# The Shoulder Charcot Arthropathy and Secondary to Hemangioblastoma in Conus Medullari with Syringomyelia

Yu Ping Bai[1,2], Yang Yu[2,3], Wenbo Qi[2,3], Le Liu[2,3] and Hao Chen[2,4]

[1]*Department of MR, Lanzhou University Second Hospital, Lanzhou, China*
[2]*Department of Tumor Surgery, Lanzhou University Second Hospital, Lanzhou, China*
[3]*The Second Clinical Medical College of Lanzhou University, Lanzhou, China*
[4]*The Key Laboratory of the Digestive System Tumors, Lanzhou University Second Hospital, Gansu Province, Lanzhou, China*

## ABSTRACT

Charcot arthropathy of the shoulder joint is usually referred to as shoulder joint involvement of Charcot arthropathy, which is a chronic, degenerative, destructive condition resulting from decreasing or loss of sensorial innervation. To date, several reports have described the shoulder Charcot arthropathy, caused by malformations of the occipital region with syringomyelia; but nobody has reported the shoulder Charcot arthropathy secondary to hemangioblastoma in conus medullari with syringomyelia. Therefore, we report a 32-year male patient who was diagnosed with Charcot arthropathy of the shoulder joint, which was misdiagnosed as a soft tissue tumor and treated surgically. After the operation, the whole-spine MRI examination was performed as a presenting feature of hemangioblastoma in conus medullari with syringomyelia.

**Key Words:** *Arthropathy, Neurogenic, Hemangioblastoma, Syringomyelia.*

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## INTRODUCTION

Charcot arthropathy, also known as neuropathic arthropathy (NA), is a destructive joint disorder that occurs secondary to neurological disorder neuropathic arthropathy.[1] Shoulder joint involvement is found in approximately 6% of cases of syringomyelia, which is the main cause of shoulder Charcot arthropathy.[2] Hemangioblastoma is a rare, benign tumor that occurs mainly in the cerebellum, but may also be found in the spinal cord and brainstem.[3] To our knowledge, there is still no literature that reports the disease of Charcot arthropathy, caused by hemangioblastoma with syringomyelia.

Therefore, we report this case and provide a review of the literature.

## CASE REPORT

A 32-year male was hospitalised for diagnosis and treatment due to presenting right upper limb numbness and weakness three years ago without any significant incentives, then complaining the progressive swelling, pain and limitation of motion in his right shoulder joint due to trauma for one year.

*Correspondence to: Hao Chen, Department of Tumor Surgery, Lanzhou University Second Hospital, Lanzhou, China*
*E-mail: chenhaodrs@163.com*

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Physical examination: The right shoulder is pparently swollen, and its motion limited; flexion contractures deformities of joints. Both superficial and deep sensations in his right extremities were slightly impaired, and ankle and knee jerks were normal. Pathologic reflex such as the Babinski sign was absent.

All the laboratory examinations were within normal limits, on the basis of which syphilis and diabetes were ruled out. Anteroposterior plain radiograph of the right shoulder revealed the disappearance of the humeral head, the swollen soft tissue of the surrounding of the joint, and sporadic calcification near the joint (Figure 1). The same signs were present in the computed tomography (CT) scan of the vertebrate column and right shoulder joint (Figure 2). The shoulder magnetic resonance imaging (MRI) revealed a large amount of effusion, synovial hypertrophy, destruction of the humeral head, and a large irregular leaf-tissue masses formation (Figure 3). Single photon emission computerised tomography (SPECT) images demonstrated a large focus of obviously increased uptake in the right shoulder joint (Figure 4).

A series of imaging examinations suggested a preliminary diagnosis of a malignant mass in the shoulder joint. Accordingly, a tru-cut biopsy was performed, and analysed by pathological examination, in which local degeneration, necrosis, fibrosis, calcification, and inflammatory cell infiltration were observed, with no finding tumor cell. The suggestion from the multidisciplinary team (MDT) was that the patient needed to make the whole-spine MRI examination to rule out Charcot arthropathy.

The whole-spine MRI revealed nodule lesion in a conus medullaris presenting as hypo-isointense signal on TIWI and T2WI, intense homogeneous enhancement, with syringohydromyelia cavity extending from C1 to T12 vertebral levels (Figure 5). There was no family history of von Hippel-Lindau disease (VHL); and the brain MRI, abdominal CT scanning, and eye funduscopy showed negative findings. Complete tumor excision and decompression of the spinal cord with syrinx proceeded successfully. The histopathologic examination confirmed that the tumor was a hemangioblastoma.

Follow-up for 12 months showed no swelling or pain of the right shoulder and no need of medication for pain management during the treatment.

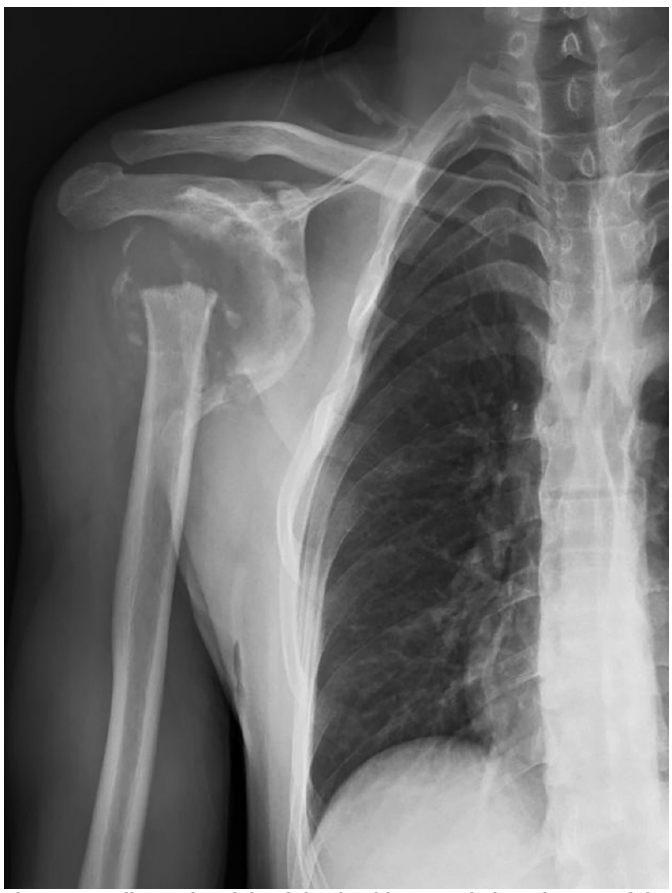

**Figure 1: Radiography of the right shoulder revealed an absence of the humeral head because of resorption, sporadic calcification near the joint.**

## DISCUSSION

Hemangioblastoma of the spinal cord occurs sporadically or is associated with VHL, accounting for about 3% of intramedullary tumors.[4] In view of the association of hemangioblastomas and VHL, MRI in the entire CNS, abdominal CT scanning, and eye funduscopy were performed before the operation to screen for other tumors. As a result, no other tumors were found except hemangioblastoma in conus medullaris. Approximately 60% of intramedullary hemangioblastomas are located in the cervical spine, followed by in the thoracic cord at nearly 40%, with the lumbar spine and conus medullaris the rare site.[5]

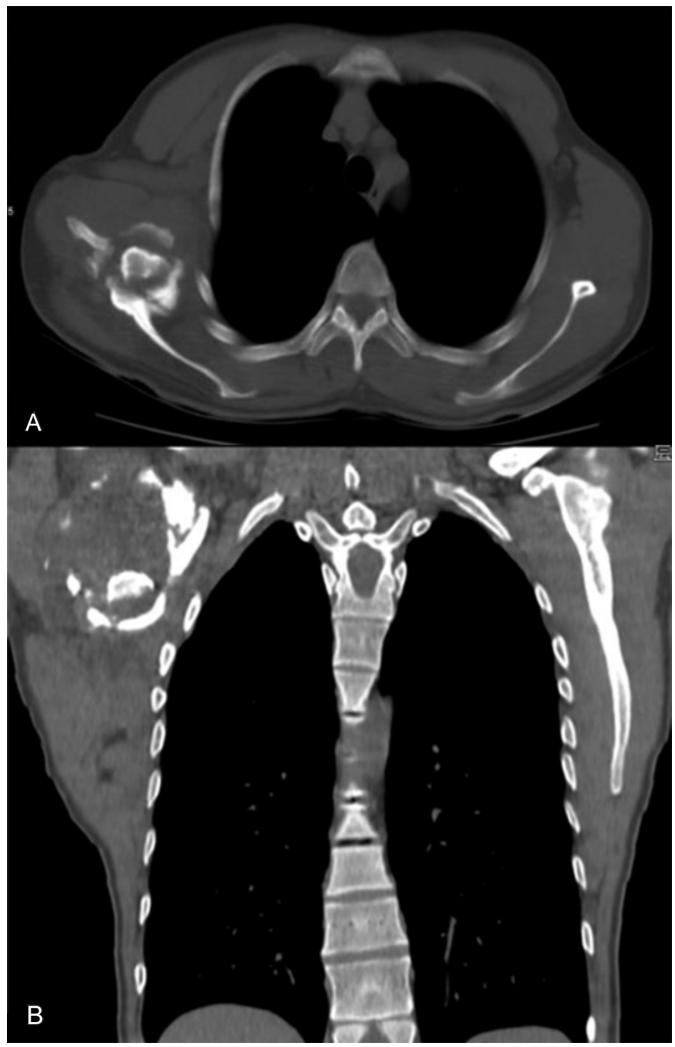


**Figure 2: Axial and coronal CT imaging of the vertebrate column and right shoulder joint (A, B) revealed a broken humeral head and sporadic calcification shadow of the surrounding of this joint.**

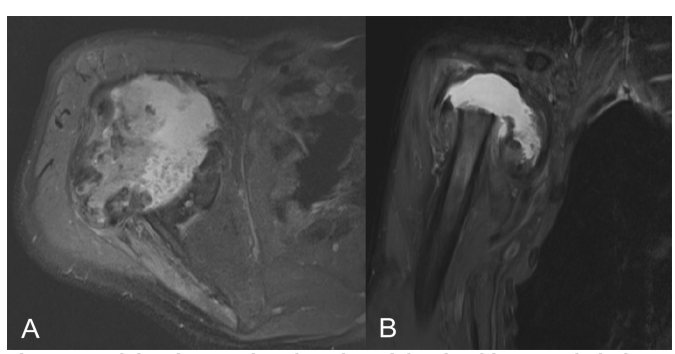


**Figure 3: Axial and coronal MR imaging of the shoulder revealed a large amount of effusion, synovial hypertrophy, destruction of the humeral head, and a large irregular leaf-tissue masses formation.**

There are some reports that about 50-70% of spinal hemangioblastomas have been associated with syringomyelia, while this rate increases to 100% in some reports. Syringomyelia is a chronic and slow progressive condition where a fluid-filled sac develops in the spinal cord. It can be congenital or may occur as a result of trauma, vascular problems, tumors, degeneration or infection.

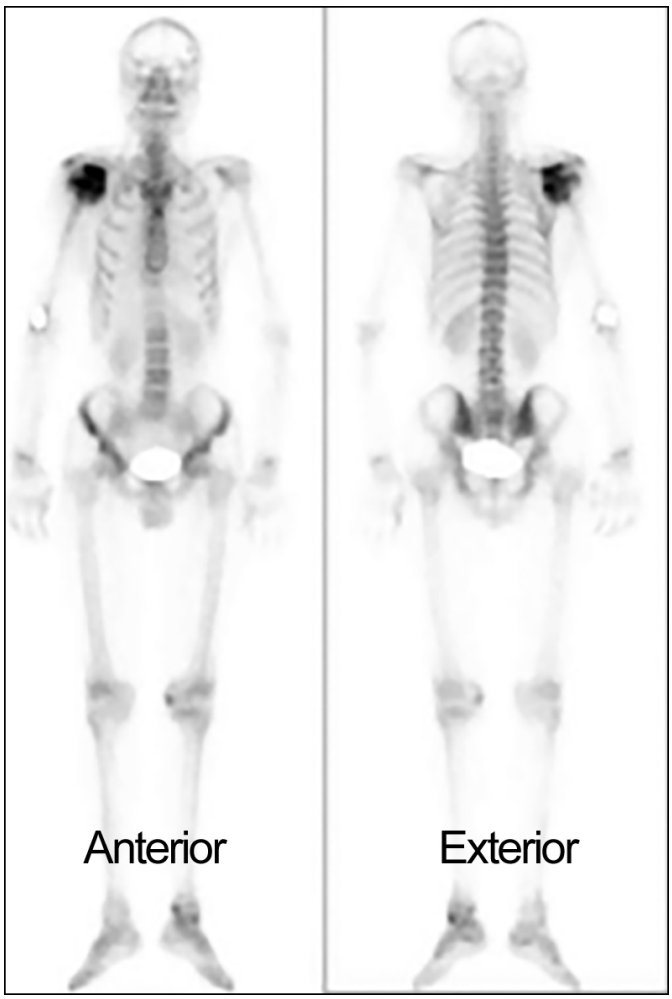


**Figure 4: SPECT images demonstrated a large focus of obviously increased uptake in the right shoulder joint.**

Several characteristic appearances of spinal hemangioblastoma on MRI have been reported. The first MRI feature is that small tumors (<10mm), mostly has low signal on T1WI and high signal on T2WI, while larger tumors usually demonstrate low signal, or equal and low signal, on T1WI, and of mixed-signal on T2WI. The second MRI feature is that flow void signals usually was shown in larger tumors. The third MRI feature is that the tumors arise in the superficial region of the spinal cord, which are most common in the posterior part of the spinal cord. The fourth is that the size of the syrinx formation is larger and the tumor is relatively smaller.[6] The pathogenesis of the large syrinx cavity relating to the small tumor nodule is still controversial and are as follows: (1) Disturbance of cerebrospinal fluid (CSF) flow by tumor occupying effect.[6] (2) Obstruction of cerebrospinal fluid circulation and malabsorption of cerebral spinal fluid CSF owing to transudation from the neoplastic nodule.[7]

NA is defined as a gradual destruction of the joint or joints, which is characterised by joint dislocations, pathologic fractures, and deformities, in patients most with sensory disturbance. NA usually occurs in load-bearing joints such as ankles, knees and hips, while the localisation of involved joint lesions being based on potential etiological factor.[8] NA is most commonly seen in patients with diabetic neuropathies, which affects the foot and ankle joints frequently. Secondly, shoulder involvement is common in syringomyelia. Charcot arthropathy secondary to syringomyelia mostly involved the shoulder and elbow.[8] The clinical manifestations of NA are varying, including painful or painless joints, joint instability, swelling, and dysfunction with or without trauma, of which joint swelling and limited activity were the commonest ones. Interestingly, the patient often visits initially to orthopedic clinics instead of neurology clinic, because joint symptoms occur earlier than neurologic symptoms. Therefore, it is essential for an orthopedic surgeon to be aware of the clinical characteristic of this rare disease.

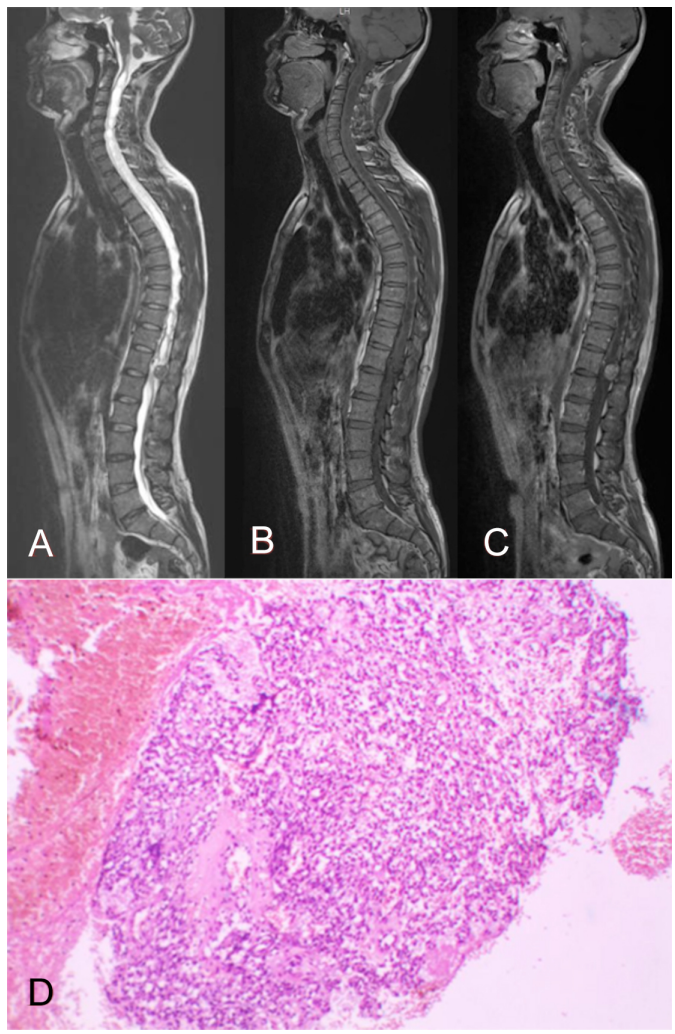


**Figure 5: Pre- and post-contrast MRI of the whole-spine (A, B, C) and pathologic specimens (D) of hemangioblastoma in the conus medullaris with syringohydromyelia. Sagittal images show a small nodular lesion of the conus medullaris with equal and high signal on T2WI (A), low signal on T1WI (B) and apparent enhancement on enhanced images (C), accompanying by high signal change of the spinal cord from C1 to T12 vertebral levels on T2WI (B). (D) H&E specimen (×100) from tumor biopsy shows syringohydromyelia.**

NA of the shoulder is usually characterised by destruction of the proximal humerus and the glenoid, swollen soft tissue of the surrounding of the joint, a large amount of effusion, with presenting as mimicking a soft tissue tumor in MRI and leading to misdiagnosis. Besides soft tissue tumor, differential diagnoses of NA should be made with septic arthritis, idiopathic osteolysis, nephropathy, synovial chondromatosis, Winchester syndrome, or Gorham disease.

It is possible that NA can be correctly diagnosed by careful history--taking, physical examination, and appropriate imaging examination. The therapeutic principles of NA include early identification and diagnosis, maintenance of joint and extremity function, and treatment of underlying disease process.[1] Our patient was eventually diagnosed as suffering from shoulder Charcot arthropathy, secondary to hemangioblastoma in conus medullari with whole-spinal cord syringomyelia. In order to eliminate the underlying disease, total removal of tumor and decompression of the spinal cord were performed. After surgery, the symptoms of the patient were improved and postoperative MRI revealed syrinx disappeared completely. As we have already seen, a recent report indicates that neurologic improvement with slowing of joint deterioration in patients after syrinx decompression.[9]

The treatment of shoulder lesion in NA is usually conservative, which is included restricted weight-bearing, immobilisation, passive stretching, physical therapy, range-of-motion exercises, strengthening and nonsteroidal anti-inflammatory medication.[1] The main goal of conservative treatment is to prevent the progression of ligamentous and soft-tissue laxity and reduce pain and swelling, thereby maintaining joint and extremity function. It is necessary to further research on conservative treatment due to the lack of its data. However, there have been some reports of unsatisfactory results for arthrodesis, resurfacing operations, and hemiarthroplasty because of a high risk of recurrence and infection.[10]

If a patient is suffering from swelling, pain, limitation of movement, and significant joint damage mimicking a soft tissue tumor in imaging, the differential diagnosis of NA of the joints should be considered and further examination such as the whole-spine MRI should be performed, in which early-stage of NA secondary to tumor with syringomyelia may be diagnosed easily, which is critical to manage patients successfully. To avoid misdiagnosis, it is therefore important to be familiar with the clinical manifestations and imaging features of this rare disease.

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**PATIENT'S CONSENT:**
Informed consent was obtained from the patient to use the descriptive information, radiological images and treatment process.

**CONFLICT OF INTEREST:**
The authors declared no conflict of interest.

**AUTHORS' CONTRIBUTION:**
YB, HC: Conceived the conception and designed the study.
YY, WQ, LL: Collected data and provided interpretations of the results.
YB: Searched the literature and drafted the manuscript.
All authors revised and approved the final version of the manuscript.

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