CLINICAL REPORT

Idiopathic spinal accessory nerve palsy. A case report

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Introduction

The spinal accessory nerve, otherwise termed cranial nerve XI, is the sole motor innervator of trapezius. This muscle is composed of three functional components and is a major stabilizer of the scapula. It contributes to scapulothoracic movements by elevating, rotating and retracting the scapula. A trapezius palsy may cause pain, loss of active shoulder abduction, and winging of the scapula. One of the most common aetiological factors is iatrogenic injury during various surgical procedures in the posterior cervical triangle of the neck, such as lymph node biopsy, tumour excision and radical neck dissection. Idiopathic paralysis of the spinal accessory nerve has also been reported. We present a scarce case study of a 42-year-old male who presented with a left-sided trapezius paralysis due to an idiopathic palsy of the distal segment of cranial nerve XI. Evaluation of such a case may pose a diagnostic and treatment challenge for any physician neurologist or orthopaedic surgeon.

Case report

A 42-year-old Caucasian male presented to our outpatients department complaining of a constant dull ache located in the left shoulder girdle and the posterolateral side of the neck, associated with sudden difficulty during overhead activities and heavy lifting. He worked as a heavy manual labourer. There was no history of trauma, falls or any precipitating events. There was not mentioned any recent vaccination. The onset of his symptoms occurred 10 days ago with an acute left-sided neck and shoulder girdle pain. He
described the pain as intermittent and sharp, ranging from 6 to 10 out of 10 on a numeric pain rating scale. The pain was worse during the night, often severe enough to prevent him from sleeping. It radiated to the left periscapular region without any neurological symptoms in the arm. A week later, he noticed weakness of his left shoulder during abduction and anterior elevation, whilst his pain subsided markedly. There were no associated headaches or left upper extremity numbness nor any severe motor weakness. The patient was self-medicated on paracetamol and nimesulide. His past medical history was unremarkable.

Physical examination revealed winging of his left scapula and severe atrophy of the left trapezius muscle. He also had an asymmetrical neckline, drooping of the shoulder girdle and weakness of forward elevation (Fig. 1). There were no signs of sternocleidomastoid muscle paresis. Passive range of motion of his left shoulder was within normal limits. Active abduction and forward elevation were minimally limited compared to his contralateral (asymptomatic) side. The patient was unable to abduct and forward flex the left arm above 70° and 85° respectively. There was no weakness of external rotation or elbow flexion. Range of motion of his cervical spine namely flexion, extension and lateral rotation was mildly limited with some end range axial pain. Spurling’s test was negative on the left. Muscle power throughout the left upper limb was 5/5, except of a mild weakness (4+/5) of the left triceps and a severe weakness (2/5) of the left trapezius. There was no sensory loss in the left upper limb. The rest of the physical examination revealed normal muscle bulk and tone, normal sensation and symmetrical upper extremity reflexes. Hawkins, Neers, Roos, Wrights and Adsons provocative tests were all negative.

Haematological investigations revealed a normal full blood count and erythrocyte sedimentation rate. Immunoglobulin levels were within normal limits. Plain radiographs and MRIs of the cervical spine and shoulder region were normal. Electromyography was carried out almost 3 weeks after the onset of symptoms and revealed denervation in the upper, middle and lower components of the left trapezius muscle with positive sharp waves and a reduced polyphasic recruitment pattern. Nerve conduction studies demonstrated slowing of the distal segment of the spinal accessory nerve. The levator scapulae, supraspinatus,
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Infraspinatus, teres major, rhomboid major and the deltoid muscles showed no signs of denervation (Fig. 2).

Consequently, the possibility was raised that our patients palsy may have been purely idiopathic. There were no signs indicative of any other pathology, such as paresis of the sternocleidomastoid muscles, which is caused as a result of a palsy of the proximal root of the accessory nerve. This is the case in neurofibromas or meningiomas in the base of the skull that cause pressure effects. Moreover, there was no history of trauma, falls or any precipitating events. There were no signs of previous surgical procedures in the trapezius, shoulder girdle or neck area. The possibility of a space-occupying lesion or a hereditary abnormality in the neck was ruled out by physical examination and radiological investigations. An idiopathic accessory nerve palsy is usually inflammatory in nature and could possibly be due to a focal mono-neuritis or as part of a brachial plexus neuritis (Parsonage-Turner syndrome) [1,3,4].

Nevertheless, with the use of analgesics and the help of physiotherapy, our patient made a full recovery 6 months following the onset of his symptoms. At his last outpatients follow up, his left trapezius muscle power tested 5/5. In addition to that, he had a full range of motion in his left shoulder and cervical spine.

Discussion

The trapezius is the largest and most superficial of the scapulothoracic muscles. Scapular and humeral motion occur simultaneously during movements of the upper extremity. The trapezius is one of the main muscles responsible for scapular movements [1]. Overall, it elevates, retracts and rotates the scapula. It consists of three parts, which all have different actions. Specifically, the upper acromionclavicular fibres raise the shoulder girdle, upwardly rotate the lateral angle and prevent it from sagging under weight. The intermediate horizontal fibres arise from the vertebral spines, bring the medial edge of the scapula 2 to 3 cm closer to the midline and press the scapula against the thorax. The intermediate fibres adduct and retract the scapula and move the shoulder girdle posteriorly. The inferior fibres pull the scapula inferiorly and medially; depress and rotate it downwards [5].

The spinal accessory nerve, or cranial nerve XI, is the sole motor innervation to trapezius. The nerve is usually joined by proprioceptive branches from the second, third or fourth cervical nerves. The accessory nerve leaves the skull through the jugular foramen. The nerve lies close to the internal jugular vein, and just below the lateral mass of the atlas it reaches the inferior surface of the sternocleidomastoid muscle. It emits motor fibres to the muscle, passes underneath it or penetrates its deep head, reappearing under its posterior edge between the upper and middle third [6]. Occasionally however, the nerve terminates in the sternocleidomastoid. In this case, trapezius is innervated by cervical nerves [1,2]. The spinal accessory nerve crosses the subcutaneous tissue on the floor of the posterior cervical triangle very superficially, before it enters the trapezius muscle. Its superficial location makes it susceptible to injury [2,6]. The nerve may be injured during various operations in the posterior cervical triangle, such as lymph node biopsy, simple tumour excision and radical neck dissection [6—12]. Penetrating injury due to trauma may also damage the spinal accessory nerve [1,6,7,12,13] and Patterson reported trapezius palsy after acromioclavicular or sternoclavicular dislocations [14]. Rare iatrogenic causes that have also been reported are during carotid endarterectomy and catheterization of the internal jugular vein [15,16]. Idiopathic paralysis of the spinal accessory nerve has also been reported [2,3,4,17—19].

In the presence of trapezius dysfunction due to an idiopathic spinal accessory nerve palsy, the patient often initially presents either with an acute sharp, and intermittent shoulder girdle pain, or with a constant, poorly localized dull ache located in the neck and periscapular area. The pain usually worsens at night, sometimes waking the patient from sleep. After a few days weakness of abduction and forward elevation of the shoulder slowly appear, whilst pain subsides. In a few weeks, atrophy of the trapezius muscle becomes clinically obvious with an asymmetric neckline, a drooping shoulder and a winged scapula [1,2,6,9,20,21].

Trapezius paralysis is frequently misdiagnosed during the initial presentation. The differential diagnoses for a trapezius palsy should include: paralysis of the serratus anterior or rhomboid muscles, herniated nucleus pulposus, scoliosis, progressive neuromuscular disorders, scapular osteochondromata, fracture malunion, stroke, herpes zoster infection and glenohumeral instability [1,20,21].

The most helpful diagnostic tests in confirming trapezius paralysis is electromyography and nerve conduction studies. They assess the condition of the trapezius, sternocleidomastoid, levator scapulae, deltoid, supraspinatus, infraspinatus, teres major, rhomboid major and minor muscles. The long thoracic nerve and serratus anterior are also tested for pathology. Abnormalities that can be associated with different neuropathies and nerve palsy include decreased amplitude and prolonged distal latency [22]. In general, the electromyographic findings of accessory nerve palsy are presented usually 3 weeks after the onset of symptoms [23]. Plain radiographs of the cervical spine, chest, and shoulder, although rarely diagnostic, are mandatory. Computed tomography (CT) and magnetic resonance imaging (MRI) scans are not necessary unless a space occupying lesion or disc disease is suspected [1,2,6,20].

For idiopathic spinal accessory nerve palsy the treatment is nonoperative. With analgesia and physiotherapy more than eighty per cent of the patients fully recover in 6 to 12 months. Physiotherapy helps to maintain a full range of motion in the shoulder and cervical spine and assists to maximise muscle function [2,18,19,21,22].

If spinal accessory nerve paralysis is caused by trauma, early surgical exploration of the nerve is indicated when there is no clinical or electrical sign of recovery up to 3 months after the injury. Microsurgical repair of the nerve (neurolysis, nerve graft or neurorrhaphy) may be performed as long as 20 months after the injury. In their study, Teboul et al. [2,18] obtained good results in the majority of their patients. In cases of spontaneous trapezius palsy, failure of surgery or when the recommended time period for microsurgical nerve repair (20 months) has elapsed, muscle transfers can be performed.

Surgical treatment of trapezius palsy has evolved from static procedures, such as scapulothoracic arthrodesis, to the current dynamic procedures of transferring the rhom-
boids major and minor and the levator scapulae, the so-called Eden-Lange procedure. Recent studies, such as Galano et al. [22], Teboul et al. [2,18] and Romero and Gerber [24] have presented pain relief, improved clinical results (shoulder function restored), with no residual winging scapula and without or very few postoperative complications. The results of such procedures may not be as good in individuals who are older than 50 years and physiotherapy should be considered as the primary treatment method in this selected group of patients [2,19—22,24,25].

Conclusion

Idiopathic spinal accessory nerve palsy is an infrequent neuromuscular disorder, which predominantly affects the shoulder girdle. It is characterized by dysfunction or paresis of the trapezius, which can be a painful and disabling condition. The shoulder droops as the scapula is translated laterally and rotated downwards. The patient also presents with an asymmetrical neckline, a drooping shoulder, winging of the scapula, and weakness of forward elevation. The diagnostic evaluation should be based on history and physical findings and is confirmed by electromyography. The prognosis is usually excellent and treatment is supportive with analgesia and physiotherapy.

Conflict of interest statement

The authors declare that they have no competing interests.

References