

Bilateral neuropathic osteoarthropathy of the shoulder due to syringomyelia

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Abstract

Neuropathic osteoarthropathy, which is known as Charcot osteoarthropathy, is a degenerative arthritis that develops as a result of proprioceptive and sensory innervation loss. A 47-year-old man was admitted to the emergency department of the hospital with left shoulder pain, which was ongoing and exacerbating for 5 days. Examination of the cervical region takes a crucial part in determining shoulder pathology. Palliative therapy is the prior treatment of choice as surgical therapy has potential risks in Charcot osteoarthropathy.

Introduction

Neuropathic osteoarthropathy, which is known as Charcot osteoarthropathy, is a degenerative arthritis that develops as a result of proprioceptive and sensory innervation loss. The diseases involving in the etiology of this denervation are diabetes mellitus, syphilis, multiple sclerosis and syringomyelia.¹ Syringomyelia is the leading etiologic factor in shoulder Charcot osteoarthropathy.² In syringomyelia, central cavity or syrinx in the spinal cord causes spinal cord compression and nerve conduction abnormalities which resulted in progressive destruction in the shoulder joint.³ As the neuronal compression in syringomyelia is on the midline, the fibers conducting pain and temperature are affected first; white and grey matter are affected later. Bilateral shoulder Charcot osteoarthropathy is a rare entity.⁴⁻⁶ A 47-year-old man who had a bilateral shoulder Charcot osteoarthropathy due to syringomyelia is presented.

Case Report

A 47-year-old man was admitted to the emergency department of the hospital with left shoulder pain, which was ongoing and exacerbating for 5 days. There were over-

sensitivity, swelling and minimally elevation of skin temperature on palpation. Range of motion of the left shoulder was 50 degrees of forward flexion, 40 degrees of abduction and 40 degrees of internal rotation. Blood examination revealed the WBC count to be 5110/mm³, CRP 19.02 mg/L and body temperature 36.7°C. Radiographic examination of the left shoulder showed destruction of the joint (Figure 1) and there was dislocation of the joint with a Hill-Sachs lesion on the computerized tomography (Figure 2). Prior diagnosis was septic arthritis according to these findings. It was also recognized that the patient was diabetic, had similar symptoms on the opposite shoulder 6 years ago, had several surgeries on the right shoulder and had ongoing discharge from a sinus. This history alerted us and a MRI examination of the left shoulder was made which displayed osteoarthropathic changes and extensive fluid. 250 mL hematoma was depleted by puncture of the joint (Figure 3). Direct microbiological examination revealed no bacteria and the culture was negative. Thus the prior diagnosis of septic arthritis was cleared off. The elevation of CRP was attributed to discontinuous discharge from the opposite shoulder. The process was considered to be chronic as the temperature of the body and WBC count were normal. When the case history was detailed, it was recognized that he had neck pain and had a diagnosis of syringomyelia. He was out of routine control for several years. A MRI of the cervical vertebrae detected the syrinx (Figure 4). Consequently, a final diagnosis of Charcot osteoarthropathy of the left shoulder due to syringomyelia was accomplished. The patient was discharged and referred to Neurosurgery for the follow-up of the syrinx and Physical Therapy and Rehabilitation for the rehabilitation of the left shoulder.

Discussion

Examination of the cervical region has an important role in the diagnosis of the shoulder disorders. As the symptoms emanated from nerve entrapment or diseases of the spinal cord appear lately, one should examine the cervical region thoroughly and go for further investigation for the differential diagnosis of the shoulder complaints. 6% of the shoulder disorders has a cervical origin and this group of patients receives inappropriate medical therapy and the results are poor.^{7,8}

Syringomyelia is the leading cause of the Charcot osteoarthropathy of the shoulder, which is rarely seen as a shoulder dis-

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order. Syringomyelia starts as a localized cavitation (syrinx) in medulla spinalis and widens proximally and distally thereafter. Destructive process in the shoulder joint begins as a result of destruction of the nerve fibers conducting pain and sensation. In the pathophysiology of neuropathic osteoarthropathy, there are changes in vasomotor reply, which lead to an increase in blood flow resulting in osteopenia. Microtraumas accompanying these changes cause both impairment in joint structure and dislocations as well.⁹

Symptoms in Charcot osteoarthropathy generally rise spontaneously or as a result of soft tissue damage. Pain is not always present, but joint swelling exists mostly. The patient can not feel pain due to neuropathy. The first symptoms are loss in joint range of motion and abnormal voices coming from the joint as they were in our case.

Radiography has an important place in achieving a diagnosis in Charcot osteoarthropathy that detects joint surface and joint structure abnormalities. If radiography is normal, MRI and bone scintigraphy can give some information about the disease process. In the present case, there was shoulder joint subluxation, which accompanied severe joint surface destruction.

Diabetes, which is one of the leading causes of Charcot osteoarthropathy, gives rise to an arthropathy that mimics infectious processes by creating an autonomic neuropathy. Rise of the blood flow and temperature around the joint and loss of range of motion resulted in a diagnosis of arthritis,

which clinically resembles septic arthritis. Charcot osteoarthropathy, which is usually a chronic process, shows exacerbations periodically. Although there is a rise in acute phase reactants in these periods, there is no leukocytosis and high body temperature. These are the main parameters that differs Charcot osteoarthropathy from septic arthritis and MRI and joint puncture support these findings. Hatzis *et al.* made a first diagnosis of septic arthritis for the patients mentioned in their study, but they changed it as Charcot osteoarthropathy after further investigation.^{8,10} Atalar *et al.*, in a case series of 6 patients with Charcot osteoarthropathy, 5 of them have got a diagnosis of malignancy and one of them was diagnosed as septic arthritis at the beginning.¹¹ In a similar study, Chakraborty *et al.* proposed the diagnosis as a stress fracture due to osteomalasia at first but changed it as

Charcot osteoarthropathy after further investigation when the treatment failed.⁴ The presented case was evaluated as septic arthritis at the first examination. As shoulder Charcot osteoarthropathy is a rare disease, false diagnosis and unsuccessful treatment may precede. Septic arthritis, malignity, metabolic problems, trauma and gorham disease are all to be kept in mind in differential diagnosis.⁹

Surgical procedures have limited performance in the management of Charcot osteoarthropathy and if performed, these procedures have poor results and have a high risk of recurrence. Neither arthroplasty nor arthrodesis have good results in the surgical treatment of shoulder Charcot osteoarthropathy.¹¹ Joint debridement which is although one of the preferred surgical procedures and performed for the purpose of palliation is rarely done. The infection

rate is also high after this kind of procedures. Clare *et al.* reported the tendency of high infection rate for the patients who undergo shoulder arthrodesis.^{6,12}

The presented case had the same clinical presentation 6 years ago and had surgical intervention from the opposite shoulder in another institution. The surgical site was infected thereafter and the shoulder joint became severely stiff after repeated debridements. This complication from the past experience supports that there is a tendency toward infection in such cases. The presented case is interesting in that it has an inappropriate past management on one side and an appropriate current management on the other. Non-operative treatment methods are generally accepted in the management of Charcot osteoarthropathy in order to prevent the advancement of joint deformity. Treatment protocols to control pain and physical therapy to enhance range of motion and patient comfort are all performed. Non-steroid anti-inflammatory drugs are given to decrease synovial inflammation.^{3,13} Protection from trauma is an important modality in conservative treatment. Joint aspiration and bandaging prevent joint laxity and progressive deformity.¹⁴

Conclusions

Examination of the cervical region takes a crucial part in determining shoulder pathology. Neuropathic osteoarthropathy should be kept in mind in the differential diagnosis of the patients referring to the hospital with shoulder complaints and neurologic examination should be made carefully. Palliative therapy is the prior treatment of choice as surgical therapy has potential risks in Charcot osteoarthropathy.

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Figure 1. Destruction of the joint in shoulder radiography.



Figure 2. Hill-Sachs lesion on the computerized tomography.



Figure 3. Aspirated hematoma of the joint.



Figure 4. Magnetic resonance imaging of cervical spine showing syrinx in the spinal cord.

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