

Neuropathic shoulder in syringomyelia treated with resurfacing arthroplasty of humeral head and soft-tissue lining of glenoid: A case report

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Syringomyelia is cavitation within the spinal cord and most commonly presents in the third to fourth decade. Usually it appears in the lower cervical spinal cord, where it interrupts the decussating spinothalamic fibers and results in loss of pain and temperature sensation in a cape-like distribution across the shoulder girdles.^{5,8} Light touch, vibration sense, and proprioception are preserved. The association of neuropathic arthropathies with syringomyelia was first noted in the 19th century.^{5,9} Neuropathic joints develop in 25% of patients with syringomyelia, of which 80% involve the upper limb, mainly the shoulder.^{4,9} Tabes dorsalis, diabetes mellitus, leprosy, multiple sclerosis, and rare congenital sensory neuropathies are recognized as other causes.^{2,3}

Neuropathic arthropathy, or Charcot joint, is a degenerative arthritis that is slowly progressive over many years. Rapid clinical deterioration and bone loss have been reported,^{3,5} however, and septic arthritis and neoplasia must be excluded by biopsy in such cases.⁹ Repetitive minor or isolated major trauma to a joint with intact motor power but impaired sensation contributes to the etiology, which is controversial and inevitably multifactorial.

Neuropathic arthropathy of the shoulder is a rare condition⁴ but most frequently occurs in patients with syringomyelia, in which it may be the first sign of this underlying condition.⁹ Most commonly, patients present with swelling or pain and often describe and demonstrate stiffness and reduced motion.^{2,9} There is often a history of injury, and radiographic features include humeral head bone resorption, abnormal osseous debris in the soft tissues, and periarticular sclerosis and swelling.^{2,3,5} Although success has been reported with arthrodesis,⁶ surgical treatment of this condition has generally been fraught with difficulty and, indeed, often

thought to be contraindicated.^{5,9} We report a case treated with novel surgical techniques and early success.

CASE REPORT

A 40-year-old, right-handed housewife with a keen interest in gardening was referred to a neurosurgeon for neuropathic pain affecting both right-sided limbs. At presentation, she had marked sensory loss in a cape-like distribution affecting the right upper chest, arm, and abdomen. Investigations revealed a large syrinx within her spinal cord, extending from the upper cervical region into the lower thoracic region. The following year, surgery was performed to drain the syrinx and was repeated 4 years later. Affected bowel function required a colostomy, and a neuropathic bladder necessitated intermittent self-catheterization.

At the age of 47, she was referred with a 4-year history of increasing activity-related and nocturnal pain in the right shoulder that was unresponsive to conservative measures. There was no history of injury or trauma of any magnitude. On examination, the shoulder was swollen, and there was crepitus with active elevation to 100°. There was good rotator cuff strength and no apparent motor or proprioceptive deficit.

Radiographs revealed flattening of the humeral head at the medial caudal aspect, bone-on-bone articular degeneration with abnormal soft-tissue calcification, and sclerosis of the glenoid (Figure 1). Advanced destruction of the glenohumeral joint was noted on computed tomography (CT) scan, with some loss of bone stock symmetrically and sclerosis of the glenoid (Figure 2).

The patient was counselled thoroughly concerning management options and 2 months later underwent uncemented Mark 3 (hydroxyapatite coated) Copeland resurfacing arthroplasty (Biomet Merck, Ltd, Swindon, United Kingdom) with a soft-tissue interposition graft of the glenoid.

Surgical technique

A standard deltopectoral approach was used, with a subscapularis tenotomy. Obvious gross destructive disease was encountered within the joint. The capsule, consisting of the middle and anteroinferior glenohumeral ligaments, was mobilized from the subscapularis tendon, reflected posteriorly onto the lightly burred surface of the glenoid, and secured with Ethibond sutures (Ethicon, Worth Ryde NSW, Australia) to the posterior glenoid labrum. A standard size Mark 3 Copeland arthroplasty prosthesis was applied to resurface the humeral head. The subscapularis tendon was repaired with nonabsorbable sutures.

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Figure 1 Preoperative plain radiograph.

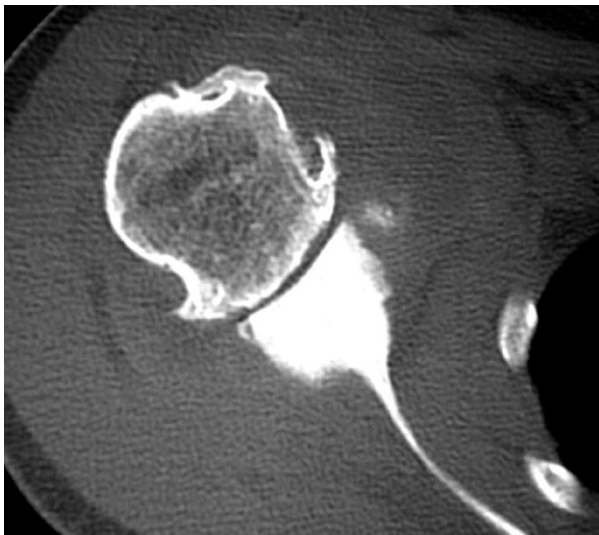


Figure 2 Preoperative computed tomography scan.

The patient's early postoperative recovery was uneventful. She experienced excellent initial relief from pain and crepitus, and the postoperative radiograph was satisfactory (Figure 3).

At 14 months, the patient reported return of mild activity related pain. At review, active elevation was 140°, and good rotator cuff power had been maintained. Radiographs at this stage showed the abnormal appearance of apparently progressive sclerosis of the entire glenoid with some new bone formation inferiorly, likely due to uncon-



Figure 3 Postoperative radiograph.



Figure 4 Radiograph at 14 months.

trolled pressure of the prosthesis on the glenoid. The prosthesis appeared quite solid, with no sign of loosening or superior subluxation (Figure 4).

The patient was advised to rest for a few weeks until symptoms settled and then return to limited daily activities with awareness of the concerns of potential loosening or increased bone loss. At her most recent review, 24 months after surgery, the patient was entirely satisfied with the shoulder and reported no pain. The American Shoulder and



Figure 5 Radiograph at 24 months.

Elbow Surgeons score (modified for no sport) was 81.5, and she had active elevation of 140° and external rotation of 50°. The most recent radiographs showed no change from the previous year's films (Figure 5).

DISCUSSION

Neuropathic arthropathy should always be considered in an evaluation of a painful swollen shoulder. The case presented here did not have humeral head destruction but did have diagnostic radiologic features previously described in the literature: mediocaudal flattening of the humeral head, periarticular soft-tissue calcification, and glenoid sclerosis.⁹ Clinically, typical features of swelling, crepitus, and pain were present in the shoulder. Syringomyelia had been proven with magnetic resonance imaging, and consultation with neurologic colleagues confirmed the diagnosis of Charcot joint due to syringomyelia.

Hatzis et al⁴ presented 6 patients with destruction of the humeral head in neuropathic shoulders. Five had proven syringomyelia with a history of injury, and the other patient had no reliable history of trauma but had a long history of alcohol abuse.⁴ We believe that the fortunate relative pres-

ervation of humeral head bone in our patient was a result of a lack of trauma to the shoulder. The radiologic features seemed to indicate potential pending deterioration.

Traditionally, orthopedic surgeons have been reluctant to operate on such cases because there have been disappointing results with both arthrodesis and arthroplasty.⁴ Joint replacement surgery is always a concern owing to the increased risk of early loosening, particularly of a glenoid component. At her relatively young age and with adequate humeral head bone, it was decided to offer the patient an uncemented Copeland resurfacing arthroplasty of the humeral head, to preserve humeral head bone stock, with soft-tissue lining of the glenoid, in the hope this would lessen any potential pain from the prosthesis–bone interface and also diminish, for a time, wear and loss of glenoid bone.

Levy and Copeland⁷ have reported good results using the resurfacing humeral head prosthesis, and Burkhead and Hutton¹ report that biologic resurfacing of the glenoid with a hemiarthroplasty may give improved results compared with hemiarthroplasty alone. To our knowledge, the 2 techniques in combination have not yet been reported, and this was conveyed to the patient. The theoretic benefit of such a surgical approach is protection of existing humeral and glenoid bone, with its preservation for use in arthrodesis in the future should the need arise.

The glenoid sclerosis needs monitoring with careful assessment of the remaining bone stock using CT scans. Although the clinical and radiologic situation is stable at present, the patient has been advised to report symptom deterioration early, and close regular review is planned.

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