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Original Article

Parsonage-Turner syndrome: a firsthand experience of an uncommon malady

Vinod P Jerath[1], Vikram K Mahajan[2]

[1]Dr Jerath's Skin, VD and Allergy Centre, Jalandhar 144001, Punjab, India; [2]Department of Dermatology, Venereology & Leprosy, Dr. Rajendra Prasad Government Medical College, Kangra (Tanda) 176001, Himachal Pradesh, India

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Abstract: Parsonage Turner Syndrome (syn. idiopathic brachial plexopathy, neurologic amyotrophy) is a rare syndrome of poorly understood etiology with a reported incidence of 1.64 in 1 lakh persons per year. It affects men more often than women with a highest incidence in the third and seventh decades of life. Its pathophysiology is obscure and the syndrome has been reported in the postoperative, post infectious and recent viral illness, and post-vaccination settings. Trauma from manipulation of tissues and various positioning techniques used to facilitate surgical techniques, or immune-mediated inflammation remains the most common associated risk factor. It mostly remains under diagnosed for lack of clinical suspicion and specific diagnostic tools. Herein, we share a personal experience of this uncommon disorder by the first author, a healthy 67-year-old man, having no significant medical or surgical disorder and presenting with several weeks of weakness of right shoulder. The diagnosis was made after its aggravation following stretch injury sustained from a fall with upper limbs in full abduction. Treatment with high dose dexamthasone (100 mg in 200 ml 5% dextrose given once by slow i.v. infusion), NSAIDs (as needed) and physiotherapy given over 12 months was remittive.

Keywords: Brachial plexus, brachial plexus neuropathy, idiopathic brachial plexopathy, neurologic amyotrophy

Introduction

Parsonage Turner Syndrome (syn. idiopathic brachial plexopathy, neurologic amyotrophy) is a rare syndrome of poorly understood etiology with a reported incidence of 1.64 in 1 lakh persons per year. It affects men more often than women of any age with a highest incidence in the third and seventh decades of life [1]. Typically, it presents with severe shoulder pain of acute onset that is commonly unilateral, often constant and self-limiting lasting for 1-2 weeks. However, it may resolve with delayed upper limb weakness, muscle atrophy, and painless paraesthesias [2]. Most patients show 80-90% recovery of muscle strength in 2-3 years but residual paresis and exercise intolerance happen in up to 70% patients [3]. Its pathophysiology remains obscure and the majority of the cases have been ascribed to surgical procedures, trauma, recent viral infections (varicella virus, herpes simplex, HIV, Coxsackie B virus, Hepatitis B virus, Hepatitis C virus, Epstein-Barr virus, cytomegalovirus, SARS-CoV2), or vaccinations, and autoimmune disorders such as systemic lupus erythematosus, polyarteritis nodosa, and temporal arteritis [1, 4-6]. Operative manipulation of tissues, various positioning techniques used to facilitate surgical techniques, and immune-mediated inflammation have been implicated in post-surgery cases and the symptoms usually develop within 24 hours to a week thereafter [2]. A possibility of underlying genetic component too has been suggested [7].

The diagnosis is primarily based on clinical history, symptomatology and thorough physical examination with extensive manual muscle testing. Radio imaging work-up will be needed to exclude other pathological abnormalities involving shoulder/glenohumeral joint or other disorders causing neuropathy/musculopathy

[8]. Magnetic resonance imaging (MRI) is a modality of choice to exclude other causes of shoulder pain such as rotator cuff tear, labral tear, or impingement syndrome and to demonstrate sequalae of denervation of skeletal muscles innervated by brachial plexus nerves [9]. The denervated muscle will appear normal but diffusely increased T2-weighted signal due to intramuscular edema will be the earliest detectable abnormality while corresponding T1-signal abnormality may be absent in acute phase [9, 10]. On the other hand, electromyography (EMG) can better identify, isolate, and grade severity of denervation, and re-innervation (after a period of time) of the involved muscles [4]. Treatment of PTS is primarily for pain management with NSAIDs, neuroleptics, and/or systemic corticosteroids with or without immunomodulators. However, recovery time reportedly lasted between 6 and 18 months with recurrence rates of 25% and 75% with these therapies [11, 12]. Additionally, there is some evidence to suggest benefits of using intravenous immunoglobulin (IVIg) [13]. However, PTS mostly remains under diagnosed for want of clinical suspicion or specific diagnostic tools. Herein, we share a personal experience of the first author of this uncommon disorder.

Case presentation

Two years back the first author aged 67 years, otherwise healthy man playing lawn tennis regularly for last 40 years, could not serve the ball to the nets on three successive days. He had no recent illness, surgery/trauma, or neck/shoulder pain. Physical examination showed muscle strength of grade 3 in right shoulder and grade 5 in the left shoulder, and normal tendon reflexes for triceps, biceps, and brachioradialis. On two-point discrimination test there was sensory loss over lateral and posterior half of right forearm over areas innervated by musculocutnaoeus nerve (C5, C6).

Lab investigations and x-rays of shoulder joints and cervical spine showed no significant abnormality. A magnetic resonance imaging showed acromioclavicular arthropathy, small intraosseous ganglion cyst in the head of humerus posterio-laterally and anterior to bicipital groove, elongated cystic lesions in the deep fibers of the infraspinatus muscle along the posterior scapular border, sub supraspinatus tendinopathy, sub-acromion bursitis of right shoulder joint, and mild edema and infiltrate of the brachial plexus. The electroneuromyography (EMG) demonstrated acute proximal multiple bilateral amyotrophic neuropathy. The overall findings were considered age-related degenerative changes and sequalae of old focal muscle tear. Use of cervical collar for neck support and physiotherapy was advised with a provisional diagnosis of radiculopathy at C5, C6 level.

After about 40 days he had a fall in prone position with arms in full abduction and remained in that position for almost 3 hr. Clinical examination showed a swollen left arm with hypoesthesia and weakness of right arm with range of motion for elevation, and internal and external rotation restricted to 80°, 10° and 10° for the left shoulder and 10°, 10° and 40° for the right shoulder, respectively. Repeat radio-imaging studies showed additional findings of intramuscular edema in the rotator cuff muscles, Bankart lesion in the anterior/inferior glenoid labarum and increased brachial plexus edema as before.

With overall features a diagnosis of Parsonage Turner syndrome (PTS) was made and he was treated with 100 mg dexamethasone in 200 ml of 5% dextrose by single intravenous infusion given over 3 hr along with NSAIDs and physiotherapy as follow-on treatment until symptomatic relief. Noticeable improvement occurred and ability to play lawn tennis ensued over next 12 months.

Discussion

Clinically, PTS presents with pain of variable intensity involving shoulder girdle of abrupt onset usually unrelated to hand dominance or the left versus the right side [4]. As it wanes over 1-4 weeks, weakness and atrophy of the shoulder girdle and upper limb muscles associated with abnormal reflexes and sensory deficit ensue in 60% cases [4]. Supra scapular nerve, long thoracic nerve, anterior interosseus nerve, axillary nerve and the phrenic nerve are the commonly affected nerves in such a scenario but it is not uncommon for ulnar, radial and median nerves to get afflicted [4, 14]. An early diagnosis and treatment is imperative to prevent neuropathy, muscle atrophy and disability in the long-term. A high index of clinical suspicion along with extensive physical examination is important for diagnosis while MRI is mostly

required to exclude other pathologies [4]. In this case, the initial presentation was confused with radiculopathy at C5, C6 level related to age-related degenerative changes and sequalae of old focal muscle tear from playing tennis. Injury to the brachial plexus may be because of a variety of mechanisms including trauma/compression. Traumatic brachial plexopathies could be from stretch injury as was from forced abduction of arms in this case. This seems most plausible explanation in this case for being akin to trauma from manipulation of tissue and various positioning techniques used to facilitate surgical techniques and/or subsequent inflammation with symptoms usually developing within 24 hr to a week thereafter [4].

In general, the initial treatment is aimed at relief in muscle/nerve pain with NSAIDs gabapentin, carbamazepine, and/or amitryptiline. Despite doubtful efficacy high-dose systemic corticosteroids early in the disease process, with or without immune-modulating therapies, NASIDs and physiotherapy remains the mainstay of treatment to preserve muscle strength and joint motions. It takes 6 months to 3 years for a noticeable effect in about 90% cases [14]. However, <10% of patients in a cohort of 246 patients continued to have pain or sustained loss of function after 3 years [11]. Treatment with single session of high dose dexamthasone i.v. infusion with follow on NSAIDs and physiotherapy for about 12 months lead to complete recovery without significant sequalae in this case.

Comments

Parsonage-Turner syndrome is a rare disorder of varied etiology and obscure pathophysiology. It may present to an orthopedist for sudden onset with or without acute shoulder pain wherein it can be mistaken for degenerative cervical spondylosis, radiculopathy or rotator cuff injury. The condition may evolve to weakness of the musculature of the shoulder girdle, and scapular belt in the absence of treatment that itself remains as varied as its clinical presentation. The loss of shoulder function may become irreversible in the absence of adequate treatment and early rehabilitation. The diagnosis of Parsonage-Turner syndrome is often delayed for want of clinical suspicion. It should be borne in mind in cases of sudden intense pain/weakness in the neck and shoulder regions, hypoesthesia and paresthesias in presence of preceding forced stretch injury. However, a multidisciplinary team of orthopedist, neurologist, rheumatologist and internist will be needed for neurological evaluation, extensive manual muscle testing, and specific diagnostic evaluation. Early administration of systemic corticosteroids, NSAIDs and physiotherapy remains preferred first-line treatment. However, well defined diagnostic tools and treatment protocols remain highly desirable.

Disclosure of conflict of interest

None.

Address correspondence to: Dr. Vikram K Mahajan, Department of Dermatology, Venereology & Leprosy, Dr. Rajendra Prasad Government Medical College, Kangra (Tanda) 176001, Himachal Pradesh, India. Tel: +91-1892-287161; Fax: +91-1892-267115; E-mail: vkm1@rediffmail.com

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