



## Case report

## Septic arthritis complicating neuropathic shoulder due to cervical syringomyelia

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## ABSTRACT

One of the main causes of neuropathic osteoarthropathy of the shoulder is cervical syringomyelia. Chronic pain and swelling of the shoulder are the most frequent manifestations, but it occasionally can develop rapid osteoarticular destructive lesions (in less than six weeks), which raise the diagnostic possibility of septic arthritis and some tumours

We present the report of two men with septic arthritis of the shoulder associated with neuropathic arthropathy secondary to syringomyelia. Both patients presented with sudden shoulder pain exacerbated by either passive or active joint movements, malaise and fever. The first patient, a 39-year-old man, suffered left shoulder arthritis due to *Staphylococcus aureus*. The second patient, a 59-year-old man presented with right shoulder arthritis caused by to *Staphylococcus epidermidis*. The last microorganism also was isolated in three blood cultures. Infection should certainly be considered as a possible complication of the natural history of the neuropathic shoulder

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### Artritis séptica como complicación de la artropatía neuropática del hombro asociada a siringomielia cervical

## RESUMEN

Una de las principales causas de osteoartropatía neuropática del hombro es la siringomielia cervical. El dolor crónico y la tumefacción del hombro son las manifestaciones más frecuentes, pero ocasionalmente pueden desarrollarse lesiones destructivas osteoarticulares de rápida evolución (menos de 6 semanas), que plantean el diagnóstico diferencial con la artritis séptica y algunos tumores.

Describimos 2 varones con artritis séptica sobre artropatía neuropática del hombro secundaria a siringomielia. Ambos cursaron con dolor agudo exacerbado por los movimientos activos y pasivos, malestar general y fiebre. El primero, un hombre de 39 años, presentó una artritis del hombro izquierdo por *Staphylococcus aureus*. El segundo, un varón de 59 años, sufrió una infección del hombro derecho por *Staphylococcus epidermidis*, microorganismo que también se aisló en los 3 hemocultivos. El clínico debe considerar la infección como una posible complicación en el curso evolutivo de la artropatía neuropática del hombro.

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## Introduction

Syringomyelia is a chronic and progressive process with many causes, characterized anatomically by the development of a dissecting cavity in the gray matter that contains cerebrospinal fluid. Its main location is the cervical spinal cord and most often affects young adults.<sup>1,2</sup> There is a moderate correlation between the

characteristics of the Syringomyelic cavity (length, disposition and morphology) and its neurological expression, which emphasizes the loss of painful sensation, proprioception and heat.<sup>2</sup> Neuropathic arthropathy complicating the development of syringomyelia can be seen in one third of patients and the shoulder is the main location,<sup>3</sup> preceding other neurological manifestations. The neuropathic arthropathy of the shoulder raises a broad differential diagnosis, which may include neoplasms and some micro-crystalline and infectious arthritis.<sup>4-9</sup>

*Staphylococcus aureus* is the primary etiologic agent of septic arthritis in all locations,<sup>10-13</sup> including the shoulder, and its participation is highly variable as seen in different series, ranging between 3% and 21%.<sup>10-14</sup> However, the medical literature (Medline, Embase-plus until

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January 2009) only provide a single description of septic arthritis of a neuropathic joint.<sup>15</sup> Due to this exceptional infectious complication of a neuropathic arthropathy and the difficulties it poses for the differential diagnosis, we considered it of interest to provide two additional cases.

### Case report 1

Male, 39, sailor, with a history of left clavicle fracture in his adolescence, admitted due to left shoulder arthritis, fever and malaise, symptoms that had their onset 72 h after suffering superficial erosions of the external side of the ipsilateral arm. At 33, he had been diagnosed with a left shoulder neuropathic arthropathy associated with cervicodorsal syringomyelia, visible on magnetic resonance imaging (MRI) to the level of the third thoracic vertebra. At that time, ultrasonography showed thinning and irregularity of the cartilage of the humeral head and partial rupture (grade I) of the supraspinatus and subscapularis tendons.

Physical examination revealed an axillary temperature of 39.1 °C, swelling and effusion of the left shoulder with active and passive restriction of all movements, decreased pain and temperature sensation in the left upper limb and moderate hyperreflexia in the upper right extremity. The remainder of the examination showed no noteworthy data. Laboratory analysis revealed an elevated ESR (132 mm/1st h) and C-reactive protein (217 mg/l), normochromic normocytic anemia (Hb: 109 g/l), leukocytosis ( $12.7 \times 10^9/\text{mm}^3$ , 89% neutrophils) and thrombocytosis ( $435 \times 10^9/\text{l}$ ). The following parameters were normal or negative: glucose, urea, creatinine, cholesterol, lactate dehydrogenase, transaminases, creatine kinase, aldolase, sodium, potassium, calcium, phosphorus, alkaline phosphatase, proteinogram, TSH, T4, immunoglobulin titers, rheumatoid factor and antinuclear antibodies. X-rays showed left shoulder subacromial space narrowing, an irregular contour of the humeral head, diffuse osteopenia and increased volume and density of soft tissue. Moreover, ultrasonography showed an abundant effusion with loose bodies in the subacromial bursa and communication with the glenohumeral cavity, complete rupture of the supraspinatus, infraspinatus and subscapularis tendons, and biceps subluxation. MRI confirmed these findings and showed periarticular soft tissue edema of the humeral head and glenoid cavity (Figure 1). Scans with <sup>99m</sup>Tc and <sup>67</sup>Ga showed tracer uptake in the left shoulder. The neurophysiological (EMG, ENG and somatosensory evoked potentials) showed evidence of denervation and neurogenic injury in C6, C7 and D1 myotomes of moderate intensity and consistent with involvement of the sensory pathway.

On admission, a 13 cc a purulent effusion was obtained after arthrocentesis in which no microorganisms were visualized after a Gram stain. However, after 48 h in culture, growth was observed and colonies were identified as *Staphylococcus aureus* susceptible to vancomycin and cloxacillin, so antibiotic therapy was started with cloxacillin (2 g/IV/4 h)+gentamicin (80 mg/IV/24/h) and the patient was subjected to surgical drainage and debridement. Although the initial progression was favorable, within 2 weeks after surgery he developed a tract that drained the surgical wound and remained open until the eighth week, when parenteral antibiotic therapy was discontinued, maintaining oral cloxacillin until completing 12 weeks. Over the 2 years of follow-up, the patient presented no recurrence, but the anatomical and functional outcome of the shoulder was poor because of the complete tear of the rotator cuff and resorption of the humeral head.

### Case report 2

59 year old male with a history of a fracture of the right clavicle during childhood, chronic alcohol abuse and COPD, had been



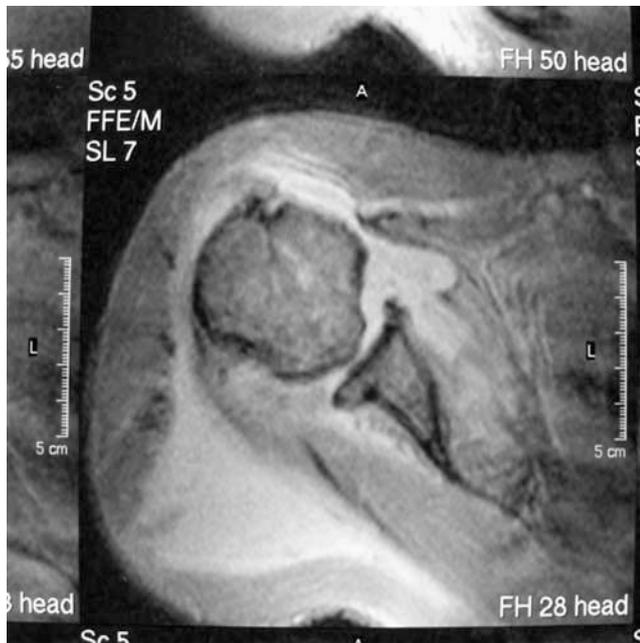
**Figure 1.** Coronal MRI of left shoulder, T2-weighted. There was an abundant effusion, loose bodies (arrow) and irregularity of the humeral head. Both this and periarticular tissues present strong Gadolinium contrast uptake.

diagnosed with right shoulder neuropathic arthropathy associated with syringomyelia at 54 (chronic right shoulder pain, reduced touch and thermal sensitivity associated with hyperreflexia of the upper extremities, consistent neurophysiological data and a cervical syringomyelic cavity on MRI). At the time, the right shoulder MRI showed complete rupture of the supraspinatus tendon, signs of the infraspinatus and subscapularis tendinitis and cortical edema and irregularity of the humeral head.

He was admitted because of right shoulder arthritis and fever. Pain and swelling in his right shoulder had begun 45 days earlier after an episode of respiratory decompensation in which he had suffered superficial phlebitis due to *Staphylococcus epidermidis* (isolated on the tip of the catheter).

Examination revealed an axillary temperature of 38.3 °C, swelling and effusion of the right shoulder with marked limitation of all active and passive movements, particularly abduction and external rotation, which did not reach 15° and 5° respectively, expiratory rhonchi and wheezing in both lungs, as well as discrete pitting edema in the right upper limb. Both the thermal and the proprioceptive sensitivity was greatly reduced in both upper limbs.

The most relevant laboratory findings were leukocytosis ( $14.2 \times 10^9$ ) neutrophilia (84%), macrocytosis (MCV of 99 fl), discrete thrombocytopenia ( $112 \times 10^9/\text{l}$ ), GPT-AST (78 UI/l, normal values < 45) and GGT (115UI/l, normal values < 65). *Staphylococcus epidermidis* was isolated both in 3 blood cultures and in the right shoulder joint effusion, with the Gram stain showed gram-positive cocci. The chest x-ray showed flattening of the domes of the diaphragm, aortic arch calcification and an increased bronchovascular infiltrate. Ultrasound visualized complete rupture and retraction of the subscapularis tendon and supraspinatus tendon subluxation of the long biceps tendon, abundant bursal effusion and heterogeneous echogenicity and irregularity of the humeral head. These findings were confirmed by MRI, which also revealed an increased uptake of gadolinium contrast in both the humeral head and glenoid cavity (Figure 2). There were no signs of endocarditis on echocardiography.



**Figure 2.** Axial MRI of the left shoulder that displays the irregularity of the humeral head and a generous effusion that stretched all of the recesses of the joint.

We proceeded to drain and debride the right shoulder and established vancomycin (1 g/IV/24 h) and rifampin (600 mg/day) for 6 weeks as treatment. The anatomical and functional result were unsatisfactory, and after treatment was complete, active abduction did not reach 20° and external rotation was only 10°.

## Discussion

About 12% of pyogenic arthritis are located in the shoulder,<sup>10-13</sup> a percentage that increases in older patients and in reached 21% in the case series by Drawer.<sup>14</sup> *Staphylococcus aureus* is the primary etiologic agent (> 75% cases).

Half of patients with shoulder septic arthritis have systemic risk factors such as diabetes mellitus, chronic renal failure, rheumatoid arthritis, liver cirrhosis, addiction to intravenous drugs and malignancy.<sup>10,12</sup> The organisms usually reach the joint through the blood, but in scapulohumeral septic arthritis, arthrocentesis with direct inoculation or infiltration is responsible for 24% to 80% of cases.<sup>10,13</sup> In the first of our patients can be considered, the eroded skin of the ipsilateral arm 72 hours before the joint symptoms was the considered point of entry. In the second patient, a superficial phlebitis associated with a catheter during a previous admission was the probable point of entry, identifying *Staphylococcus epidermidis*, a micro-organism that was recovered in the joint fluid and 3 blood cultures. The isolation of these bacteria in shoulder septic arthritis is rare.<sup>10</sup>

Although the presence of a previous joint injury promotes the development of pyogenic arthritis,<sup>16</sup> we have found only one case of mixed Neuropathic arthropathy and septic arthritis in the medical literature.<sup>15</sup> In that case it was a young man who suffered a *Staphylococcus aureus* arthritis on a Charcot shoulder. The two patients described were male and had also Neuropathic shoulder arthropathy associated with cervicodorsal syringomyelia. The time between diagnosis of the neuropathic arthritis and the infectious complication was 6 and 5 years, respectively.

The majority of cases of cervical syringomyelia have a prior injury of the cervical spine, arachnoid or spinal cord and, above all, are

associated with Arnold Chiari type I or II abnormalities, not seen in our 2 patients. The mechanism is controversial but involves both the loss of pain sensation and proprioception and autonomic nervous system disorders, with regional vasodilation and hyperemia, which promote bone resorption.<sup>17</sup>

The diagnosis of septic arthritis on a Charcot joint presents special difficulties, since clinical and radiological findings similar to those of shoulder septic arthritis can be seen clearly in pseudoseptic forms of neuropathic arthropathy<sup>4,6</sup> and in some microcrystalline arthritis, such as with "Milwaukee shoulder". In addition, fever has little predictive value because its specificity and sensitivity in pyogenic arthritis is less than 50%.<sup>18,19</sup> Tuberculous arthritis, some cancers<sup>5,6,8</sup> and hemophilia may also present with clinical and radiographic manifestations similar to those of neuropathic arthropathy of the shoulder, including swelling, pain, joint instability, glenohumeral subluxation, loss of function and serohematic effusion.

The progression of neuropathic arthropathy is usually slow and may be delayed by preventive measures, orthosis and physiotherapy and, as has been shown in some studies, by reducing bone resorption with bisphosphonates.<sup>20</sup>

In summary, the complicated course of neuropathic arthropathy with septic arthritis is rare, with only three cases having been reported. Diagnostic delay accelerates functional and anatomical articulation deterioration and worsens prognosis. The 2 patients described reflect this possibility as well as the need to quickly study, including joint fluid culture, any episodes of acute inflammation complicating chronic arthropathy.

## Disclosures

The authors have no disclosures to make.

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