LESSON OF THE MONTH

Destructive large joint arthritis

Marian Regan, J K Lloyd Jones, J Snape

Case reports

PATIENT 1

A 72 year old man presented to an orthopaedic unit with a 10 day history of pain in the right shoulder after a fall. He admitted to discomfort in the shoulder for two months before the fall. His right shoulder was swollen and tender. Plain radiography (fig 1) revealed fragmentation and destruction of the humeral head, epiphyseal plate and metaphyseal region of the humerus, with surrounding soft tissue swelling. There was no calcification or periosteal reaction, and minimal irregularity of the glenoid. The appearances suggested a soft tissue tumour. Computed tomography (CT) scan showed fragmentation of the humeral head, a fracture through the head, and loss of fatty interspace between the lesion and deltoid, consistent with a neoplastic lesion. A biopsy specimen was taken and showed fibrous and granulation tissue with fragments of necrotic bone and fibrinous material, but no evidence of malignancy. On review of the radiographs and scans, the possibility of a neuropathic joint was raised. Detailed neurological examination of the patient revealed a mild spastic paraparesis with reduced reflexes in the upper

Differential diagnosis of destructive large joint arthritis

Infection
Tumour
Avascular necrosis
Apatite associated destructive arthritis
Neuropathic joint

limbs. There was patchy loss for light touch and pinprick to T10. The neurology suggested a lesion in the low cervical or mid thoracic spine. Magnetic resonance imaging revealed a large syrinx extending from C2 to the mid thoracic spine, with an associated Chiari malformation.

PATIENT 2

A 50 year old woman presented to casualty with a short history of a painful right shoulder. She had a history of ethanol abuse, but denied having fallen. Examination revealed a swollen shoulder with a large cool effusion. Movements were globally restricted and painful. Plain radiography (fig 2) revealed a destructive process at the humeral head, with flattening and deformity. There was periosteal reaction along the shaft, with extensive soft tissue calcification around the humeral head and neck, and associated soft tissue swelling. The initial diagnosis was of a synovial tumour. Magnetic resonance imaging showed fluid and

Figure 1 Radiograph of the right shoulder in patient 1, showing destruction of the humeral head without substantial periosteal reaction or calcification.

Figure 2 Radiograph of right shoulder in patient 2, showing destruction of the humeral head, periosteal reaction plus soft tissue swelling and calcification.

Department of
Rheumatology,
Harlow Wood
Orthopaedic Hospital,
Mansfield,
United Kingdom
M Regan
J K Lloyd Jones

Department of
Medicine,
Kings Mill Hospital,
Mansfield,
United Kingdom
J Snape

Correspondence to:
Dr Marian Regan,
Department of
Rheumatology,
Derbyshire Royal Infirmary,
Derby DE1 2QY,
United Kingdom.

Accepted for publication
24 March 1995

detritus within the shoulder joint, with erosion of the humeral head and proximal subluxation of the humerus; there was no evidence of tumour. The possibility of a neuropathic joint was suggested.

On questioning, the patient admitted to difficulty lifting her arms and walking in a straight line. On examination she had burns on both arms which had previously been unnoticed. She had nystagmus, weak arms with absent reflexes, absent pain sensation to T4 and a mild spastic paraplegia. Magnetic resonance imaging of the cervical cord and brain revealed an extensive syrinx in the cervical and thoracic cord, communicating hydrocephalus, and an associated Chiari malformation.

Discussion

The now disputed theory that neuropathic joints arise through subclinical trauma in an insensitive joint has led to the common belief that such joints are not painful. Arthralgia is a well documented feature of neuropathic arthropathy, arising in 32% and 50% of affected patients in two studies.^{1,2} Charcot joints are seen in 25% of patients with syringomyelia; in 80% of these the shoulder is affected² as in the patients described. The radiological picture in affected upper limb joints is predominantly a resorptive pattern, with marked loss of bone and little osteophytosis. Severe destruction of the joint can occur over a period as short as six weeks.³ As in our patients, the diagnosis of a neuropathic joint may be the first indication that a patient has an underlying neurological disorder. The development of a neuropathic joint may predate by several years the development of neurological signs.⁴ As syringomyelia is a potentially treatable disorder and surgery is more successful in patients with little neurological disability,⁵ physicians should be alert to this possibility in patients presenting with a destructive arthropathy.

The primary pathogenesis of neuropathic joints is believed to be neurovascular; a neurally initiated vascular reflex leads to very active bone resorption by osteoblasts.^{1,6} This belief is supported by the changes seen on bone scans and angiograms of affected joints suggesting increased blood flow.^{7,8} There may be secondary pathological fractures (as in patient 1) in as many as 23% of cases.¹

Radiographic appearances of a Charcot joint are characteristic. In the initial phase when soft tissue swelling is present, radiograms show only periosteal and parosteal calcification, and sometimes joint effusion. After a few weeks the calcification becomes more dense and the humeral epiphysis loses its profile. Ultimately, the joint ends disappear, the residual bone may be fractured, and free fragments may be present in the joint cavity. There is usually marked soft tissue and periosteal calcification (fig 2). In a patient with extensive bone destruction, the appearances may resemble septic arthritis—particularly that caused by tuberculosis. However, laboratory evidence of acute phase response and systemic upset are absent. The destructive arthritis associated with hydroxyapatite crystal deposition or 'Milwaukee shoulder' may produce a similar picture.⁹ The diagnosis of malignancy may be queried, particularly when the typical soft tissue calcification is absent, as in patient 1. Avascular necrosis may give a similar appearance, and a history of steroid therapy or excessive alcohol intake should be sought.

The lesson

- Neuropathic arthropathy may present as a painful swollen joint.
- Careful neurological examination is particularly warranted in patients presenting with an upper limb, rapidly destructive, large joint arthritis.

The authors acknowledge the help of Mr Simon Frostick, under whose care both patients were originally admitted.

- 1 Brower A C, Allman R M. Pathogenesis of the neuropathic joint: neurotraumatic vs neurovascular. *Radiology* 1981; **139**: 349–54.
- 2 Meyer G A, Stein J, Poppel M H. Rapid osseous changes in syringomyelia. *Radiology* 1957; **60**: 415–8.
- 3 Norman A, Robbins H, Milgrain J E. The acute neuropathic joint arthropathy: a rapid severely disorganising form of arthritis. *Radiology* 1968; **90**: 1159–64.
- 4 Arlecchini S, Bussanelli L, Masetti C. Neurogenic arthropathy: differential diagnosis. *Chir Organi Mov* 1992; **77**: 317–20.
- 5 Burnett H J M, Foster J B, Hodgson P. *Syringomyelia*. Philadelphia: W B Saunders Co, 1973; 3–7.
- 6 Charcot J. Sur quelques arthropathies qui paraissent dépendre d'une lésion du cerveau ou de la moelle épinière. *Arch Physiol Norm Pathol* 1968; **1**: 161–78.
- 7 Sackrelles J C, Swift T R. Shoulder enlargement as the presenting signs in syringomyelia. A report of two cases and review of the literature. *JAMA* 1976; **236**: 2878–9.
- 8 Nigrisoli M, Moscato M, Padovani G. Syringomyelic arthropathy: a description of two cases and a review of the literature. *Chir Organi Mov* 1991; **76**: 237–44.
- 9 Dieppe P A, Doherty M, MacFarlane D G, Hutton C W, Brandfield J W, Watt I. Apatite associated destructive arthritis. *Br J Rheumatol* 1984; **23**: 84–91.