CASE REPORT

Neuropathic arthropathy and syringomyelia complicating a case of ankylosing spondylitis with peripheral arthritis

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BACKGROUND

Ankylosing spondylitis (AS) can present with various spinal complications due to ligamentous calcifications, fractures and extradural compression. However, syringomyelia is a rare association in patients of AS. It is also unusual for a patient to present with two forms of arthropathies due to different aetiologies at one point of time.

CASE PRESENTATION

A 30-year-old man presented with a 6-year history of inflammatory back pain, enthesitis and with a history of peripheral arthritis since 1 year. A diagnosis of AS was carried out with the presence of inflammatory back pain, limited lumbar flexion and bilateral grade 3 sacroiliitis on pelvic radiography (not shown). He had a history of being treated with steroids and sulfasalazine with partial relief. On enquiry for his predominant hand wasting, he revealed having tingling sensation in the left-upper limb followed by right-upper limb since 2 years. He also noticed thinning of hand muscles with weakness in hand grip (left>right) since last 6 months.

On examination his modified Schobers's test and chest expansion test were limited to 3.5 and 4.2 cm, respectively. The peripheral swollen joint count was 12. He also had mild-left shoulder pain. Further neurological evaluation revealed wasting of the periscapular muscles, deltoid, biceps, triceps and brachioradialis. Intermittent fasciculations were present in upper limbs. He had bilateral hand muscle wasting with hand grip weakness disproportionate to the degree of hand joint involvement ([figure 1](#)). Bilateral biceps and triceps reflexes were absent. There was dissociated, suspended anaesthesia, with preserved posterior column sensations in bibrachial distribution on the lateral aspects of the arms and forearms. There was no lower limb or bladder bowel involvement with flexor plantar responses.

INVESTIGATIONS

The investigations revealed raised erythrocyte sedimentation rate (100 mm in 1 h), increased C reactive protein (12 mg/L) with normal haematological parameters including blood sugar, renal and liver function tests. His Bath Ankylosing Spondylitis Disease Activity Index and AS disease activity score-C reactive protein (ASDAS-CRP) were 8.6 and 4.5, respectively, suggesting high disease activity. X-ray of the left shoulder revealed severely deformed humerus head along with its articular surface with preserved glenoid articular margins

Figure 1 Wasting of bilateral hand muscles.

([figure 2](#)). Minimal amount of fluid was aspirated from the left shoulder joint, fluid was non-inflammatory, crystals were absent and evaluation for infections including tuberculosis was negative. The radiological findings of left shoulder with minimal pain and associated neurological findings suggested a diagnosis of neuropathic arthropathy/Charcot's joint. MRI of cervical spine revealed a syrinx extending from C1 to D1 level with no associated Arnold-Chiari malformation ([figure 3](#)).

DIFFERENTIAL DIAGNOSIS

Neurological complications in AS have been well described. They have been attributed to

Figure 2 Plain radiograph showing neuropathic left-shoulder joint with severely deformed humerus head and its articular surface (arrows).

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Figure 3 MRI with T2-weighted images showing syrinx extending from C1 (cervical) vertebra to D1 (thoracic) vertebra (arrows).

ligamentum flavum calcification, vertebra-osseous deformities, osteoporotic vertebral fractures and increased kyphosis of the spine. There are various reports of extradural compressive myelopathy and radiculopathy.¹ However, AS and syringomyelia is a rare association.

Syringomyelia has been associated with Arnold-Chiari malformations, intramedullary tumours, Klippel feil or other spinal abnormalities, cervical spondylosis, trauma, arachnoiditis and even rheumatoid arthritis.²

There was no history of fever, constitutional symptoms or tuberculosis in our patient. MRI excluded possibilities of intramedullary tumour or any congenital abnormalities. Plain radiograph of cervical spine (not shown) and MRI in our patient did not show significant spinal ligament calcification or subarachnoid indentations so as to postulate cerebrospinal fluid (CSF) obstruction as a possible cause of syrinx.

OUTCOME AND FOLLOW-UP

He was given non-steroidal inflammatory drugs, local intra-articular steroids and sulfasalazine for his AS. He was referred to neurosurgery for surgical intervention of syringomyelia, but he refused for the same.

DISCUSSION

AS and syringomyelia is a rare association. To the best of our knowledge this is only the second case so far described.

The authors of the previously described case hypothesised that obstruction to the flow of CSF in subarachnoid space caused by spinal ligament calcifications and facet joint arthropathy could have led to syringomyelia in their patient.³ Their patient had developed spasticity and weakness in lower limbs along with bladder bowel symptoms before the diagnosis was considered.³ Our case was difficult to diagnose as the patient had predominantly articular symptoms and his hand wasting was previously ascribed to disuse atrophy due to chronic hand joint arthritis. There was no obvious evidence of CSF obstruction and the exact cause of syringomyelia remained unclear in our patient.

Neuropathic arthropathy, especially shoulder joint arthropathy is a well-described phenomenon with syringomyelia.^{4 5} In fact, arthropathy associated with syringomyelia can be misdiagnosed as rheumatoid polyarthritis, polyosteoarthritis and even septic arthritis.^{4 6} The diagnosis can become more challenging if the patient has pre-existing arthritis as in our patient. Our patient failed to give his history of upper-limb sensory symptoms and hand grip weakness initially. The patient had minimal left shoulder pain in contrast to the severity of radiographic picture. The possibility of neuropathic arthropathy of left shoulder along with predominant hand wasting prompted us to revisit patient's history, examination and finally reach a correct diagnosis.

Learning points

- ▶ Ankylosing spondylitis can present with various complications involving the spine.
- ▶ Neuropathic arthropathy should always be suspected in a destructed and relatively painless joint.
- ▶ Syringomyelia is the most common cause of neuropathic arthropathy of upper-limb joints.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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