OLGU SUNUMU CASE REPORT

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Postpartum Acute Brachial Plexus Neuritis Mimicking Anterior Interosseous Nerve Palsy

Anterior İnterosseöz Sinir Paralizisini Taklit Eden Postpartum Akut Brakiyal Pleksus Nöriti

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ABSTRACT Acute brachial plexus neuritis also known as neuralgic amyotrophy (NA) is one of the rare neuromuscular diseases characterized by sudden onset of severe pain and sensory deficits in the upper extremities and is followed by muscle weakness and atrophy. The diagnosis is based on the patient's history and physical findings and is corroborated by electromyography. In this case, we present a 35-year-old female who developed idiopathic NA manifesting predominantly with the involvement of the anterior interosseous nerve in the postpartum period following normal pregnancy and delivery with cesarean section under epidural anesthesia.

Keywords: Acute brachial plexus neuritis; neuralgic amyotrophy; postpartum; anterior interosseous nerve

ÖZET Nöraljik amyotrofi olarak da bilinen akut brakiyal pleksus nöriti, üst ekstremitelerde ani başlayan şiddetli ağrı, duyu bozukluğunu kas güçsüzlüğü ve atrofinin takip ettiği nadir görülen nöromüsküler hastalıklardan biridir. Tanı hastanın öyküsüne, fiziksel bulgularına dayanır ve elektromiyografi ile doğrulanır. Bu olguda, normal bir gebeliği takiben epidural anestezi altında sezaryen ile doğum yapan, ağırlıklı olarak anterior interosseöz sinir tutulumu ile seyreden postpartum idiyopatik nöraljik amyotrofi tespit edilen 35 yaşında bir kadın hastayı sunuyoruz.

Anahtar Kelimeler: Akut brakiyal pleksus nöriti; nöraljik amyotrofi; postpartum; anterior interosseöz sinir

Neuralgic amyotrophy (NA) is an acute neuropathy mainly affecting the brachial plexus or its branches and is characterized by non-specific and painful clinical manifestations.[1] The etiology of this disease is still unclear. Some precipitating events (immunization, infection, surgery, pregnancy, trauma, etc.) seem to trigger the syndrome but its pathophysiology remains uncertain.[2] Clinically, symptoms with abrupt onset of shoulder pain, usually unilaterally are followed by progressive neurologic deficits of motor weakness, dysesthesias, and numbness.[3] The diagnosis of NA is typically based on history and physical examination; electrodiagnostic testing is crucial to confirm the diagnosis.[4] Proximal entrapment neuropathies of the upper limb, compressive cervical root disorder, rotator cuff syndrome, meningo-radiculitis, facio-scapulo-humeral myopathy, infiltrative or post-radiation plexopathy, and vasculitis should be considered in differential diagnosis.[1] The following case report focuses on NA which presented as mimicking anterior interosseous nerve (AIN) palsy in an active female in the postpartum period.

CASE REPORT

A 35-year-old woman presented to our outpatient clinic complaining of a weakness to flex the terminal phalanges of her right index finger and thumb which

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started 6 weeks ago. Before her complaints onset, she had delivered a full term baby with a cesarean section under epidural anesthesia. There were no complications. The patient and baby were discharged after one day in hospital. On detailed questioning, she described an acute onset sharp pain in right arm two days after delivery. There was no trauma to her right upper extremity by way of positioning while on the operating table. She told that her arm pain remitted within two weeks. But she noticed a lack of strength in the right hand especially her right thumb and index fingers. There was no sign of systemic disease, vasculitis or malignancy in her past medical history and no family history of neuromuscular disease. There was also no sign of infection. The patient had no fever, headache, muscle soreness, arthralgia, malaise, or rash. She had no history of smoking or alcohol intake. On physical examination, she had full range of motion in the neck and shoulder. Neurological examination showed weakness of the right flexor pollicis longus (FPL) and the flexor digitorum profundus (FDP) to the index finger as Medical Research Council (MRC) grade 2/5. She was unable to form an O with the thumb and index finger (O sign or Spinner's sign), and had abnormal pinch grip strength (Figure 1). No other muscle weakness, sensory loss, or reflex abnormality was detected. Cranial nerve examination was normal. She could ambulate independently. Complete blood tests including autoimmune screen and biochemical parameters were unremarkable. Radiographs of the chest, shoulder and neck were noncontributory. Magnetic resonance imaging (MRI) of the neck and the right shoulder revealed mild degeneration in the intervertebral discs and acromioclavicular joint. Her right arm ultrasonography showed no abnormalities. Nerve conduction study (NCS) and needle electromyography (EMG) were performed 4 weeks after onset the patient's symptoms. Motor NCS showed decreased amplitudes of compound motor action potentials (CMAPs) in both right median and ulnar nerves recorded on the levels of wrist and elbow (Table 1). In the needle EMG, intense abnormal spontaneous activity was detected in the right FPL and FDP-II muscles, while motor unit potential (MUP) was not observed. Mild polyphasic MUPs were observed in the right APB, pronator quadratus (PQ) and ADM muscles with a mild abnormal spontaneous activity. Recruitment was reduced in a neurogenic pattern in right PQ and no motor units were recruited in right FPL, FDP-II (Table 2). Electrodiagnostic studies revealed a pattern of plexopathy not readily localizable to one or more specific trunks, divisions or cords. The patient was diagnosed with right NA predominantly involving the nerve fibers belonging to AIN. Physical therapy including electrical stimulation, joint range of motion and strengthening exercises for the denervated muscles was started. On control visit 3 months after the treatment, she started to use her right thumb and index fingers effectively during daily activities with a motor strength of grade 4/5 (MRC) in interphalangeal joint flexion. Reinnervation MUPs were seen in the muscles innervated by the AIN in the control EMG which was performed 3 months after the treatment (Table 2). Both clinically and electrodiagnostically, recovery was observed with improvement in strength and recruitment of motor units. The patient told that she almost returned to normal activities. A written informed consent was obtained from the patient.

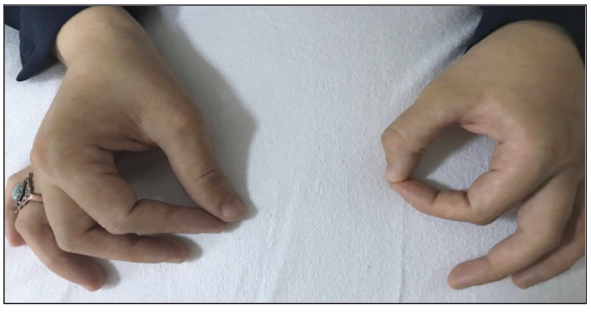

FIGURE 1: Abnormal pinch grip (right hand) due to weakness of the right thumb interfalengeal and index finger distal interfalengeal joint flexion.

DISCUSSION

NA, is a rare disorder that occurs most often in healthy individuals.[5] It was first described by Dreschfeld in 1887. He reported recurrent episodes of the condition in 2 sisters.[6] Several other reports followed, but it was Parsonage and Turner who clearly detailed the clinical features of the condition in a cohort of 136 patients in 1948.[7,8] Hence, this clinical condition has become commonly known as Parsonage-Turner syndrome.[7,9]

TABLE 1: Nerve conduction study.

Nerve stimulation	DML (ms)	AMP (2-4uV)	NCV(m/s)
Sensory			
Right median -2nd finger	2.34	19.5	40.7
Right ulnar-2nd finger	2.29	30.2	43.6
Right radial superficial	3.05	12.5	43.9
Right medial antebrachial cutaneous	2.9	10	41.4
Right lateral antebrachial cutaneous	2.3	14	60.9
Motor			
Right median-APB			
1. Wrist	3.18	**1.9**	
2. Elbow	7.19	**1.1**	62
Right ulnar-ADM			
1. Wrist	1.98	**3.7**	
2. Below elbow	5.57	**3.3**	64
3. Above elbow	7.14	**3.1**	58
3rd month			
Right median-APB			
1.Wrist	3.33	4.6	
2.Elbow	7.50	4.0	60
Right unlar-ADM			
1.Wrist	2.19	7.2	
2.Below elbow	5.94	6.6	64
3.Above elbow	7.71	6.4	62

DML: Distal motor latency; **AMP:** Amplitude; **NCV:** Nerve conduction study; **ADM:** Adductor digiti minimi; **APB:** Abductor pollicis brevis.

TABLE 2: Needle EMG findings.

	Spontaneous					MUAP			Recruitment
Muscles (Right side)	**IA**	**Fib**	**PSW**	**Fasc**	**H.F**	**Amp.**	**Dur.**	**PPP**	**Pattern**
C5 paraspinal	N	None	None						
C6 paraspinal	N	None	None						
C7 paraspinal	N	None	None						
C8 paraspinal	N	None	None						
Deltoid	N	None	None	None	None	N	N	N	N
Triceps brachii	N	None	None	None	None	N	N	N	N
Biceps brachii	N	None	None	None	None	N	N	N	N
Pronator teres	N	None	None	None	None	N	N	N	N
FCU	N	None	None	None	None	N	N	N	N
FDP-digit II	I	4+	4+	None	None	N	N	N	No Activity
FPL	I	4+	4+	None	None	N	N	N	No Activity
ADM	I	1+	2+	None	None	N	N	N	Reduced
APB	I	2+	2+	None	None	N	N	N	Reduced
PQ	I	2+	2+	None	None	N	N	N	Discrete
EİP	N	None	None	None	None	N	N	N	N
3rd month									
FDP-digit II	I	2+	2+	None	None	1-	1+	1+	Reduced
FPL	I	2+	2+	None	None	1-	1+	2+	Discrete
APB	N	1+	1+	None	None	1+	1+	1+	Reduced
PQ	N	1+	1+	None	None	1+	1+	1+	Reduced
ADM	N	1+	1+	None	None	1+	1+	1+	Reduced

MUAP: Motor Unit Action Potential; **IA:** İnitial Activity; **Fib:** Fibrillation; **PSW:** Positive Sharp Wave; **Fasc:** Fasciculation; **H.F:** High Frequency; **Amp:** Amplitude; Dur: Duration; **PPP:** Polyphasic Potentials; **N:** Normal; I: Increased; **FCU:** Flexor carpi ulnaris; **FDP:** Flexor digitorum pollicis; **FPL:** Flexor pollicis longus; **ADM:** Adductor digiti minimi; **APB:** Abductor pollicis brevis; **PQ:** Pronator quadratus; **EİP:** Extansor indicis proprius.

The incidence of NA is approximately 2 to 3 per 100.000 person years.[10] The classical clinical pattern of NA includes 3 consecutive phases: painful phase, then weakness, amyotrophy and sensory complaints, then recovery. Pain is the first symptom in 90% of cases, it has an acute onset.[1] It is typically severe, neuropathic and unrelenting, often waking patients from sleep. Its duration varies from 1 day to 2 months.[7] Weakness may precede the pain in 5% of cases but occurs within 24 hours in 34% of cases, after 1-7 days in 39% and after 1-4 weeks in 27%. The amyotrophy appears generally between 2 and 6 weeks and indicates the significance of the axonal loss.[1,10] In this case, the patient's pain terminated in 2 weeks, and then weakness appeared.

The reason of this syndrome is unclear although it often suggests an autoimmune origin such as in Guillain-Barre syndrome.[1,5] The current evidence suggests that NA has a complex pathophysiology that includes an underlying predisposition, susceptibility to dysfunction of some peripheral nervous system structures, and an autoimmune trigger.[11] Precipitating conditions are common benign trauma, simple or strenuous exercise, pregnancy, childbirth, any kind of surgery, several bacterial or viral infections, and also various vaccinations.[1] In this case, the patient's inciting event appeared to be a delivery with cesarean section and childbirth. The patient was not vaccinated recently before the delivery. There was also no sign of infection and malignancy. The patient had no fever, headache, muscle pain, fatigue, or rash. Concerning postsurgical origins, traction injury due to intraoperative positioning and an immune-mediated inflammation of the brachial plexus are main factors. In this case the patient's arms were resting. Traction was not applied, and there was no positioning of the arms for a long period of time. In a study conducted by Lederman and Wilbourn, it is supported that neither the route of delivery nor the type of anesthesia is important in causing or triggering an attack of NA. Both vaginal deliveries and cesarean sections have been related with the condition.[11] The current hypothesis in this case is that NA may have been caused by an underlying susceptibility to mechanical injury of the brachial plexus, and caused by an immune-mediated response due to pregnancy. Pregnancy also increases the risk of compression, stretch, or entrapment of various nerves. Delivery and labor and the postpartum period create additional conditional risk.

NA has both an idiopathic and hereditary form.[7] Hereditary neuralgic amyotrophy (HNA), is an autosomal dominant disorder, which predisposes to recurrent attacks of peripheral nerve damage. HNA is mainly linked to a mutation in the gene of the septin-9 protein (SEPT9) on chromosome 17q25.[10] Surgery and childbirth are known triggers for HNA attacks. Some prior reports have suggested the hereditary forms of NA as being especially likely to predispose to postpartum events.[11] But this form is differentiated from idiopathic form with familial recurrence, earlier age of onset.[12] In this case, the patient had no family history. Given the lack of family history, our suspicion of HNA was low, though we cannot rule it out as a contributing factor as no genetic testing was done.

In the differential diagnosis, cervical radiculopathy, complex regional pain syndrome involving the shoulder or arm and mononeuritis complex (peripheral nervous system vasculitis) should be considered. According to the clinical and additional investigations, meningo-radiculitis, neoplastic plexopathy, or vasculitis, for example, may be considered. With painless "NA", chronic idiopathic demyelinating polyneuropathy, multifocal motor neuropathy, Lewis Sumner syndrome, and hereditary neuropathy with liability to pressure palsy or a facio-scapulohumeral myopathy may be considered.[1] In some cases, NA may involve nerves not in the brachial plexus. Extra-brachial forms are particularly those affecting the lumbosacral plexus, and the phrenic nerve. In these cases, diabetic amyotrophy may be considered which is characterized by acute onset of proximal, unilateral pain of the hip, buttocks, or thigh, progressing to sensory disturbance of the affected limb and followed by weakness and atrophy of the proximal lower extremity muscles.[1,13] In this case, the patient's deep tendon reflexes and sensory examination were normal and the patient had no weakness in the lower extremities. Therefore, diabetic amyotrophy was ruled out.

The diagnosis of NA is essentially based on history and physical examination; electrodiagnostic testing is crucial to confirm the diagnosis and monitor recovery.[3,4] The distribution of abnormalities in NA can vary from an isolated nerve to the widespread involvement of the brachial plexus.[14] It has been reported that paresis of the upper part of the brachial plexus, affecting the shoulder girdle muscles, is the most common (71.1%) form of it and involvement of the lower part of the plexus is less likely. A few cases of NA presenting as AIN compression have been reported.[10,15] In this patient, EMG indicated that the median and ulnar nerves were also involved, but the AIN-innervated muscles were severely damaged. Sometimes, MRI of the cervical spine and brachial plexus is helpful to exclude alternate diagnoses, such as a compressive lesion.[3,9] There are no specific laboratory tests to guide diagnosis. Blood investigations may be helpful to define the recent infection (IgM bodies). Searching for diabetes mellitus and antibodies such as ANCAs, is useful to rule out some autoimmune disorders. In this case, the patient's complete blood tests, biochemical parameters including fasting blood sugar, glycated hemoglobin A1c, autoimmune screen were normal. Lumbar puncture findings are usually normal in NA and the procedure is rarely performed but is necessary with extensive NA, with involvement of the lumbosacral plexus or the cranial nerves, or with chronic erythema migrans to search for Lyme disease and other causes of meningo-radiculitis. Genetic investigations of the SEPT9 protein will be performed when HNA is suspected.[1,10]

There is no specific treatment protocol established for NA. First-line treatment is based on analgesia together with anti-inflammatory drugs and other co-analgesics. Some articles have reported favorable outcomes of oral or intravenous steroid treatment for NA. Considering the rarity of the disorder, there is poor literature evidence to support its efficacy.[16,17] Physical therapy is also an important adjunct in the treatment of NA to enhance range of motion and strength of the shoulder girdle, scapular stabilizers, and rotator cuff.[5] Passive joint range of motion exercises for the shoulder and elbow can be added with the reduction of pain. Considering the recovery status of the affected muscles, active-assisted and active range of motion exercises should be initiated. Other modalities such as massage, ultrasound, and electrical stimulation may be helpful, but there are insufficient studies in the literature on their use.[18,19] The clinical course of this condition varies from patient to patient. When the nerve lesion is partial, the recovery is provided by collateral reinnervation and may be completed in 6 to 12 months. In contrast, when the lesion is severe, recovery is based on nerve growth and direct reinnervation, requires 1 to 3 years, and is not always as good.[1,7] If there is no evidence of regeneration with EMG after 6-9 months, nerve transfers or decompression surgery can be considered.[20] Drug treatment could not be given in this case because the patient was breastfeeding her baby. So, the main treatment of the patient was given by physical therapy. A significant recovery was observed in the patient 3 months after the treatment.

CONCLUSION

In the present report, we discussed the case of a woman diagnosed with NA in the postpartum period. Lack of knowledge of this condition among physicians and clinical variability often result in the lack of recognition of this syndrome. In a patient presenting with shoulder pain followed by weakness, NA should always be included in the differential diagnosis. This case report highlights the importance of considering the association of NA with postpartum events and may presenting as mimicking AIN palsy.

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