

# Neuralgic Amyotrophy (Parsonage-Turner Syndrome): An Often Misdiagnosed Diagnosis

Pages with reference to book, From 101 To 103

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Neuralgic amyotrophy (Parsonage-Turner syndrome) is an idiopathic brachial plexus neuritis which commonly affects the upper trunks<sup>1</sup>. Since the first description during World War II<sup>2,3</sup> there have been multiple reports highlighting the manifestations of this syndrome<sup>4-7</sup>. The clinical presentation may be individual mononeuropathy, multiple mononeuropathies or involvement of a single trunk, most commonly the upper trunk<sup>8</sup> have all been reported as presentations or neuralgic amyotrophy. We report 4 cases of neuralgic amyotrophy presenting to the electrodiagnostic laboratory at the Aga Khan University within the past 2 months. We re-emphasize the varied presentation, including diaphragmatic paresis in one patient. We also question the pathologic localization of this entity, as our experience does not support a brachial plexus origin.

## Case Report

**Patient 1:** A 40-year-old male presented for evaluation of inability to move right upper arm for the past month. He denied any history of fever or trauma preceding this weakness. Examination revealed impaired abduction at the shoulder joint and impaired external rotation of the humerus. Atrophy of the right infraspinatus, supraspinatus and deltoid was seen. Sensory loss extended over the right forearm laterally. Electrodiagnostic study revealed normal right median, ulnar motor and median, ulnar sensory nerve conductions. Right axillary and musculocutaneous motor nerve conductions revealed decreased compound motor action potential amplitudes. EMG revealed severe denervation in the right deltoid and infraspinatus and mild denervation in the right biceps and supraspinatus.

**Patient 2:** A 51-year-old male underwent splenectomy for splenic vein thrombosis and bleeding gastric varices. Seven days post operatively he developed right shoulder pain. The pain was so severe that he refused to move his right arm. He denied a history of traction, neck injury or trauma. Examination revealed limited abduction at the shoulder joint. External rotation of the right humerus was weak. No atrophy was seen. Right biceps reflex was diminished. A patchy sensory loss was noticed over the deltoid muscle. Electrodiagnostic studies revealed normal median and ulnar motor and sensory nerve conductions. EMG revealed active denervation in the infra and supraspinatus on the affected side.

**Patient 3:** A 70-year-old male presented with inability to move his right arm for the past one month. He had a febrile illness with productive cough 6 weeks prior to this presentation. Examination revealed inability to abduct or externally rotate right arm. There was wasting of the right supra and infraspinatus muscles. No sensory loss was detected and reflexes were normal. Motor and sensory nerve conductions of the right median, radial and ulnar nerves did not reveal any abnormalities. EMG revealed frank denervation in the right infraspinatus only. Mild changes were seen in the right deltoid.

**Patient 4:** A 35-year-old male developed sudden onset breathlessness while jogging. This was preceded by mild neck ache for about a week. He denied any history of major trauma or febrile illness. He felt more comfortable in erect posture. He complained of dyspepsia. Examination revealed no restriction of movements at the shoulder joint. Muscle strength was normal. Investigations revealed a right hemidiaphragmatic paralysis. He was managed conservatively. A repeat evaluation at six months revealed improved movements of the right hemidiaphragm. Bilateral phrenic nerves studies revealed

normal onset latencies and compound motor action potential amplitudes. EMG did not reveal any active denervation, but recruitment frequency was decreased.

## Discussion

Major Spillane in his classic report<sup>3</sup> described 46 cases from the Middle East Expeditionary Force with “unusual neuritic features”. In some the symptoms started while the soldiers were recovering from some other malady in the field hospital. In others, the symptoms started in the field and they were not all admitted for these complaints. Common theme in all these patients was a sharp pain around the affected shoulder. Pain was followed by neuritic weakness of the shoulder girdle muscles in about 4 to 5 days. There was a high incidence of serratus anterior palsy followed closely in numbers by weakness of the spinati. Spillane<sup>3</sup> also commented on isolated nerve palsies and the relative sparing of the pectoralis, latissimus dorsi, triceps, brachialis and brachioradialis.

Five years later, Parsonage and Turner, reported their experience with 136 cases, from the British Army in the United Kingdom and in India Command<sup>2</sup>. The clinical phenotype was remarkably similar to Spillane’s. They commented that “without any constitutional disturbance pain starts suddenly across the top of the shoulder blade. This pain lasts from a few hours to a fortnight or more and then a flaccid paralysis of some of the muscles of the shoulder girdle and often of the arm develops”. They labeled the syndrome as “Neuralgic Amyotrophy”. The disease was found in all ethnic populations and 66 out of 136 patients were in the hospital at the time of onset of symptoms for minor surgical procedures or unrelated medical illnesses. They commented on the differing patterns of involvement. In some patients, a single nerve was affected whereas in others multiple nerves or nerve-root involvement were seen. They also highlighted the differential muscle involvement within a single nerve distribution. Prognosis was good and most of their patients, recovered to a considerable degree within 24 months. Many reports have since confirmed the existence and importance of this form of neuropathy in the post World War era<sup>1,4,7,9-12</sup>. Scattered reports have also highlighted phrenic nerve involvement, in isolation or in combination with other nerves<sup>13</sup>. Cases of neuralgic amyotrophy have been misdiagnosed as diaphragmatic ruptures<sup>14</sup> and some of these patients were subjected to extensive investigations to elucidate the cause for the diaphragmatic palsy.

Our cases followed the same pattern. In all patients, the symptoms occurred on the right side starting with shoulder pain or neck pain. The weakness crept on insidiously. The disability, as in Spillane and Parsonage-Turner’s experience, was considerable. The syndrome followed surgical procedure in Case 2 and febrile illness in Case 3. In the other two patients no immediate cause could be determined. The illness was monophasic in all cases. All patients are pain free now but have variable degree of weakness and disability on follow up examinations. Conventional motor and sensory nerve conductions were not helpful. Even recording sensory antidromic potentials from digit 1 (upper trunk), 3 (middle trunk) and 5 (lower trunk) did not yield any significant side to side difference. Our experience with the antebrachial nerves<sup>15-17</sup> (data not shown) has not been positive, but we are looking at it prospectively. Use of appropriate proximal motor nerve conductions and extensive needle EMG study, including sampling of the unaffected side, was the most useful electrodiagnostic evaluation. EMG in all patients confirmed the neuropathy with either a mononeuropathy or a multiple mononeuropathy pattern with variable muscle involvement. In case 3, infraspinatus was more involved than the supraspinatus, in keeping with the experience of Parsonage and Turner and many others”<sup>18</sup>.

The etiology of this syndrome is not clear but may be related to post-inflammatory immune mono or multiple mononeuropathies<sup>8</sup>. A single case report showed pathological changes in the brachial plexus<sup>18</sup>. In general, mononeuropathy or selective involvement of muscles within a nerve distribution is more common. Our cases also failed to show the sensory nerve conduction abnormalities that would

be expected if this was truly a brachial plexus, especially upper trunk lesion. The support for a plexus origin for this form of neuropathy in the electrodiagnostic literature is fast dwindling<sup>8,15</sup>. We support the notion that a proximal lesion of individual or multiple nerves causes this neuropathy and sometimes selective involvement may be seen within a nerve, as in case 3. As the lesions in this entity sometimes cannot be localized to the trunks and cords of the brachial plexus, the term “neuralgic amyotrophy” may be preferable to the term “brachial plexus neuropathy<sup>8</sup>”. Some of these cases may be misdiagnosed as entrapment neuropathies whereas in others, iatrogenic factors (such as malpositioning during general anesthesia or improper intramuscular injection or vaccination) are blamed for the presentation. It is not unusual for some of these cases to be under orthopedic or physical therapy care for “frozen shoulders”. Knowledge of this entity will help prevent over investigation and improper referrals. Reassuring the patients regarding the good prognosis is important. Simple use of analgesic agents and physical therapy is all that is needed to help patient recover.

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