NEUROPATHIC ARTHROPATHY OF THE SHOULDER CAUSED BY SYRINGOMYELIA: A CASE REPORT

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ABSTRACT Neuropathic or Charcot arthropathy is a chronic degenerative disease that causes joint destruction in patients with abnormal sensation and proprioception. Charcot’s shoulder is rare, and its most common cause is syringomyelia. Patients present insidiously with joint instability, swelling and dysfunction. Pain may not be present due to sensory loss. The key to successful management is an early diagnosis by a thorough history, physical examination and imaging after exclusion of inflammatory, infectious and malignant aetiology. Most commonly, Charcot’s shoulders are managed conservatively. However, surgical intervention has recently proven to be a promising treatment for recovering function in selected patients. We present a 52-year-old female with chronic complaints of sensory loss and swelling of her right upper limb that progressively evolved into shoulder pain and marked dysfunction, which led to the diagnosis of syringomyelia associated with Chiari I malformation and for which she underwent posterior fossa craniectomy. Considering her past medical history and after exclusion of other aetiology, she was diagnosed with neuropathic shoulder arthropathy. She initiated conservative treatment, but the symptomatic and functional improvement was poor, highlighting the importance of early diagnosis and management of these patients.

KEYWORDS Charcot, Shoulder, Neuropathic arthropathy, Syringomyelia

Introduction

Neuropathic arthropathy, also known as Charcot arthropathy, is a chronic degenerative disease characterized by progressive joint destruction with bone and soft tissue involvement. Any condition that causes sensory or autonomic neuropathy can lead to a Charcot’s joint. Most commonly occurs as a complication of Diabetes. Other conditions include chronic alcoholism, leprosy, meningomyelocoele, myelodysplasia, spinal cord injury, syringomyelia, end-stage renal disease and congenital insensitivity to pain [1]. It is most common in weight-bearing joints. While upper limb involvement is rare, syringomyelia is the main cause of neuropathic shoulder arthropathy, with a reported prevalence of 6% [2]. Therefore, early diagnosis is crucial and should be considered after excluding infectious and rheumatologic aetiology in a patient with arthralgia and sensory changes.

Case report

A 52-year-old female, the domestic worker, presented with a 3-year history of pain and limited range of motion of her right shoulder. Past medical history revealed chronic complaints of numbness and swelling of a right hand that led to the diagnosis of syringomyelia extending from C1 inferiorly to T6 associated with Chiari I malformation (Fig.1), diagnosed 18 months before. The patient underwent surgery for posterior fossa craniectomy, removal of the posterior arch of C1 and dural graft by the neurosurgery department. She was taking pregabalin 600mg/day.

Physical examination showed pain on palpation of the anterolateral aspect of the right shoulder with no associated swelling or visible muscle atrophy. The shoulder range of motion was limited, with pain during active and passive mobilization, actively performing 60° of forward flexion, 20° of internal rotation and 20° of external rotation. No pain on cervical mobilization...
was elicited, and Spurling’s test was negative. The overall upper limb muscle strength was grade 4. Tendon reflexes were normal. Both pain and temperature sensation in the upper limb was decreased. However, the deep sensation was preserved. Hoffman’s test was negative. Impingement Hawkins-Kennedy, Neer and Jobe’s tests were positive.

The plain radiograph showed erosions of the glenoid and humeral head (Fig.2). Computed tomography (CT) scan showed marked diffuse sclerosis of the glenoid and humeral head associated with irregularity with geodes and osteophytosis, fracture along the anterior edge of the glenoid, periosteal peri-glenoid reaction, joint effusion with synovial thickening and intra-articular fragments (Fig.3). Magnetic resonance imaging (MRI) demonstrated a full thickness tear of the infraspinatus tendon, supraspinatus tendinitis, heterogeneity of the entire humeral head with multiple foci of increased uptake, muscle atrophy and fatty degeneration (Fig.4). Bone scintigraphy showed increased uptake in the humeral head and surrounding soft tissues.

Arthrocentesis was performed with 18 mL of citrine fluid aspiration and a synovial membrane biopsy under ultrasound guidance. No crystals were detected by light microscopy. Chemical, microbiologic and anatomopathological examinations were also negative. Complete blood count, biochemical analysis, erythrocyte sedimentation rate, C-reactive protein and rheumatoid factor were normal. Considering history and clinical and radiological findings, the patient was diagnosed with neuropathic shoulder arthropathy.

The patient started nonsteroidal anti-inflammatory drugs for the synovial inflammatory process and was referred for a physical therapy program including prevention of joint trauma and stretching, range of motion exercises and strengthening to reduce pain and promote functionality. At a 36-month follow-up, the patient reported partial relief of pain complaints. Physical examination revealed a very slight improvement of active mobilization, with forwarding flexion to 90°, internal rotation to 30° and external rotation to 30°. Rotator cuff muscle strength with internal and external rotation was preserved. There was no swelling. Both pain and temperature sensation in the upper limb remained decreased. She reported being functionally independent in self-care, maintaining major limitations in daily activities requiring overhead mobilization.

Discussion

Charcot arthropathy is characterized by the gradual destruction of the joints associated with neurosensory loss. Usually affects weight-bearing joints such as the ankle, knee, and hip. However, it occurs more frequently in diabetic patients in the foot and ankle [3].

Charcot arthropathy may also be secondary to syringomyelia, a rare disease (8.4/10000) that consists of developing a fluid-filled cavity or syrinx within the spinal cord. In most cases, syringomyelia is associated with Chiari malformation type I, involving the cervical region [4]. Other causes include basal arachnoiditis, basilar impression or masses such as arachnoid cysts and tumours, spinal pathology following trauma, haemorrhage, infection, radiation necrosis, ischemic, degenerative disease or tumour [5]. In addition, neuropathic arthropathy will develop in 25% of patients with syringomyelia, with 80% of these cases occurring in the upper extremity [6].

The pathogenic mechanisms involved are related, on the one hand, to the abolition of the neurovascular response caused by
Early diagnosis is widely recognized as the cornerstone to allowing treatment of the underlying cause, and joint protection measures and rehabilitation exercises are adopted to promote function [6, 18, 19]. Patients should therefore be referred immediately for neurosurgery to syrinx decompression. Several reports show decompression surgery slows down the degeneration process and maximises joint function. On the other hand, conservative measures must be addressed, such as restricted weight bearing, immobilization, passive stretching, physical therapy and nonsteroidal anti-inflammatory medication. For example, Deng et al. [11] presented clinical data from 12 patients with Charcot joints caused by syringomyelia, in which 5 underwent neurosurgical decompression and all reported neurologic improvement over a follow-up of 30 months. The remaining 7 patients underwent conservative treatment, 5 of whom experienced worsening symptoms during a 47-month follow-up. Atalar et al. [15] also reported 4 patients out of 5 with neuropathic arthropathy of the shoulder joint who had undergone neurosurgical decompression, and all showed improved range of motion and function. Accordingly, current studies advocate that early neurosurgical syrinx decompression may halt joint degeneration and thus stabilize or improve symptoms in patients.

However, and interestingly, in a recent report of a series of 10 patients with neuropathic shoulder secondary to syringomyelia, Wawrzyniak et al. [19] showed significant improvement in pain complaints in 8 patients who did not undergo decompression surgery and were only referred for reverse arthroplasty or conservative treatment with physical therapy and anti-inflammatory medication, in a 3-year follow-up. Until recently, arthroplasty or resurfacing surgery was not recommended because of the risk of implant loosening or failure due to muscle weakness and sensory changes [6, 20]. There are actually already promising results in hip and knee neuroarthropathy. Matsushashi et al. [16] reported satisfactory pain control and improved range of motion after an 8-year follow-up of 3 patients who underwent humeral head replacement and rotator cuff repair. Ueblacker et al. [21] also achieved a significant functional gain after bilateral reverse shoulder arthroplasty in a patient with severe neuroarthropathy. One of the requirements for reverse shoulder arthroplasty’s success is the glenoid’s preservation. Therefore, its indication should not be delayed until advanced stages of joint deterioration have been established. Once again, the literature is not yet solid enough to recommend a specific treatment. Nevertheless, it seems prudent to consider surgical treatment for severe bone destruction, instability and soft tissue damage, mainly to gain joint mobility and function after conservative treatment has failed.

We present the case of a patient with progressive limitation of shoulder mobility and function months later who started complaining of dysesthesia and swelling of the upper limb. She was then diagnosed with syringomyelia and had undergone surgery. When neuroarthropathy was detected, it was already at an advanced stage. After decompression surgery and several years of conservative treatment with physiotherapy and adjuvant medication, the patient improved slightly, still complaining of pain and significantly limited range of motion. Given the negative impact on daily life activities, the failure of conservative treatment and the satisfactory results that have emerged recently, one should consider proposing surgical treatment with reverse arthroplasty.

Despite the extensive differential diagnosis, symptoms and the cranium and cervical spine imaging could have allowed early...
detection of syringomyelia complicating Chiari malformation and its surgical treatment, delaying joint deterioration. We want to raise awareness of this rare disease and the importance of diagnosing it early.

Conclusion

Neuropathic arthropathy should be considered in patients suffering from pain, swelling, weakness and limited range of motion. Syringomyelia is the main cause of shoulder neuroarthropathy. In these cases, a thorough history, examination and imaging of the cranium and cervical spine enable an early diagnosis, which is crucial for therapeutic success. In addition, surgical decompression and joint protection measures should be performed, which is essential in delaying joint destruction and maintaining limb function.

Conflict of interest

There are no conflicts of interest to declare by any of the authors of this study.

References