Neuropathic Shoulder Arthropathy Associated with Syringomyelia and Arnold-Chiari Malformation (Type I)

LUCREZIA RIENTE, MD; SABRINA FRIGELLI, MD; ANDREA DELLE SEDIE, MD, University of Pisa, Rheumatology Unit, Dipartimento Medicina Interna, Via Roma 67, 56126 Pisa, Italy. Address reprint requests to Dr. Riente. E-mail: l.riente@int.med.unipi.it

Neuropathic arthropathy is a destructive joint disease frequently associated with syringomyelia (up to 25% of cases)[1]. Syringomyelia may sometimes be a complication of Arnold-Chiari malformation[2], a condition characterized by herniation of the cerebellar tonsils into the foramen magnum. We describe a patient in whom all 3 conditions were present.

A 53-year-old woman developed pain, burning paresthesia, and swelling of the left shoulder, which was also limited in motion. Serum fibrinogen, cholesterol, and IgM concentrations were moderately elevated, and syphilis serology was negative. Left shoulder radiograph showed absorption of the humeral head and glenoid and calcifications of the periarticular soft tissue (Figure 1), confirmed by computed tomography scans. Mycobacterial, fungal, and bacterial infections were excluded on the basis of cultures of a specimen obtained by open synovectomy of the left

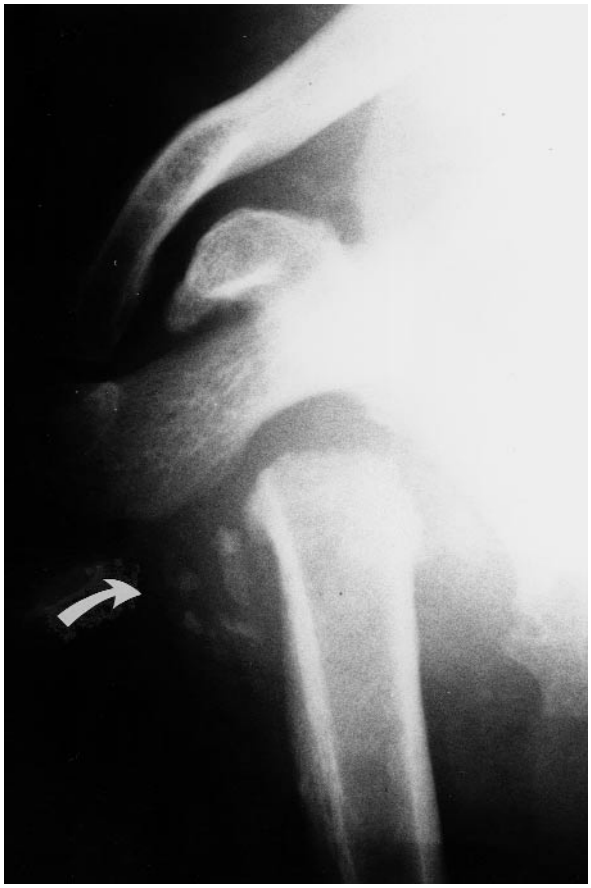

Figure 1. Left shoulder radiograph showing absorption of the humeral head and glenoid, and calcifications of the periarticular soft tissue.

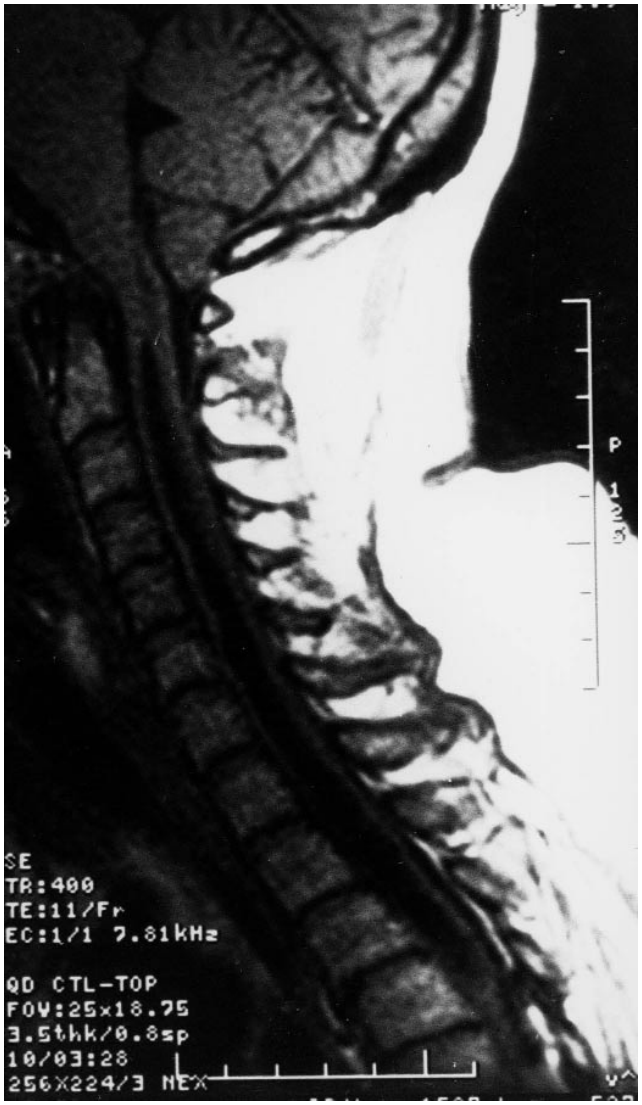

Figure 2. MRI of the cervicothoracic spine showed syrinx spreading from C2 to T7, with cerebellar tonsillar herniation into the foramen magnum.

shoulder, and no signs of villonodular synovitis were found on histology.

After 6 months she complained of reduction of strength and exacerbation of the paresthesia in both arms. Neurological examination revealed limitation of motion of both arms, fingers slightly "en griffe," hypotrophy of the thenar and interosseous muscles of the hands, and hypoactive deep tendon reflexes in the upper extremities and brisk deep tendon reflexes in the lower extremities. Electromyography of arms and legs showed abnormalities compatible with denervated muscles in the left hand and abnormal motor and sensory conduction of the right ulnar nerve. A gadolinium enhanced magnetic resonance study of the cervicothoracic spine disclosed syrinx spreading from C2 to T7, with cerebellar tonsillar herniation into the foramen magnum (Figure 2). A diagnosis of syringomyelia associated with Arnold-Chiari malformation (type I) was made; she underwent cervical-spinal decompression that resulted in partial recovery of her sense of pain and temperature[3].

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