IDIOPATHIC BRACHIAL NEURITIS

Austin J. Sumner, M.D.

Department of Neurology,
Louisiana State University
Health Sciences Center,
New Orleans, Louisiana

Reprint requests:
Austin J. Sumner, M.D.,
Department of Neurology,
Louisiana State University
Health Sciences Center,
2820 Napoleon Avenue,
Suite 400,
New Orleans, LA 70115.
Email: asumner@lsuhsc.edu

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Parsonage-Turner syndrome (PTS) is a rare syndrome of unknown cause, affecting mainly the lower motor neurons of the brachial plexus. The brachial plexus is a group of nerves that conduct signals from the spine to the shoulder, arm, and hand. PTS is usually characterized by the sudden onset of severe 1-sided shoulder pain, followed by paralysis of the shoulder and lack of muscle control in the arm, wrist, or hand several days later. PTS can vary greatly in presentation and nerve involvement. Also known as brachial plexus neuritis or neuralgic amyotrophy, PTS is a common condition characterized by inflammation of a network of nerves that control and supply, or innervate, the muscles of the chest, shoulders, and arms. Individuals with the condition first experience severe pain across the shoulder and upper arm. Within a few hours or days, weakness, wasting (atrophy), and paralysis may affect the muscles of the shoulder. Although individuals with the condition may experience paralysis of the affected areas for months or, in some cases, years, recovery is usually eventually complete.

KEY WORDS: Brachial, Idiopathic, Neuritis, Parsonage-Turner syndrome

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In 1948, a classic paper appeared in *The Lancet* titled "Neuralgic Amyotrophy. The Shoulder-girdle Syndrome," written by M.J. Parsonage (5), a registrar at Guy's Hospital and John W. Aldren Turner, consultant neurologist at St. Bartholmew's Hospital. Their syndrome comprised pain and flaccid paralysis of the muscles around the shoulder girdle, which occurred fairly often during the years of World War II (1941–1945), but had previously been rare. They observed 136 cases during neurological work in the army in the United Kingdom and India. In addition to muscle paralysis around the shoulder girdle, they noted involvement of the thumb and index finger in 5 cases. In 1 of these cases, the paralysis of the thumb and index finger was an isolated finding. They stated, "This localized paralysis cannot anatomically be of peripheral nerve or nerve root distribution and is only explicable by an anterior horn lesion" (5). Of course, it was a short time later that Kiloh and Nevin (4) described this pattern of paralysis as consistent with involvement of the anterior interosseous nerve. Thus, their name for the condition, *neuralgic amyotrophy*, was based on the concept that the condition did not arise from involvement of discrete individual nerves. Perhaps the most influential study on this condition published in the United States was that of Tsairis et al. (8) when they reported 99 patients in a publication titled "Natural History of Brachial Plexus Neuropathy" in 1972. In their excellent clinical description of patients, they noted that the disease may involve the upper, lower, or entire brachial plexus, sometimes bilateral, and showed a male predominance (70 males versus 29 females). They made an important observation that in 8 of their patients, strenuous exercise preceded the onset of pain and weakness by 1 to 7 days. They noted that sensory involvement, although not prominent, often involved the lateral upper arm and the radial lower arm when present. They noted that, although they determined clinically that the lesions lay within the brachial plexus, they were diffuse and incomplete with sparing of some muscles of the same root distribution. In fact, they noted paralysis localized to a single nerve in 10 patients: the radial nerve in 5 cases, the long thoracic nerve in 3 cases, and the suprascapular nerve in 2 cases. Also, 6 patients exhibited pain that was most severe not at the shoulder, but rather at the elbow. However, they did not correlate this with the presence of an anterior interosseous neuropathy.

On January 5, 1940, the first echelon of the 2nd New Zealand Expeditionary Force embarked to Maadi Camp near Cairo. These forces were accompanied by 2 young physicians from New Zealand, Captains E.D. Burnard and T.G. Fox. While these New Zealand forces were in training in the Middle East, Burnard and Fox published their observations in the *New Zealand Medical Journal Vol. XLI, 1942*, in which they described a condition called "multiple neuritis of the shoulder girdle."(1) It seems, however, that their detailed description has not been widely acknowledged as a classic initial description of the condition, now generally attributed to Parsonage and Turner. They also reached the important conclusion that the shoulder girdle paralysis typically involved the distribution of individual nerves arising from the brachial plexus or close to it and not from a localization within the brachial plexus. They found that

ABBREVIATIONS: PTS, Parsonage-Turner syndrome

trapezius paralysis was the most common single nerve secondary to involvement of the accessory nerve: the suprascapular nerve in 8 cases, the long thoracic nerve to serratus anterior nerve in 5 cases, the axillary nerve in 4 cases, and the musculocutaneous nerve in 4 cases. They never encountered involvement of the dorsal scapular nerve to the rhomboids or the medial or lateral pectoral nerves as exceptions. Soon after, the condition became widely recognized in soldiers in the Middle East forces, and Major J.D. Spillane (6) described a further 46 cases of late localized neuritis of the shoulder girdle in *The Lancet*. The prevailing theory was that these cases represented an infectious etiology, and the term *infective neuritis* appeared in "Notes on Nervous Diseases and Head Injuries in the Middle East" as an official general headquarters publication of the *Middle Eastern Forces Pamphlet, 1943*. (3)

Dr. John D. England and I attempted to expand the concept of Parsonage-Turner Syndrome (PTS) beyond that prevalent at the time in a 1987 publication (7), titled "Neuralgic Amyotrophy: An increasingly Diverse Entity." We described more complex patterns of nerve involvement than had been described previously. For example, we found cases presenting with isolated paralysis of the pronator teres muscle, the muscle innervated by 3 distinct nerve branches of the median nerve. We noted that the lateral brachial cutaneous nerve was involved in several instances in the absence of involvement of the whole musculocutaneous nerve and that in 1 case, there was a small cutaneous branch lesion of the recurrent palmar branch of the median nerve in the palm accompanying otherwise typical features of PTS. These observations led to the conclusion that the distribution and location of individual nerves or branch lesions in the upper extremity could be quite complex and were not restricted to the brachial plexus. These themes were expanded by Cruz-Martinez et al. (2) in their article titled "Neuralgic Amyotrophy: Variable Expression in 40 Patients." They also documented that the clinical entity seemed to consist of a mononeuritis multiplex affecting 81 nerves in 40 patients, in whom 33 were unilateral and 7 were bilateral. The most common individual nerves in rank order were the suprascapular, axillary, musculocutaneous, long thoracic, and radial nerves. Small numbers of cases of the anterior interosseous and dorsal interosseous, lateral antebrachial cutaneous, phrenic and axillary nerves were also present in their cases.

I was struck by a single case report of a 35-year old man who developed sudden onset severe pain in both shoulders without obvious cause published by Yamamoto et al. (9). The next day, the patient's pain appeared at both elbows and was associated with an inability to extend the left elbow due to pain. Two days later, he was unable to flex the left thumb and index finger. The pain diminished over the next 2 weeks, but the weakness did not recover. Neurological examination showed focal tenderness over the median nerve in the antecubital fossa. His examination of the left arm showed FPL measuring 0/5, FDP measuring 0/5, pronator teres measuring 1/5, and flexor carpi radialis measuring 1/5, but all other median nerve muscles were 5/5. In the contralateral right arm, there was isolated complete paralysis of flexor carpi radialis with all other muscle groups exhibiting 5/5 strength and normal sensation throughout both extremities. Electromyography confirmed complete acute denervation in the paralyzed muscles. What was unusual about this case is that the patient underwent surgical exploration 14 weeks after the onset which showed, as expected, no extrinsic nerve compression. However, interfascicular neurolysis was undertaken, which revealed 2 hour glass-like fascicular constrictions in the anterior interosseous nerve at 2 levels: adjacent to the medial epicondyle and 2 cm distal to that point. There were also constrictions of the motor branches, 2 in the pronator teres and the flexor carpi radialis above the medial epicondyle. This was the first pathological evidence indicating multifocal involvement of terminal branch lesions underlying the complex patterns of paralysis often encountered clinically. A perineurial biopsy showed mild perivascular lymphoid cells in the epineurium. More recently, further examples of nontraumatic anterior interosseous palsy have been described with multiple constrictions along the course of the nerve which Ysunaga et al. (10) from Fukuoka, Japan have described as fascicular torsion in the median nerve within the distal one-third of the upper arm. Similar lesions have been described in the posterior interosseous nerve by Yongwei et al. (11) from Beijing, China in a publication titled "Nontraumatic Paralysis of the Radial Nerve with Multiple Constrictions." Again, there was evidence of inflammatory infiltration of affected nerves, but these authors were of the opinion that the constrictions were the result of partial pronation of nerve fascicles with secondary constriction which they modeled by twisting a sausage balloon. This remarkable and unexpected nerve pathology appears to the basis of lesions in PTS, which could be thought of as a mononeuritis multiplex of the upper extremity with multifascicular inflammatory and constrictive lesions.

I would pose the following questions:

1) Are these nerve lesions the results of combined inflammatory and mechanical interactions?
2) What is the role of soft tissue injury (e.g., trauma, surgery, or child birth) as etiological factors in the onset of PTS?
3) The role of extreme muscular exertion or sustained repetitive activity has frequently been noted as an antecedent event in the onset of acute idiopathic brachial neuritis. Why are these nerve lesions usually located close to joints or other mobile locations (e.g., scapula, shoulder joint, or elbow joint)?

I do not have the answers to these questions, but idiopathic brachial neuritis, or PTS, is turning out to be a much more complex syndrome than previously thought. I would simply leave off with the thought that a more complete name of the condition might be multifocal multifascicular inflammatory and constrictive brachial neuritis.

Disclosure

The author has no personal financial or institutional interest in any of the drugs, materials, or devices described in this article.

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