Neuropathic arthropathy caused by syringomyelia

Clinical article

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Object. Neuropathic arthropathy (Charcot joint) caused by syringomyelia is rare and commonly misdiagnosed. Few cases have been reported by neurosurgeons. The aims of this study were to analyze the clinical and imaging presentations of neuropathic arthropathy and to discuss the effect of surgical management of the primary neurological deficits on neuropathic arthropathy.

Methods. The authors retrospectively reviewed clinical and imaging data of 12 patients with neuropathic arthropathy caused by syringomyelia who were referred to the department of neurosurgery between January 2003 and September 2012. Radiographs revealed destruction, dislocation, disorganization, and increased density or debris in the joints. Magnetic resonance imaging showed a syrinx of the spinal cord in all patients, with Chiari malformation in 11 patients and tethered spinal cord in 1 patient. Neurosurgical operations were performed in 5 of 12 patients, including posterior fossa decompression in 4 patients and syrinx-subarachnoidal shunt placement in 1 patient. Surgical management of the neuropathic joints was not performed in any of the patients. All patients were followed up, with a mean duration of 39 months.

Results. Sixteen joints were involved, including 10 elbows, 3 shoulders, 2 interphalangeal joints, and 1 wrist. The side of the syrinx on cervical axial MRI was consistent with the side of the affected limb in every patient. Five patients who underwent neurosurgical treatments stated improvement in neurological dysfunctions and no deterioration in symptoms related to neuropathic arthropathy. In the 7 patients without neurosurgical treatments, 5 reported aggravation of neuropathic arthropathy manifestations, with deterioration of neurological symptoms in 4 of the 5 patients. The condition of the other 2 patients remained stable.

Conclusions. The elbow is the most frequently involved joint in neuropathic arthropathy caused by syringomyelia, followed by the shoulder. The authors speculate that the side of the syrinx determines the side of the neuropathic arthropathy. A detailed medical history and a careful physical examination are crucial for differentiating neuropathic arthropathy from other joint lesions. This study suggests that early management of the primary neurological condition may play an important role in preventing the development of neuropathic arthropathy and avoiding disease progression.

KEY WORDS • neuropathic arthropathy • Charcot joint • syringomyelia • Chiari malformation

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EUROPATHIC arthropathy, also known as Charcot joint, is a chronic form of degenerative arthropathy with destructive and productive (hypertrophic joints, such as sclerosis and debris) articular abnormalities, caused by an underlying neurological disorder. Painless joint swelling and limited joint mobility are the main manifestations of this affliction. Typical imaging features include joint destruction, disorganization, and effusion with osseous debris. Patients with diabetes mellitus, syringomyelia, tabes dorsalis, and other neuropathies are particularly prone to developing this joint disease. The joints most frequently involved are the hips and knees in tabes dorsalis, the ankles and feet in diabetes mellitus, and the shoulders and elbows in syringomyelia.

For patients, manifestations of neurological deficits are often occult and frequently overshadowed by symptoms related to neuropathic arthropathy. Thus, the orthopedic surgeon is often the first physician to evaluate patients with this condition. Improved outcome depends on early diagnosis and proper management of both the affected joints and syringomyelia. However, the under-
ing neurological disorders are easily overlooked, and thus neuropathic arthropathy is easily misdiagnosed; therefore, it is very important to determine the primary neurological disease and differentiate neuropathic arthropathy from other joint lesions.

In our experience, most cases of neuropathic arthropathy were reported by orthopedic surgeons, and manifestations around the affected joints were considered as the chief complaints. Few cases were reported by neurosurgeons. In addition to joint lesions, we pay more attention to neurological deficits in our department. In this article, we report on 12 patients who were diagnosed with neuropathic arthropathy secondary to syringomyelia, in which 5 patients were treated with neurosurgical operations. Clinical presentations, imaging findings, and outcomes are reviewed and discussed.

Methods

After the study was approved by the Institutional Review Board of Beijing Tiantan Hospital, Capital Medical University, we retrospectively reviewed the medical records, radiographs, and MR images of 12 patients with neuropathic arthropathy secondary to syringomyelia who were referred to our department from January 2003 through September 2012. A detailed medical history was recorded, with emphasis on neurological dysfunctions, including paresthesias, dysesthesias, pain and temperature sensory deficits, muscle atrophy, weakness of limbs, and others. A history of joint lesions was also investigated, focusing on the presence or absence of pain, swelling, stiffness, motor limitations, and traumatic events. Radiographs of the involved joints and MR images of spinal cords of all 12 patients were reviewed and discussed.

Syringomyelia was mainly located in the center of the spinal cord, but we found it to be always slightly asymmetrical. The side of the syrinx on cervical axial MRI was noted in this study. The primary neurological conditions were treated surgically in 5 of the 12 patients (Table 1). Posterior fossa decompression was performed in Cases 1, 5, 8, and 10, using procedures including bone decompression (2.5–3 cm diameter), and duraplasty with autologous graft. Syrinx-subarachnoidal shunt placement was performed for Case 9. Surgical management of the neuropathic joints was not conducted in any of the patients. All patients were followed up, with a mean duration of 39 months.

Moreover, 169 patients without preoperative joint lesions who were diagnosed with CM and syringomyelia and who underwent posterior fossa decompression in our department from January 2008 to December 2009 were followed up, focusing on manifestations of joint lesions.

Results

Clinical data of the 12 patients are summarized in Table 1. There were 4 male patients and 8 female patients in this series. The average age was 45.8 years (range 13–76 years). Sixteen joints were involved, including 10 elbows, 3 shoulders, 2 interphalangeal joints, and 1 wrist.

All of the patients were right-handed. Right limbs were involved in 3 patients, and left limbs were involved in 9 patients.

The onset of symptoms related to the involved joint was acute in 1 case (Case 7), and chronic in the other cases. Only 1 patient was able to recall a traumatic injury (Case 1). All of the affected joints were markedly swollen. Only 2 patients had mild pain and the others were pain free.

All 12 patients had neurological symptoms. Sensory changes occurred in 11 patients and motor weakness occurred in all 12 patients. Neurological dysfunctions occurred earlier than joint lesions in all patients. The average duration between the onset of neurological dysfunctions and joint lesions was 9 years. Asymmetrical reflexes were noted in all patients, with 3 patients having absent reflexes and 9 patients having hypoactive reflexes in the involved limbs. Lower extremities were involved in 1 case (Case 9), and the main manifestation was motor weakness.

Radiographs of the involved joints showed destruction, dislocation, disorganization, and increased density or debris. Atrophic changes occurred in 8 patients and hypertrophic changes in 4 patients. Magnetic resonance images of the spinal cord demonstrated syringomyelia in all cases, with CM in 11 patients and tethered spinal cord in 1 patient. In the 11 patients with CM, the length of tonsillar herniation ranged from 5 to 16 mm (mean 9.1 ± 3.3 mm), and a cervicothoracic syrinx was found in all of these patients. No other craniovertebral anomalies were detected, such as basilar invagination, Klippel-Feil syndrome, instability of the craniovertebral junction, and others. A holocord syrinx was found in the patient with a tethered spinal cord (Case 9). The side of syrinx was noted, and it was consistent with the side of the affected limbs in all patients.

Five patients underwent neurological operations, and the average follow-up duration was 30 months in these 5 patients. All of these patients stated improvement in neurological dysfunction and no deterioration of the symptoms related to the affected joints. Postoperative MR images showed reduction of the syrinx in all 5 patients. Seven patients who did not undergo surgical treatment were also followed up, with an average duration of 47 months. Five patients complained that symptoms related to joint lesions were aggravated, and 4 of the 5 patients stated they experienced deterioration of neurological symptoms. No obvious change of symptoms was noted in the other 2 patients.

In addition to these 12 patients, 169 patients without complaints of joint lesions preoperatively were diagnosed with CM and syringomyelia, underwent posterior fossa decompression, and were followed up for an average duration of 42 months. None of these 169 patients complained of manifestations of arthropathy, and 143 patients stated obvious postoperative improvement of neurological dysfunctions.

Illustrative Cases

Case 1

This 44-year-old, right-handed woman was admitted to our department with limited movement and painless
swelling of elbow and interphalangeal joints of the middle finger on her upper right side for 6 years. She was referred to several orthopedic departments at different hospitals but no definite diagnosis was given. The patient had a history of weakness and loss of pain and temperature sensations in the right upper limb for 16 years, with no history of diabetes mellitus, sexually transmitted disease, or traumatic events. The patient was treated with massage and manipulation of the elbow but no improvement was observed.

On examination, the right elbow joint was markedly painless and swollen, and there was a 30° limitation in active extension (Fig. 1A). The internal and external rotation of her elbow was also limited. In addition, there was a flexion deformity and a motor limitation of the interphalangeal joints of the right middle finger (Fig. 1B arrow). The left upper limb and lower limbs were not involved.

On neurological examination, there was a loss of superficial touch, pain, and thermal sense on the right half of her face, extending to the T-8 dermatome on the right half of her trunk and on the right upper extremity. Muscle power was Medical Research Council Grade 4 in the right upper limb and Grade 5 in other limbs. Deep tendon reflexes were hypoactive in her right upper extremity but normal on the other side.

Radiographs of the elbow revealed a loss of normal architecture, with bone fragments lying in joint space and heterotopic ossification in the adjacent tissues. No bone resorption or dislocation was observed (Fig. 2A). A radiograph of the right hand showed deformity of the right middle phalange with destruction in joint spaces and sclerosis of articular surfaces of the proximal and distal interphalangeal joints (Fig. 2B arrows). Magnetic resonance imaging of the spinal cord revealed a right-sided syrinx extending from C-2 to the thoracic region and a Type I CM with a 7-mm descent of the cerebellar tonsil below the foramen magnum (Fig. 3A and B). The motor evoked potentials test revealed a nerve conduction disorder of the right upper limb, while the somatosensory evoked potentials test result was normal. Other examinations and laboratory test results were within normal ranges.

Posterior fossa decompression was performed on this patient in May 2012. Postoperative MR images showed reduction of syrinx volume (Fig. 3C). The patient stated improvement in pain and temperature sensations, and no obvious change in the symptoms of the elbow and the interphalangeal joints was observed.

Case 2

This 48-year-old, right-handed woman was admitted to our department with painless swelling of the shoulder, elbow, and wrist on the upper left side for 8 years, and limited movement of these joints for 2 years. The patient had a history of weakness and loss of pain and temperature sensations in the left upper limb for 15 years. No traumatic event to her left arm could be recalled.

On examination her left shoulder, elbow, and wrist were markedly swollen, but there was no tenderness or pain with motion. Active forward elevation of the shoulders was to 60° on the left and to 160° on the right. Active external rotation was to 30° on the left and to 50° on the right. The internal rotation of the left shoulder was also limited. The active elbow range of motion was limited, from 20° of extension to 90° of flexion. The right upper limb and lower limbs were not involved.

On neurological examination there was a loss of superficial touch, pain, and thermal sense extending from C-3 to the T-8 dermatome on the left half of her trunk and on the left upper extremity. According to the Medical Research Council scale, muscle strength was 4/5 in the left upper limb and 5/5 in other limbs. Deep tendon reflexes were hypoactive in her left upper extremity but normal in

### TABLE 1: Summary of clinical data in 12 patients with neuropathic arthropathy*

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (yrs), Sex</th>
<th>Etiology</th>
<th>Joints Involved</th>
<th>Side of Syrinx</th>
<th>Traumatic Events</th>
<th>Pain</th>
<th>Sensory Changes</th>
<th>Main Radiographic Change</th>
<th>Surgical Treatment</th>
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<tbody>
<tr>
<td>1</td>
<td>54, F</td>
<td>CM</td>
<td>elbow (rt), interphalangeal joints (middle finger, rt)</td>
<td>rt</td>
<td>no</td>
<td>no</td>
<td>yes</td>
<td>hypertrophic</td>
<td>PFD</td>
</tr>
<tr>
<td>2</td>
<td>48, F</td>
<td>CM</td>
<td>shoulder, elbow, wrist (lt)</td>
<td>lt</td>
<td>no</td>
<td>no</td>
<td>yes</td>
<td>atrophic</td>
<td>none</td>
</tr>
<tr>
<td>3</td>
<td>57, F</td>
<td>CM</td>
<td>shoulder (rt)</td>
<td>rt</td>
<td>no</td>
<td>no</td>
<td>yes</td>
<td>atrophic</td>
<td>none</td>
</tr>
<tr>
<td>4</td>
<td>53, F</td>
<td>CM</td>
<td>elbow (rt)</td>
<td>rt</td>
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<td>yes</td>
<td>hypertrophic</td>
<td>none</td>
</tr>
<tr>
<td>5</td>
<td>52, F</td>
<td>CM</td>
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<td>yes</td>
<td>hypertrophic</td>
<td>PFD</td>
</tr>
<tr>
<td>6</td>
<td>76, F</td>
<td>CM</td>
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<td>yes</td>
<td>atrophic</td>
<td>none</td>
</tr>
<tr>
<td>7</td>
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<td>lt</td>
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<td>atrophic</td>
<td>none</td>
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<tr>
<td>8</td>
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<td>no</td>
<td>yes</td>
<td>atrophic</td>
<td>PFD</td>
</tr>
<tr>
<td>9</td>
<td>13, M</td>
<td>TSC</td>
<td>elbow (lt)</td>
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<td>atrophic</td>
<td>syrinx-subarachnoidal shunt placement</td>
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<td>10</td>
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<td>yes</td>
<td>hypertrophic</td>
<td>PFD</td>
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<td>11</td>
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<tr>
<td>12</td>
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<td>CM</td>
<td>elbow (lt)</td>
<td>lt</td>
<td>no</td>
<td>no</td>
<td>yes</td>
<td>atrophic</td>
<td>none</td>
</tr>
</tbody>
</table>

* All patients were right-handed, and all suffered from swollen joints and motor weakness. Abbreviations: PFD = posterior fossa decompression, TSC = tethered spinal cord.
other limbs. Other examination and laboratory test results were within normal ranges.

Radiographs and 3D CT images of the left shoulder revealed destruction, dislocation, periarticular ossification, effusion, and partial resorption of the humeral head and glenoid fossa (Figs. 4A and 5). Radiographs of the left elbow showed destruction of joint spaces and sclerosis of articular surfaces (Fig. 4B). Sclerosis, debris, destruction of joint spaces, and partial absorption of the distal radius were detected on the anteroposterior radiograph of her left wrist (Fig. 4C). Cervical MRI revealed a left-sided spinal syrinx extending from C-1 to the thoracic region and a Type I CM with a 9-mm descent of the cerebellar tonsil below the foramen magnum (Fig. 6).

The patient was admitted to our department 2 years ago, and the diagnosis of neuropathic arthropathy secondary to syringomyelia with CM was considered. However, no surgical treatment was performed on this patient because of hypertension and arrhythmia. The patient was followed up for 2 years, and she complained her condition was getting worse. She had voice hoarseness and trouble walking in those 2 years. In addition, the motion of her left upper limb was further limited and swelling of the involved joints was aggravated.

**Discussion**

Neuropathic arthropathy secondary to syringomyelia is a rare condition, and only a few cases have been reported in the literature. From January 2003 to September 2012, 715 patients with CM and syringomyelia were admitted to our department, but only 11 patients (1.54%) had neuropathic arthropathy. During the same period, 1832 patients were diagnosed with arthropathy in our hospital, so neuropathic arthropathy accounted for approximately 0.66% of patients with arthropathy. Our study confirmed this disorder was most commonly related to CM, including 11 of 12 cases in our series. Neuropathic arthropathy can also result from other spinal cord lesions, such as tethered spinal cord.

Some authors report that in syringomyelia, neuropathic changes are more commonly noted in the shoulder, followed by the elbow and wrist. In our study, the elbow was the most frequent location: of the 16 affected joints, 10 were elbows, 3 were shoulders, 2 were interphalangeal joints, and 1 was the wrist.

Bone resorption of Charcot joints, especially in the shoulders, has been emphasized by many authors. In our series, atrophic changes were the main radiographic manifestations in 10 joints, including all 3 shoulders, 6 elbows, and 1 wrist. However, the other 6 joints revealed hypertrophic changes. These data demonstrate that a Charcot joint does not necessarily characterize bone resorption, especially in elbows, wrists, and hands.

The pathogenesis of neuropathic arthropathy remains a controversial issue. There are 2 main theories: the neurotraumatic and neurovascular theories. The neurotraumatic theory suggests that somatic muscular reflexes that normally protect joints from exceeding certain safe limits in range of movement are lost because of absent or decreased sensations, leading to repeated subclinical trauma and ultimately joint destruction. The neurovascular theory states that a Charcot joint develops when the sensory deficits disrupt the normal neurovascular reflex, resulting in persistent hyperemia and active bone resorption by osteoclasts. It is believed that both of these theories play an important role. The neurovascular theory plays an initial role and the neurotraumatic theory plays an auxiliary or supporting role on the basis of the neurovascular theory. However, the pathogenesis of this condition is not fully understood and there are still some questions; for example, why does neuropathic arthropathy usually occur monolaterally?

According to the neurotraumatic theory, weight-bearing joints and frequently used joints are more prone to be affected. The more the joints are used, the more traumas the joints suffer. This explains why most affected joints are located in the lower extremities. However, in this series, all 12 patients were right-handed, but only 3 had involvement of their right limbs, which contradicts the neurotraumatic theory. To explain this phenomenon, we paid attention to the primary neurological disorder. Although syringomyelia was mainly located in the center of the spinal cord, we always found it to be slightly asymmetrical. In our study, the side of the syrinx on cervical axial MRI was noted (Figs. 3B and 6B), and it
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was consistent with the side of the affected limb in every patient. In addition, the literature on neuropathic arthropathy secondary to syringomyelia was reviewed, and 4 cases with axial MR images were found. In these 4 cases, the side of the syrinx was also identical to the side of the Charcot joints in these 4 patients. Therefore, we speculate that the side of the syrinx determines the side of neuropathic joints. In other words, the primary neurological condition, rather than traumas, plays a more important role in determining the development of neuropathic arthropathy. This also explains why neuropathic arthropathy usually occurs monolaterally. In most cases, syringomyelia is asymmetrical, where the nerve pathway is more easily affected, and then neuropathic arthropathy develops on the same side. This explains why neurological presentations are asymmetrical in most patients with syringomyelia.

Some authors report that symptoms related to neuropathic joints manifest earlier than neurological symptoms in most cases. However, in our study, although the primary complaint of most patients was symptoms of neuropathic arthropathy, a history of neurological dysfunctions could be recalled in every patient when asked in detail. In our experience, the neurological symptoms are usually occult and easily overlooked. A detailed neurological history should be taken and neurological examinations should be performed on patients with painless joint destructions. In addition, pain and temperature sensory deficits are not limited to just those in the involved limb, but also include the ipsilateral trunk in most cases. In this series, sensory changes occurred in 11 of 12 patients, and all 11 had ipsilateral trunk manifestations. Thus, sensory changes are a very important clue in determining the underlying neurological disorder.

![Fig. 3. Case 1. Cervical T2-weighted MR images obtained in the patient. A: Preoperative sagittal image revealing a syrinx extending from C-2 to the thoracic region, with a Type I CM. B: Preoperative axial image demonstrating the syrinx was right-sided. C: Postoperative sagittal image showing a reduction in syrinx volume.](image1)

![Fig. 4. Case 2. Plain anteroposterior radiographs obtained in the patient. A: Image of the left shoulder showing destruction, dislocation, periarticular ossification, and partial resorption of the humeral head and glenoid fossa. B: Image of both elbows revealing sclerosis and destruction of the left elbow. C: Image of both hands showing sclerosis, debris, destruction of joint spaces of the left wrist, and partial resorption of the distal radius.](image2)
In our experience, early management of the primary neurological disorder is very important to avoid disease progression. Five patients who underwent neurosurgical treatments showed improvement in neurological symptoms and no deterioration in presentations related to neuropathic arthropathy. In the 7 patients without neurosurgical treatment, 5 complained of aggravation of neuropathic arthropathy symptoms, with deterioration of neurological symptoms in 4 of the 5 patients. This further proves the significance of early diagnosis of neuropathic arthropathy and detecting the underlying neurological disorder. Moreover, in the 169 patients without neuropathic arthropathy symptoms preoperatively who were diagnosed with CM and syringomyelia, and who underwent posterior fossa decompression, none complained of manifestations of arthropathy during postoperative follow-up. This result suggests that early management of the neurological deficits may prevent the development of neuropathic arthropathy.

The present study has several limitations. First, it is a retrospective case series with limitations inherent to the study design. Second, because of the rarity of neuropathic arthropathy secondary to syringomyelia and the lack of case controls, no statistical analysis was performed, and no definitive statement regarding management can be made from this Level IV study. Third, although we noted that the Charcot joints occurred ipsilateral to the side of syrinx, the mechanism is still unclear, which needs further study.

Conclusions

Neuropathic joints caused by syringomyelia are more commonly observed in the upper extremities, and the elbow is the most common site of involvement. Syringomyelia tends to be asymmetrical in most cases, which determines the side of the Charcot joints. Neurological dysfunctions always occur before joint lesions but are often overlooked. We believe a detailed medical history and a careful physical examination are important for correctly diagnosing neuropathic arthropathy. In our experience, early management of the primary neurological condition may play an important role in preventing the development of neuropathic arthropathy and avoiding disease progression.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: Deng, Wu. Acquisition of data: Deng, Yang. Analysis and interpretation of data: all authors. Drafting the article: Xu, Deng. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Xu. Statistical analysis: Deng. Administrative/technical/material support: Yang. Study supervision: Xu.

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