

Parsonage: Turner Syndrome or Amyotrophic Neuralgia

M. Fournier Mehouas*

Department of Physical Medicine and Rehabilitation Service, CHU, Nice, France

Abstract

Parsonage and Turner syndrome or amyotrophic neuralgia manifests itself by the sudden onset of violent pain in the shoulder area, followed by paralysis and amyotrophy of the muscles of the scapular belt, frequently accompanied by sensory disturbances. It is essentially an attack on the brachial plexus, most commonly on the upper trunk, but it can also be located outside the brachial plexus (cranial nerves, phrenic nerve, or even lumbosacral plexus). The diagnosis is based above all on the clinical history and physical examination, confirmed by additional tests: Electromyogram, high-resolution ultrasound, magnetic resonance imaging, which will make it possible to specify the topography of the damage and eliminate differential diagnoses, mainly shoulder tendon damage and cervical radiculopathies. In addition to idiopathic forms, there are more severe and recurrent familial forms with autosomal dominant transmission, often associated with dysmorphia. Their genetic diagnosis is based on the mutation of the Septin Gene (SEPT9). The etiology remains unknown at present, a dysimmunity hypothesis associated with certain favourable factors, essentially infectious and vaccinal, is suspected. The treatment is symptomatic, based in the acute phase on analgesic treatments or even corticotherapy and on re-education adapted according to the phase. The evolution is favourable in the majority of cases, with recovery over a few months and in 70% of cases a total recovery at 3 years. However, motor after-effects and recurrences are possible.

Keywords

Parsonage and turner syndrome; Amyotrophic neuralgia; Brachial plexus; Neuropathy

Introduction

Parsonage and Turner Syndrome (PTS) is a rare disease, characterised by the onset of sudden and extreme pain in the upper limb, followed by progressive deficits including weakness, atrophy and sometimes sensory disturbances. The cause is unknown. The distribution of nerve damage is variable. The most common site is the upper trunk of the Plexus Brachial (PB). Recovery is long and incomplete. A hereditary form of the syndrome also exists but is much less frequent with an identical clinic but in younger subjects with a high incidence of recurrent damage. This clinical picture is commonly called Parsonage and Turner Syndrome (PTS), but also amyotrophic neuralgia, Brachial Plexus neuropathy, acute brachial neuropathy, idiopathic brachial plexopathy, paralyzing brachial neuritis, brachial radiculitis, scapular belt syndrome.

History/epidemiology

One of the first descriptions dates back to 1887, when Dreschfeld [1] described two cases of recurrent non traumatic episodes of brachial plexus paralysis. In 1948 MJ Parsonage and J Turner published an article: "Neuralgic amyotrophy: The shoulder girdle syndrome" which analysed 136 patients with Parsonage and Turner Syndrome [2]. It is from this work that Parsonage and Turner Syndrome was described in details.

A rare disease with an annual incidence of 2/100 000 but more recently an incidence of 1/1000 has been reported and is considered to be underestimated due to the unknown diagnosis. More common in males, dominant (58%) and unilateral (97%) [3].

Clinical Diagnosis

The classic form requires the indispensable condition of the 3 successive phases: Painful, then deficit and finally recovery.

Painful phase

The pain, the first symptom in 90 to 95% of cases, is brutal, of the scapular belt, permanent, with nocturnal recrudescence and which can even wake the patient up, violent (EVA7/10), without general signs, of a neuropathic type lasting from a few hours to several months. 30 to 70% of patients present with persistent pain (neuropathic or compensatory muscle pain or even chronic pain that is not systematised). It can radiate into the neck or upper limb homolateral to the deficit. Some have bilateral pain with unilateral motor impairment.

Deficiency phase

Weakness, amyotrophy and sensory disorders after the onset of pain which increases or persists the location of the initial pain does not necessarily correspond to the muscular damage. The amyotrophy appears rapidly. Most often, it is a multifocal attack on the predominant upper BP (long thoracic nerve). The sensory damage is variable (hypoesthesia and unevenly distributed paresthesia).

Recovery phase

Complete between 6 months and 3 years (75%). Recidivism increases the risk of after-effects.

Clinical Forms

Painless, pure sensitive, topographical (bilateral with damage to the anterior or posterior interosseous nerve damage outside the Brachial Plexus) familial with mutation of the septine-9 protein on chromosome

***Corresponding author:** M. Fournier Mehouas, Department of Physical Medicine and Rehabilitation Service, CHU, Nice, France, E-mail: fournier-mehouas.m@chu-nice.fr

Received January 11, 2021; **Accepted** January 25, 2021; **Published** February 02, 2021

Citation: Mehous MF (2021) Parsonage: Turner Syndrome or Amyotrophic Neuralgia. J Clin Exp Neuroimmunol. 6: 124.

Copyright: © 2021 Mehous MF. This is an open-access article distributed under the terms of the creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.

17q25 with repetitions which alter the micro-tubules. These patients may present a dysmorphic syndrome: Epicanthus, hypotelorism, cleft palate, partial syndactyly, facial asymmetry, bifid ovary. (comparison with Modigliani's portraits) dysautonomic [4-7].

Paraclinical Diagnosis

Investigations are useful when faced with an atypical symptomatology.

Electrophysiology proves the peripheral origin of the disease. Signs at the level of clinically affected muscles but sometimes at a distance or on the contralateral side proof of infraclinical damage.

EMG stages

0: Complete denervation,

1: Beginning of reinnervation with the presence of nascent and polyphasic potentials,

2: Almost complete reinnervation defined by decreased recruitment and motor unit potentials of large amplitude or duration of normal or elongated motor unit potentials,

3: Complete reinnervation defined by absence of abnormal spontaneous activity and complete recruitment with normalised appearance of motor unit potentials [8].

Biological balance sheet

- Blood tests are not specific.
- There is no immune bioassay to look for specific antibodies.
- But antibody testing can be useful in defining a recent viral infection that triggers PTSD. Hepatitis E infection should be investigated if bilaterally affected with elevated transaminases. If diaphragmatic paralysis or extended form, type 1 diabetes or autoimmune disease should be investigated.
- Lumbar puncture is not indicated unless there is extensive damage to the lumbosacral plexus or cranial pairs.

Histological study

The nerve biopsy has no indication. The histological alterations are those of a marked axonal degeneration with loss of myelinated fibres in certain bundles.

Genetic study

The study of SEPT9 protein, if suspicion of a familial PTSD is useful and it may reveal a deletion of the PMP22 gene.

The images

• X-rays (shoulder, cervical spine) are normal. Chest X-ray may show the ascension of a diaphragmatic dome, indicating phrenic involvement.

Magnetic Resonance Imaging (MRI) is an aid to early diagnosis and differential diagnosis.

MRI of the cervical spinal cord may show a disc pathology or compression of a nerve root.

MRI of the shoulder can identify other causes of shoulder pain.

• MRI of the Brachial Plexus can show a T2 hypersignal of certain trunks in 6% of cases, with no specificity demonstrated. But [9], it can identify a little-known sign, the hourglass deformity of the nerve, which suggests that MRI can contribute to diagnosis, prognosis and therapeutic decisions.

• Muscle MRI can help in diagnosis and follow-up because of the anomalies related to denervation [10] which appear chronologically: early muscle oedema, then amyotrophy and then fat degeneration.

• Muscular oedema: From the 15th day [11], with a diffuse hypersignal in T2 in the muscular territory innervated by the affected nerves, making it possible to determine the topography of the nerve damage. Non-specific, it is the clinical context and the topography which will guide the diagnosis.

• Muscle atrophy: Sometimes associated with signs of fat involution, it is fairly rapid and is best assessed on T1-weighted sequences without fat saturation [12].

• Fatty degeneration results in linear hypersignal ranges in T1, T2 and proton density which can appear as early as 3 weeks after denervation. Amyotrophy does not necessarily lead to fat degeneration, which is irreversible [4].

• These signs can lead to the diagnosis of PTSD when it is not suspected, whereas MRI is performed for another reason.

Ultrasonounds

The study of the nerve by high-Resolution Ultrasound (HRUS) is potentially useful in the diagnosis of SPT but operator dependent.

There was a significant difference ($p < 0.0001$) [13] in nerve with compared to control subjects or the contralateral limb of the affected limb. Sensitivity is 85.7% and specificity 96.7%.

Differential Diagnosis

It occurs in atypical forms with mainly other neurological disorders of the scapular belt and other peripheral neuropathies [4].

It will be necessary to eliminate:

- Pathologies of the shoulder Pathology of the rotator cuff Complex regional pain syndrome.
- Post-operative damage to the upper limb (excessive traction or prolonged nerve compression due to poor posture).
- In the case of other BP disorders in front of lower BP disorders, Pancoast Tobias syndrome will have to be ruled out and a chest tumour will have to be looked for.

Family forms

- Neuropathies of the scapular belt
- Compression of nerve trunks from PB

Distal forms

- Anterior or posterior interosseous nerve compression
- Damage to the musculocutaneous nerve

Cervical radiculopathy

Cervicobrachial neuralgia deficiency (especially C6-C7)

Finally, we must not forget a possible meningo-radiculitis, vasculitis or facio scapulo humeral dystrophy.

The occurrence of a PTSD has been observed in the context of a systemic disease, so the symptomatology will not be considered as a PTSD but as part of the known systemic disease.

Prognosis

Symptoms may persist for more than a year [14]. Complete recovery is observed in more than 80% of cases at 3 years with a more severe evolution in hereditary forms. The muscle least likely to recover is the anterior serratus (large serratus). The after-effects depend on the extent of the initial damage. Recent studies report a poorer recovery compared to older studies.

Treatment

The processing is currently not coded [15]. It is purely symptomatic and aims to reduce pain, the consequences of neurological deficits and to improve the prognosis of PTSD.

Pharmacological treatments

In the painful phase and if the patient is seen within the first month, corticotherapy (prednisolone per os 60 mg/day 8 days, then 10 mg/day 8 days) [15] is the treatment of choice; it acts on the pain and allows a better recovery after one year.

The following treatments have been proposed:

Intravenous bolus of corticoids (1 g/day 3 days) intravenous immunoglobulins, its a combination of non-steroidal anti-inflammatory drugs and opioids.

Anti-epileptics (gabapentin, carbamazepine) as well as tricyclic antidepressants are often used in neuropathic pain.

Immunosuppressants or plasma exchanges have been tried in a few isolated cases [4] but seem to represent a disproportionate risk compared to the most often favourable evolution of the disease.

The main limitation of these treatments is linked to the fact that these patients are rarely seen at an early stage.

Non-pharmacological treatments

Based on small, non-randomised, double-blind studies [16].

Active and passive functional re-education of the shoulder muscles to combat deficiency and amyotrophy in a prolonged manner, as well as passive joint maintenance of the glenohumeral and stretching to prevent capsular retraction. It is important to prevent secondary pathology of the rotator cuff. Rehabilitation can play a role in pain control. Desensitizing exercises can reduce allodynia.

It would appear that physical therapy does not accelerate recovery, while electro-stimulation for muscle strengthening is controversial.

Surgery (neurolysis, grafting or nerve transfer) is considered to be of uncertain value in exceptional cases (lack of recovery or secondary complications) [17].

Chronic electrostimulation using electrodes implanted at the roots of the brachial plexus has been proposed in a hyperalgesic form with refractory chronic neuropathic pain [18].

Genetic counselling is offered in family forms with possible antenatal diagnosis by amniocentesis.

In total, the most appropriate curative treatment is not known, and the treatment consists of a multidisciplinary symptomatic approach centred on the patient, combining pharmacological treatment, physical therapy and personalised psychological care [19].

Conclusion

The SPT is based on a clinical diagnosis with acute and severe pain in the scapular belt, followed by amyotrophy and paresis of uneven distribution in the territory of the BP. Confirmation is provided by EMG, high-resolution ultrasound and MRI, which in atypical forms allow a differential diagnosis to be made with pathologies of the shoulder, cervical radiculopathies and other neurological compressions of the scapular belt. The evolution is most often favourable with a complete recovery in 75% of cases at 3 years. There are also other atypical clinical forms, autosomal dominant familial forms by mutation of the septine gene. Its etiology remains unknown to this day, even if favourable factors have been identified. The treatment is symptomatic and not codified, as there is no therapy to shorten its evolution.

References

- Rubin DI (2001) Neuralgic amyotrophy : Clinical features and diagnostic evaluation. *Neurologist* 7: 350-356.
- Parsonage MJ, Turner JW (1948) Neuralgic amyotrophy : Shoulder girdle syndrome. *Lancet* 1: 973- 978.
- Seror P (2016) Neuralgic amyotrophy. An upodate. *Joint Bone Spine* 84: 153-158.
- Legré V, Azulay JP, Serratrice J (2009) Syndrome de parsonage et turner (névralgie amyotrophique). *Appareil Locomoteur*, 14-347-A-10.
- Dunn HC, Daube JR, Gomez MR (1978) Heredo familial brachial plexus neuropathy (hereditary neuralgic amyotrophy with brachial predilection) in childhood. *Dev Med Child Neurol* 20: 28-46.
- Serratrice G, Baudouin D, Pouget J, Blin O, Guieu R (1992) Typical and atypical forms of neuralgic amyotrophy of the shoulder : 86 cases. *Rev Neurol* 148: 47-50.
- Laccone F, Hannibal M, Neesen J, Grisold W, Chance P F, et al. (2008) Dysmorphic syndrome of hereditary neuralgic amyotrophy associated with a SEPT9 gene mutation – a family study. *Clin Genet* 74: 279-283.
- Feinberg JH, Nguyen ET, Boachie-Adjei K, Gribbin C, Lee SK, et al. (2017) The electrodiagnostic natural history of parsonage - turner syndrome. *Muscle Nerve* 56 : 737-743.
- Sneag CB, Saltzman EB, Meister DW, Feinberg JH, Lee Sk, et al. (2017) MRI bullseye sign : An indicator of peripheral nerve constriction in parsonage-turner syndrome. *Muscle Nerve* 56: 99-106.
- Cruz-Martinez A, Barrio M, Arpa J (2002) Neuralgic amyotrophy variable expression in 40 patients. *J Peripher Nerv Syst* 7 : 198-204.
- ArAnyi Z, Csillik A, DeVay K, Rosero M, Barsi P, et al. (2017) Ultrasonography in neuralgic amyotrophy : sensitivity, spectrum of findings and clinical correlations. *Muscle Nerve* 56: 1054-1065.
- Lieba-Samal D, Jengojan S, Kasprian G, Wober C, Bodner G (2016) Neuroimaging of classic neuralgic amyotrophy. *Muscle Nerve* 54: 1079-1085.
- Gruber L, Loizides A, Loscher W, Glodny B, Gruber H (2017) Focused high-resolution sonography of the suprascapular nerve : A simple surrogate marker for neuralgic amyotrophy ? *Clin Neurophysiol* 128: 1438-1444.
- Tjoumakaris FP, Anakwenze OA, Kancherla V, Pulos N (2012) Neuralgic amyotrophy (Parsonage-Turner syndrome). *J Am Acad Orthop Surg* 20: 443-449.
- Van Alfen N, Van Engelen BG (2006) The clinical spectrum of neuralgic amyotrophy in 246 cases. *Brain* 129: 438-450.
- Smith CC, Bevelacqua AC (2014) Challenging pain syndromes, Parsonage-Turner Syndrome. *Phys Med Rehabil Clin N Am* 25: 265-277.
- Pan YW, Wang S, Tian G, Li C, Tian W, et al. (2011) Typical brachial neuritis (Parsonage-Turner syndrome) with hourglass-like constrictions in the affected nerves. *J Hand Surg Am* 36: 1197- 1203.
- Bouche B, Manfrotto M, Rigoard P, Lemarie J, Dix-Neuf V et al. (2017) Peripheral nerve stimulation of brachial plexus nerve roots and supra-scapular nerve for chronic refractory neuropathic pain of the upper limb. *Neuro Modulation* 20: 684-689.
- Janssen RMJ, Satink T, Ijspeert J, Van Alfen N, Groothuis JT, et al. (2018) Reflections of patients and therapists on a multidisciplinary rehabilitation programme for persons with brachial plexus injuries. *Disabil Rehabil* 31: 1-8.