Neuropathic arthropathy of the shoulder joint secondary to a syringomyelia

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DESCRIPTION

A 29-year-old male brick-layer presented with a 5-month history of atraumatic right anterolateral shoulder pain and swelling.

Pain was present at night and rest but did not prevent him from working. He had no significant medical history. He was a smoker and non-diabetic.

On examination, his right shoulder was swollen with minimal irritability but a limited abduction range. His neurological examination showed normal tone, power, proprioception, sensation and reflexes of the upper and lower limbs.

X-ray showed an aggressive, lytic appearance of the proximal humerus and glenoid (figure 1). MRI confirmed a destructive lesion involving both sides of the joint with a large, thick walled fluid filled cavity (figure 2).

A nuclear medicine scan was performed to further assess whether the lytic lesion was isolated or widespread. Localised high activity suggested an isolated right shoulder arthropathy.

Blood test investigations demonstrated C reactive protein <2 (normal <5) and a white-cell count of 8.5 (normal 4–10). A syphilis screen, B12, folate, HBA1c and serum electrophoresis were normal.

A fine needle aspirate of joint fluid demonstrated no malignant tumour cells, no crystals and a negative culture for organisms.

Further MRI of the whole spine revealed an extensive syringomyelia extending from C2-T10 (figure 3), with a basilar herniation of the cerebellum consistent with a type 1 Chiari malformation.

Neuropathic (Charcot) arthropathy is a rare degenerative process secondary to a loss of innervation to a joint, causes of which include diabetes, Chiari malformation, end stage renal disease, tabes dorsalis, leprosy, traumatic spinal cord injury and multiple sclerosis. Shoulder neuropathic arthropathy has been noted in approximately 6% of cases of syringomyelia.[1] A syringomyelia can involve the decussating fibres of the lateral spinothalamic tract (pain and temperature fibres) resulting in abnormal innervation of the shoulder joint.[2] This is thought to cause abnormal neurovascular reflexes, resulting in hyperaemia and activation of osteoclasts and thus

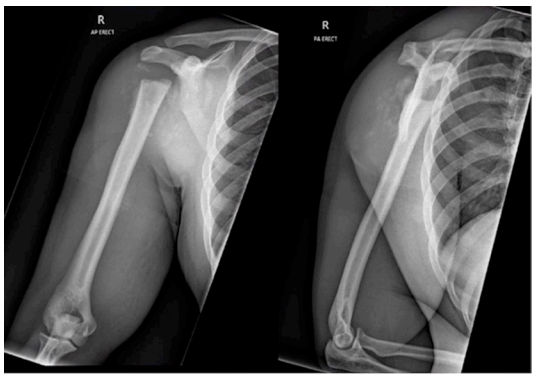

Figure 1 Plain film radiograph showing an aggressive, lytic appearance of the proximal humerus and glenoid.

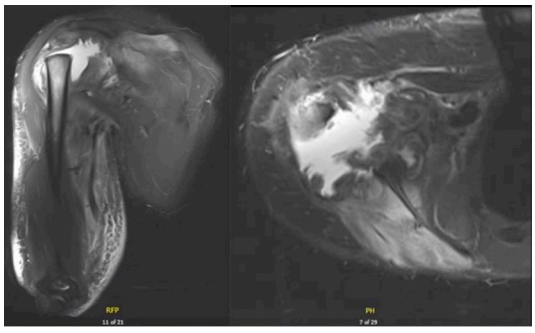

Figure 2 MRI showing a destructive lesion involving both sides of the joint with a large, thick walled fluid filled cavity.

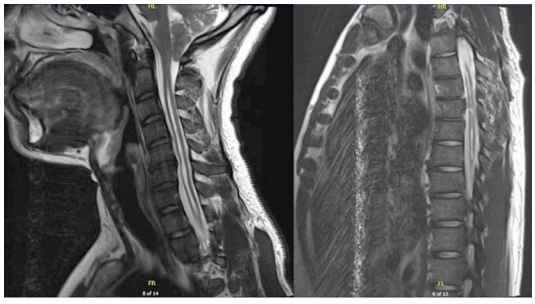

Figure 3 MRI demonstrating an extensive syringomyelia extending from C2-T10.

Learning points

- Charcot arthropathy is a process of joint destruction secondary to a peripheral or central nervous system pathological process.
- The most common cause of Charcot arthropathy in the developed world is diabetes but a syrinx should be excluded when it affects the upper extremities.
- Other causes of destructive joint processes, particularly infection and tumour, should be excluded.
- Treatment of the cause of the neuropathic process is key to preventing progression of disease.

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bone resorption.[3] Treatment of the syringomyelia is required to prevent progression of the disease. This patient was referred to the neurosurgical service for consideration of posterior fossa decompression of the Chiari malformation and shunting of the syrinx.

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REFERENCES

1 Hatzis N, Kaar TK, Wirth MA, *et al*. Neuropathic arthropathy of the shoulder. *J Bone Joint Surg Am* 1998;80:1314–9.
2 Cullen AB, Ofluoglu O, Donthineni R. Neuropathic arthropathy of the shoulder (Charcot shoulder). *MedGenMed* 2005;7:29.
3 Brower AC, Allman RM. Pathogenesis of the neurotrophic joint: neurotraumatic vs. neurovascular. *Radiology* 1981;139:349–54.