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Neuralgic Amyotrophy in a 66-year-old Hiker: a Case Report

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ABSTRACT

Introduction: Neuralgic amyotrophy (NA) or Parsonage Turner syndrome is a clinical syndrome characterized by sudden attack of neuropathic pain, motor weakness and sensory loss that could be more or less clinically present. Different interpretations regarding the differential diagnosis, symptoms, cause and treatment were given till now. **Aim**: We report our experience with a 66-year-old male who had a sudden pain attack and palsy in shoulder region, without sensory loss, and associated calcific tendinitis of rotator cuff and degenerative changes in cervical spine. **Case report**: Patient came to our hospital with strong pain in shoulder area and signs of frozen shoulder. Active abduction and anteflexion was only to 30 degrees. He experienced the intense pain with visual analogue scale (VAS) 10/10 a night before, during his sleep. No trauma. Neurontin (gabapentin) was given to the patient. After 4 days, he felt better with abduction and anteflexion to 90 degrees. After 2 weeks VAS was 3/10, abduction and anteflection to 100 degrees. **Conclusion**: Neuralgic amyotrophy (NA) is a self-limiting inflammatory disorder usually with idiopathic etiology. The condition *can be challenging* to treat since many associated symptoms and diagnostic tests and procedures may mimic NA. In that case, accurate differential diagnosis is essential.

Keywords: Neuralgic amyotrophy, CT Arthrography, Neurontin.

1. INTRODUCTION

Neuralgic amyotrophy (NA) or Parsonage Turner syndrome is a clinical syndrome characterized by sudden attack of neuropathic pain, motor weakness and sensory loss that could be more or less clinically present (1, 2)its pathophysiology remains unknown. An inflammatory (auto. Different interpretations regarding the differential diagnosis, symptoms, cause and treatment were given till now (2)pathophysiology, epidemiology, and diagnostic and therapeutic strategies in neuralgic amyotrophy (NA. After more than a century, the pathophysiology of the syndrome is still unknown. Neuralgic amytrophy in the literature also stands for the Parsonage-Turner syndrome or brachial plexus neuritis.

Neuralgic amyotrophy is more frequent in men than women, and is almost always asymmetric. NA has an annual incidence of 2/100.000 people, but recently it goes up to 1/1000 people, as more physicians (especially general practitioners) start to recognise this syndrome (1-3). The classical clinical pattern of NA includes 3 successive phases: painful phase, then weakness, amyotrophy and sensory complaints, then recovery (1). These 3 phases occur similar both in idiopathic neuralgic amyotrophy (INA) and hereditary neuralgic amyotrophy (HNA).

Patients with INA usually suffer only one attack in their life, and the rate of recurrence varies from 5% to 26%, compared with 74% in HNA patients who are subject to frequent recurrences (3).

In INA, an age of onset is usually in the second or third decade, but ranging from the neonatal age to the seventh decade. Patients with HNA tend to be younger when their first attack occurs, usually in their 2nd decade (1-4). An inflammatory (auto)immune pathophysiology is presumed, with mechanical or infectious precipitating conditions, which triggers attacks.

2. AIM

We report our experience with a 66-year-old male who had a sudden pain attack and palsy in shoulder region, without sensory loss.

3. CASE REPORT

Patient came to our hospital with strong pain in shoulder area and signs of frozen shoulder. He could hardly move his arm. Active abduction and anteflexion was only to 30 degrees. He experienced the intense pain with visual analogue scale (VAS) 10/10 a night before, during his sleep. No trauma. This was his first episode of such a problem. Familiar anamnesis was negative. Clinically, he had unilateral motor palsy without sensory loss and amyotrophy in periscapular region. X-ray showed calcific deposit in supraspinatus tendon in the process of absorption (Figure 1).

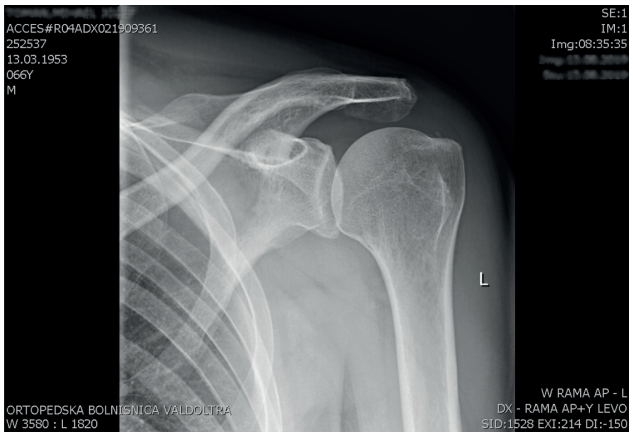

Figure 1. X-ray–calcific deposit in resorptive phase.

We assumed that the pain was due to the calcific absorption and applied corticosteroid with 2%lidocaine injection in subacromial space and glenohumeral joint. Patient did not feel better. Due to the age of patient and unclear anamnesis of the pain during the night, we made a CT arthrography (Figure 2) which was normal, without rotator cuff tear signs, no compression or cyst in the area of suprascapular nerve or any associated lesion.

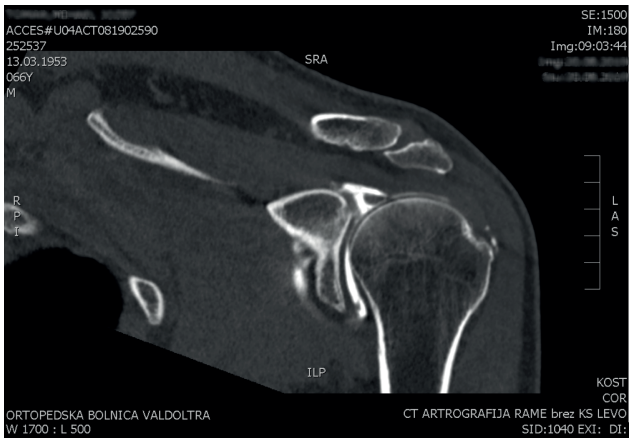

Figure 2. CT arthrography–left shoulder with calcific deposit in resorptive phase, no rotator cuff tear.

Chest X-ray (Figure 3) excluded compression mass in the upper lungs (Pancast tumor) which may compress the brachial plexus creating similar symptomatology. Neurological tests for the involvement of the peripherical nerve of the upper limb were negative except for the suprascapular nerve with involvement of both supraspinatus and infraspinatus muscle unilateraly.

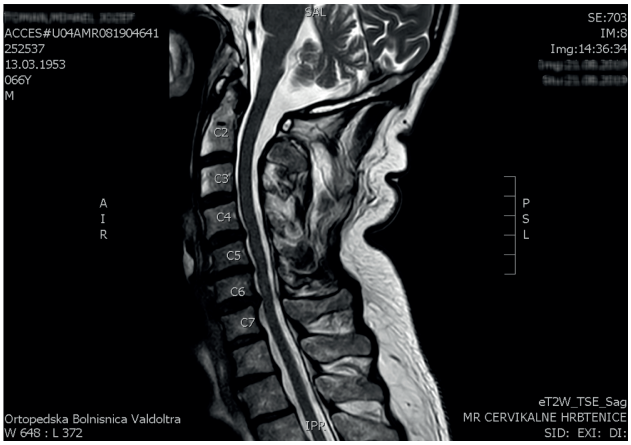

Figure 3. Chest X-ray was normal.

MRI of the cervical spine (Figure 4) showed slight degenerative changes, slight diffuse stenosis at the level C4-C5 and C5-C6. Disc herniation was excluded. MRI was shown to a spinal surgeon who concluded that the stenosis was inconsistent for such a symptomatology.

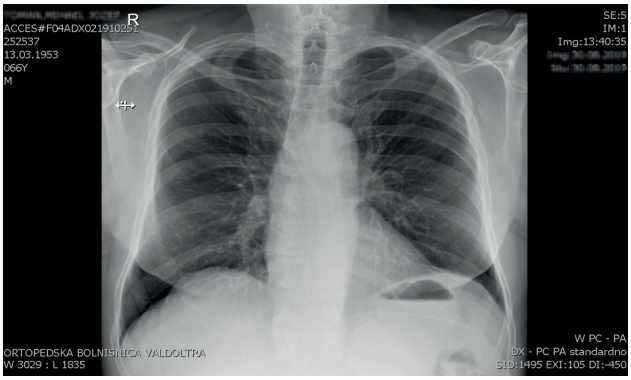

Figure 4. MRI shows diffuse stenosis at C4-C5 and C5-C6.

EMG of the upper limb and periscapular area was also normal, except two muscles: infra- and supraspinatus, where acute isolate motoric dysfunction was seen. Extended blood exams, including also borreliosis test, were negative.

Without trauma, but with sudden onset of shoulder pain, the treatment choice was very challenging. Neurontin (gabapentin) was given to the patient. After 4 days, he felt better with abduction and anteflexion to 90 degrees. After 2 weeks VAS was 3/10, abduction and anteflection to 100 degrees. Patient was send to physiotherapy for further rehabilitation. Control EMG showed some slight improvement but with still present neural inflammation. He started reducing pain killers. Globally he felt better and fully return to his everyday activities.

4. DISCUSSION

Our patient was referred to our hospital as capsulitis of unknown reason. Acute onset, very severe pain without trauma with good passive range of motion and almost absent active anteflexion and abduction of the shoulder. At anamnesis and clinical examination our first hypothesis was a tendinitis of supra- or infraspinatus tendon. Blood exams excluded infection of the glenohumeral joint.

Neuralgic amyotrophy can be recognized due to the extreme neuropathic pain at onset and the rapid development of uni or multifocal paresis and atrophy, usually in the

upper extremity, as well as the slow recovery in months to years. From the sensoric point of view, the presentation is atypical. Patchy hypaesthesia can be found (5,6).

The crucial problem of this pathology is that NA is still relatively unknown to many physicians. This makes the diagnosis challenging. Patients with NA are usually first seen by a family physician or physical therapists and after a first failure of conservative treatment referred to neurologists and/or orthopaedic surgeons.

The localisation and course differ from patient to patient and even among different attacks in the same patient.

More than 95% of patients complain of pain in neck, shoulder and/or arm regions. The differential diagnosis become difficult when patient has associated pathologies that may mimic NA (1,6).

Tendinitis is a pathologic condition of the tendon which compromises its function. The physiopathology is not completely clear. The cause seems to be a failure of the biologic process with uncontrolled reparation of damaged tendon (4,5).

One of the most common cause of Rotator cuff tendinitis is a Calcific tendinitis (CT). This condition is characterized by deposit of hydroxyapatite into the tendon (5). CT physiopathology goes through three phases. The first is a Calcific phase, where the deposit is creating. This phase is asymptomatic and the patients usually do not have symptoms. Second phase is a resorptive phase. This phase may have a rapid onset with intense pain that can radiate from the shoulder till the neck proximally and till the elbow distally. The third phase is a phase of rigidity, with contracture of the articular capsule. X-ray that was made showed a calcification in the supraspinatus tendon.

Considering the anamnesis, clinical manifestations and radiologic findings, it seemed logical that the patient had pain due to the resorptive phase. In the acute phase with intensive pain, corticosteroide with 2%lidocaine injection is indicated. We applied the intraarticular injection in the soft spot posteriorly and in the subacromial area. The patient was sent home and in case of persistent pain, the control visit was suggested. Non-steroid antirheumatic drug (NSAID) was also prescribed. After four days, patient had still severe pain and the NSAID, corticosteroid and lidocain injection did not make clinically positive effect.

Because the diagnosis was still unclear we proceeded with exclusion of other possible causes. Cervicobrachialgy is another possible cause of sudden pain in the periscapular-upper limb region. Herniation of the intervertebral disc in the cervical region produce usually symptoms involving ipsilateral dermatomes and myotomes that correspond to the cervical levels affected. Pain is a result of direct impingement of nerve roots against the hernia and associated inflammatory processes. MRI of the cervical spine is the diagnostic gold standard (7).

MRI show slight degenerative stenosis in the area C4-C5, C5-C6. Due to sudden onset of symptoms, slight compression and atypical clinical presentation, the cervical involvement was excluded.

EMG of the upper limb and periscapular region are the best modality to confirm a diagnosis of NA. Abnormal sensory potentials, lack of paraspinal denervation potentials, and abnormal conduction velocities are typical findings. Neurophysiologic studies should be performed 2-3 weeks after the onset of symptoms because there is no degenerative action potential in the acute stage (10,11). Due to positive anamnesis, clinic and EMG findings, NA was with high probability the right diagnosis.

Another possible cause of severe and unrelenting shoulder and arm pain can be Superior sulcus tumor, or as otherwise known Pancoast tumor. The clinical distribution of that chest apex tumor is usually in the region of the eighth cervical and first and second thoracic nerve trunks. Horner's syndrome (ptosis, miosis, and anhidrosis) and atrophy of the intrinsic hand muscles comprises a clinical entity named as "Pancoast-Tobias syndrome". It is a rare tumor, represent 3% to 5% of all lung cancers. In the more advanced stages we can detect it with chest X-ray. CT and MRI are more specific diagnostic tools (8,9). Due to atypical clinical presentation, negative chest X-ray and positive EMG, we excluded that diagnosis.

Treatment of NA is challenging. The pain is usually very resistant to pharmacological and even to sophisticated (e.g. nerve block) pain management. It is crucial to re-establish the normal biomechanics of the shoulder. Preventing the formation of the adhesions, maintain fluent periscapular motion and prevent dysfunctional compensating strategies (1).

5. CONCLUSION

Neuralgic amyotrophy (NA) is a self-limiting inflammatory disorder usually with idiopathic etiology. The condition can be challenging to treat since many associated symptoms and diagnostic tests and procedures may mimic NA. In that case, accurate differential diagnosis is essential.

- **Author's contribution:** All authors gave substantial contribution to the conception or design of the work, in the acquisition, analysis and interpretation of data for the work. All authors had role in drafting the work and revising it critically for important intellectual content. Each author gave final approval of the version to be published and they agreed to be accountable for all aspects of the work in ensuring the questions related to the accuracy or integrity of any part of the work. Final proof reading was made by the first author.
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