, brachialis plexitis, nervus interosseus posterior (PIN)

Neuralgic amyotrophy (NA) is characterized by neuropathic pain attacks and subsequent patchy paresis in the upper extremities. Although the exact mechanism of NA is not yet known, it is thought that the brachial plexus or its branches may develop idiopathic inflammation in response to intrinsic or extrinsic conditions, resulting in extreme pain at symptom onset and rapid paresis of upper extremity muscles[1]. NA is associated with immune-mediated disorders of the peripheral nervous systems; the extent and distribution of affected nerves are quite variable. The distribution of abnormalities in NA can vary from an isolated nerve (suprascapular, long thoracic, and anterior interosseous nerves) to the widespread involvement of the brachial plexus[2].

PIN paralysis can usually result from direct injury, repeated overuse of wrist and forearm muscles, compression, mass effect tumors, and sometimes NA.

The purpose of this case report is to present a rare case of recurrent NA that is characterized by right brachial plexitis and left PIN palsy despite being negative for the SEPT 9 gene.

Case report

A 43-year-old male patient was presented with severe right neck and shoulder pain that began 7 days ago and woke him from sleep. His medical history and family history were unremarkable. Previously, there was no triggering cause such as fever, infection, or physical activity. After 5 days, the severe pain subsided, but the patient was unable to raise his right arm. On neurologic examination according to the Medical Research Council (MRC) muscle strength on the examination scale, right arm abduction was 0/5, right arm external rotation was 0/5, right arm flexion was 4/5, and right arm extension was 4/5. The patient was primarily suspected of having a cervical disc herniation, and magnetic resonance imaging (MRI) of the cervical spine was performed. No compressive lesion was noted on the cervical MRI.

The nerve conduction study did not reveal sensory nerve action potential (SNAP) in the right lateral antebrachial cutaneous nerve. The right medial antebrachial cutaneous nerve SNAP, median and ulnar nerve SNAP and compound muscle action potential (CMAP) were normal. Electromyography performed on day 15 showed signs of abnormal spontaneous activity in the right deltoid and infraspinatus, and there was no motor unit potential (MUP). MUP loss was observed in the biceps, brachioradialis, pronator teres, and triceps muscles. Electrophysiological findings were consistent with the involvement of the superior and middle trunks of the right brachial plexus. Increased focal thickness and enhancement in the right brachial plexus were found to be significant for plexitis **(Figure 1)**.

He was diagnosed with NA mainly at the upper and middle trunks of the brachial plexus and was treated with high-dose steroid (7 days of pulse therapy) and active physical therapy. After a few months, partial movement of the shoulder was observed.

The patient presented severe left arm pain and an inability to lift the fingers of his left hand 4 months after his first application. The patient was examined on the 3th day of his complaint. The neurological examination according to the MRC scale of muscle strength on the examination of the extension of the left fingers were 1/5. Electrophysiological findings showed a complete conduction block between the forearm and elbow on the left radial nerve, with recording from the extensor indicis proprius muscle **(Figure 2)**. Sensory findings of the left superficial radial nerve and sensory and motor findings of the median and ulnar nerves were normal. On the control EMG performed 2 weeks later, CMAP was not detected in the left radial nerve. Total denervation was noted in the left extensor indicis proprius and extensor digitorum communis muscles. The extensor carpi radialis muscle was normal. Electrophysiologic findings were consistent with total axonal degeneration of the left posterior interosseous nerve in the acute phase. Because the conduction block detected on electrophysiological testing on day 3 disappeared on day 15, it was considered a pseudoblock. For the left PIN lesion, an MRI of the forearm was performed in the early phase and no compression was detected.

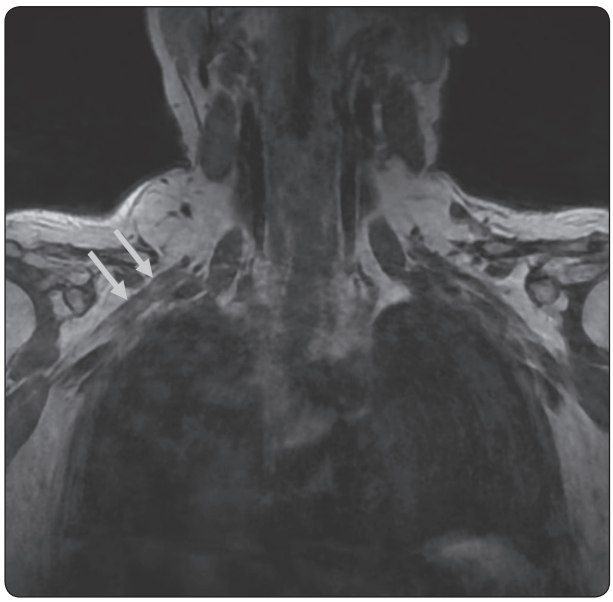

Figure 1. *Focal thickness increase and contrast enhancement in the right brachial plexus on T2-weighted MRI image*

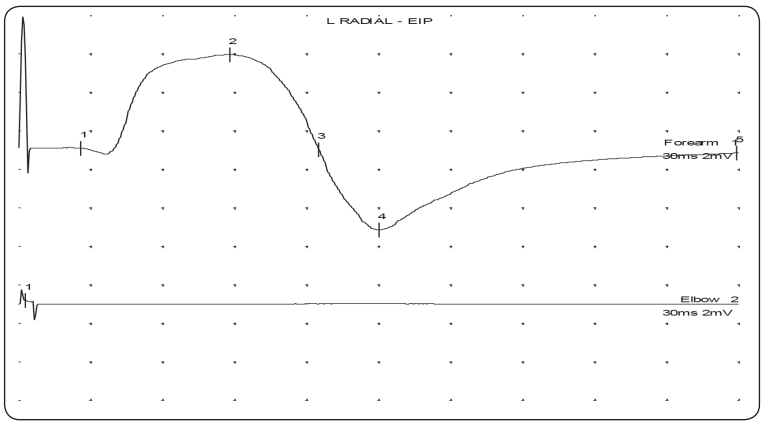

Figure 2. *Motor conduction of the left radial nerve (pseudo-block) with the recording from the EIP muscle; A normal CMAP response is obtained with stimulation from the distal to the radial tunnel (upper trace), whereas no response is obtained with stimulation from the proximal elbow (lower trace)*

Serologic testing, including autoantibodies and vasculitis markers, revealed no significant findings. HEV serology was negative. The patient with a history of recurrent NA (with brachial plexitis and isolated PIN lesion) underwent SEPT9 testing for hereditary neuralgic amyotrophy, which was negative.

The patient received IVIG for 5 days and treatment with 64 mg prednol was started. The patient partially benefited from the treatment and follow-up is continued.

Discussion

The classic presentation of neuralgic amyotrophy is a patient with acute onset of asymmetric upper extremity symptoms with excruciating pain during most attacks, rapid multifocal paresis often involving a winged scapula, and a monophasic disease course[3]. After the pain is disappeared, NA usually presents as the involvement of the superior brachial plexus, with weakness of the peri-scapular and glenohumeral muscles, including the infraspinatus, supraspinatus, serratus anterior, and deltoid.

The etiology of NA is unclear and the pathophysiology of the upper part of the brachial plexus, which is frequently affected, is still unclear. *Alfen* et al. report that NA affects the brachial plexus due to erosion and weakening of the blood-nerve barrier, which normally prevents immune factors from reaching the peripheral nervous system. Disruption of the blood-nerve barrier is more common, especially in the upper extremities, as shoulder movement is greater[4]. PIN paralysis can result from acute or structural compression of the nerve in Frohse's arch, resulting in poor finger extension. True neurogenic PIN palsy is not usually accompanied by widespread pain or tenderness. Therefore, if the physician is unaware of the clinical feature of NA, which is an episodic pain followed by rapid onset weakness with muscle atrophy, he/she may perform unnecessary testing and misdiagnose the patient with a herniated cervical disc, PIN syndrome, or other musculoskeletal problems.

The four-month interval between brachial plexus palsy and PIN in our case suggests that peripheral nerve involvement may recur in different nerves at NA. Several case series suggest that corticosteroids and intravenous immunoglobulin can be effective in the acute phase of NA, and the earlier treatment initiation, the higher the chance of a favorable response[5]. Up to 30% of NA patients have permanent motor deficits, and nonhealing nerves often have focal hourglass stenosis, which can lead to severe nerve stenosis[6]. If there is no significant clinical improvement, surgical neurolysis is indicated within 6-12 months to restore reinnervation[7].

Recurrent attacks occur in a substantial proportion of patients, with at least 25% of idiopathic cases and 75% of hereditary cases experiencing a second attack within the first few years after the first attack[4]. But if we search the literature, this case is the first report of the SEPT9 negative recurrent NA with brachial plexus involvement on one side and isolated PIN involvement with early conduction block on the other side.

The clinical features and diagnostic studies, in this case, support the contention that NA may present as multiple mononeuropathies associated with inflammation of the peripheral nerves and the plexus.

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