Answer to Case Study

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Presentation of the case on page 2.

Diagnosis

Parsonage-Turner Syndrome.

Discussion

The Parsonage-Turner syndrome (PTS) is a rare disorder of unknown aetiology characterized by acute and intense sudden-onset pain in shoulder or upper limb, showing later progressive muscle weakness and hypotrophy.

Feinberg describes this entity in 1897 reporting a case of complete unilateral brachial neuritis in brachial plexus associated with the flu. Afterwards, the most relevant report on acute brachial neuritis was Spillane's, in 1943, who reported on localized shoulder neuritis in 46 soldiers. However, this disease was not named as we know it today until 1948, when Parsonage and Turner described a series of 136 cases during Second World War, naming their findings neuralgic amyotrophy or scapular girdle syndrome. Other terms used to describe this entity are brachial plexus neuropathy, paralytic brachial neuritis, acute idiopathic brachial neuritis, and acute brachial radiculits. The entity prefers the cervicobrachial plexus, but it is not always limited to it. It can involve multiple nervous that originate in the brachial plexus. According to publications on the subject, the long thoracic, suprascapular and axilar—individually or combined— nerves are the ones affected to the greatest extent, as medical assessment and electromyographic findings show. However, there is no agreement on the works that we consulted as regards the peripheral nerve most frequently affected. According to the earliest Turner and Parsonage's studies, the long thoracic nerve is the most frequently involved. Later reports suggest that it is the suprascapular nerve the one most frequently affected. On the other hand, the nerves least frequently affected are the ulnar, the radial and the median nerves. It is worth highlighting that the effects on the long thoracic nerve —which innervates the anterior serratus muscle—, usually are not appropriately evaluated by images because in general this muscle is excluded from the vision field in routine shoulder MRI studies. If the long thoracic nerve is compromised, the patient can develop any scapular defect right up to scapula alata (winged scapula). With this medical finding, MRI studies could be complemented widening the vision field. There are also reports on the lesion of the phrenic nerve with diaphragmatic dysfunction.

Since in most cases only one or two nerves are compromised, the PTS can be interpreted as a mononeuropathy or a polyneuropathy instead of a true plexopathy.

Although the precise causes of PTS remain unknown, in specialized bibliography it has been attributed to (viral and bacterial) infections, and also to autoimmune processes, as well as immunization. Some pathogenic agents have been correlated with PTS, such as smallpox, typhoid fever, flu virus, coxsackie virus, B19 parvovirus, cytomegalovirus, human immunodeficiency virus and *Borrelia burgdorferi.* There is plentiful evidence of the PTS association with viral infections—there are publications about epidemic outbreaks in isolated populations such as the one that occurred in a native population in the South-western US, with eight cases of PTS.

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A genetic component could predispose an individual to PTS: there are reports on a hereditary condition that is similar to it from the medical point of view, although it is a different one, called hereditary neuralgic amyotrophy. This autosomal dominant disorder is characterized by painful, episodic, recurrent brachial neuropathy in association with mild dysmorphic features.

Likewise, there are reports on the association between PTS and recent surgery plus anaesthesia, delivery and even extenuating physical activity.

However, the role that all these factors play in the PTS is still hypothetical, because they could not be proved.

Most patients are going through from the third to the seventh decade of life, but reported ages oscillate between three months and 82 years of age. There is a clinical study reporting incidence rates of 1.64 cases per 100,000 people. The PTS prevails in males, with a 2:1 to 11.5:1 male-to-female ratio, according to different studies. There is no evidence of any PTS tendency to compromise one upper limb rather than the other one, nor is there any of PTS correlating with hand dominance. In up to one third of the patients, there are reports on bilateral involvement, but these conditions are usually asymmetric in terms of the affected muscles and the degree of damage.

The main symptom at the time of presentation is shoulder pain. The PTS pain onset is usually sudden and, oftentimes, very intense, awaking the patient in the middle of the night. It worsens progressively in hour or days, it usually lasts from few hours up to three weeks, and it usually recedes suddenly as well, although sometimes, a mild residual pain remains. It exacerbates with the upper limb movements and by pressure on muscle masses.

Pain is distributed across the shoulder blade to the acromion, and radiates along the external aspect of the arm and the neck and, rarely, it is distal to the elbow. There is no exact correlation between pain localization and subsequent distribution of paresis. However, pain radiation below the elbow is associated with the damage of the biceps or the triceps muscles, whereas pain radiation to the neck implies that the ones compromised are the sternoclestomastoid and the trapezius muscles. Time up to paresis onset is highly variable. Palsy is flaccid, patchy and, in general, progressive, affecting especially shoulder elevation and upper limb abduction. The PTS is a condition of the lower motor neuron type, with hypoactive reflexes and sometimes fasciculations.

Paresis —also of variable degree— in most cases continues spreading throughout the month consecutive to the symptoms onset. Paresis can be associated with atrophy.

On the basis of the anatomic location of the lesion, there are reports on several patterns of paresis, whether there is just peripheral nerve or roots damage, or both of them are compromised. The most frequently described pattern is that of damage of several peripheral nerves (suprascapular, axilar, long thoraxic and musculocutaneous nerves).

With respect to nerve roots, the most frequently affected are C5 and C6, which are included in the upper trunk of the brachial plexus.

Regarding the affected muscles, the most frequently reported is the deltoid muscle, followed by the supraspinatus, the infraspinatus, the serratus major, the biceps, the triceps and the extensor digitorum muscles.

At the onset, many patients report localized hypoesthesia, although such sensitive disorders are discrete; the area most severely compromised is that of the distribution of the axilar nerve.

Oftentimes, the PTS is taken mistakenly by other disorders which are better known and more common, such as cervical spondylosis, rotator cuff disorders, shoulder impingement syndrome, adhesive capsulitis, and acute calcific tendinitis of the shoulder. Therefore, knowledge about the PTS and its characteristic findings in imaging studies determine that, more often than not, it is the specialist in imaging diagnosis the first one that suggests the right diagnosis, preventing the patient from receiving inappropriate treatments or even an unnecessary surgery.

Nowadays, there is no test to diagnose the PTS in a specific way. Medical history and findings in physical examination, electromyography and imaging studies are used to confirm PTS diagnosis. Among them, MRI is the diagnostic method of choice, because it shows the shoulder anatomy in multi-planar sections and it is sensitive enough so as to evidence muscle oedema in acute cases (in T2-weighted sequences, with or without fat suppression), as well as fat hypotrophy in chronic cases (in T1-weighted sequences). It is also useful to rule out any other structural disorder that can emulate this condition, such as glenoid labrum lesion with synovial cyst, associated with secondary compression on the nerve path, as well as general pathology with rotator cuff injury.

MRI findings in the PTS convey lesions by denervation. It has been proved that, after an isolated acute neurologic lesion, at the beginning the denervated muscle looks normal. Usually changes in the MRI signal start at approximately 48 hours after the denervation lesion. There is controversy over the increase in MRI signal in fluid-sensitive-weighted sequences (T2, STIR, proton density and T2 with fat suppression) being due to an increase in the extracellular fluid or due to an increase in the volume of capillary blood circulating in the partially denervated muscle. Most probably, it is a combination of both.

Either in one case or the other one, the neurogenic abnormality that underlies the increase in signal in fluid-sensitive-weighted MRI sequences is not specific in itself and can be caused by entities other than the PTS—traumatism, entrap-

ment neuropathy caused by the effect of a local mass (due to a cyst in the spinoglenoid notch, for instance), and cervical slipped discs.

The quadrangular space syndrome can emulate the PTS too, although in general, it has an insidious onset and compromises only the axilar nerve.

There are also reports on suprascapular neuropathy as a scarcely frequent chronic lesion associated with a combination of predisposing anatomic conditions and sport activities (particularly volley ball and, especially, throwing in baseball). In general this neuropathy shows in overhead sports, and patients show painless atrophy in the infraspinatus muscle (nerve damage around the spinoglenoid notch). Proximal nerve lesions (around the suprascapular notch) will include the supraspinatus muscle too.

Predisposing anatomic conditions might include: a thick or calcified superior transverse scapular ligament, the presence of an anterior coracoscapular ligament, narrow spinoglenoid or suprascapular notches, and a hypertrophic subscapular muscle (which may cover the anterior aspect of the suprascapularis notch). These conditions can emulate the PTS in images. However, if they do, in general they are associated with different medical findings and neither the axilar nor the subscapular nerves are compromised.

Likewise, there are other causes for abnormal T2-signal within the muscle that should be excluded, such as myopathy, myositis, and tumors.

In acute/subacute stages, MR neurography can also be useful, since it shows diffuse thickening and increase in signal intensity in the T2-weighted sequences of the affected nerves in case of brachial plexus pathology.

Electromyography and nerve conduction velocity can show changes associated with acute denervation in the distribution of the brachial plexus.

Not only electromyography but also MR images should be interpreted in the context of medical history.

All in all, the PTS can be suspected when:

1) MRI reveals in muscle tissue an abnormal increase in signal in fluid-sensitive-weighted sequences (T2, STIR, proton density and T2 with fat suppression) distributed in a pattern consistent with an injury of a peripheral nerve.

2) Medical findings are consistent with PTS, what include the patient lacking in history of traumatism or overhead sport activity.

3) MRI confirms the absence of any morpholgical cause of denervation (such as entrapment neuropathy caused by a space-occupying lesion).

The treatment of the PTS is not a surgical treatment, and includes pain-killers and physical therapy for weakness. In general is a self-limiting condition, although there are reports on low recurrence rates.

Conclusions

The PTS, also known as acute idiopathic brachial neuritis, is a non-traumatic painful disorder that affects the scapular girdle. Oftentimes patients consult for sudden-onset pain in his or her shoulder or weakness (or both) in the muscles of their scapular girdle. From the medical point of view, making diagnosis can come as a challenge, because symptoms are unspecific and can emulate those in other disorders of the scapular girlde, such as glenoid labrum tear (with or without associated paralabral cyst), rotator cuff rupture, impingement, and adhesive capsulitis. The assessment of a patient with pain and weakness in his or her shoulder includes medical history, physical examination, imaging studies and sometimes electrophysiology studies.

MRI is the imaging technique of choices in patients with shoulder pain and weakness, and it is the most complete evaluation method for the scapular girdle due to the possibility it gives to get images at different-plane level and characterize soft tissues. Familiarization with the characteristics of the PTS in MRI is essential for specialists in imaging diagnosis, because they may be the very first ones in suggesting diagnosis. These characteristics can actually be pinpointed by screening muscle oedema with specific distribution in one or in several muscles with denervation pattern.

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