Neuropathic arthropathy. Answer to the e-quad “A highly damaged shoulder”☆

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Case report

A 53-year-old woman was hospitalised for an assessment of monoarthritis of the left wrist. Among the antecedents, tubercular meningitis was treated in 1988 with the insertion of a ventriculoperitoneal shunt. The clinical examination revealed necrotic pulp of the 3rd row of the left hand, and sensory disorders (dysesthesia and hypoesthesia) of the left hand. An EMG found a conduction block of the left median and cubital nerves crossing the wrist and signs of nerve root at C7-C8 on the left and C8 on the right.

The X-rays revealed an infiltration of the soft parts of the left wrist without significant bone destruction (Fig. 1a). An MRI of the hands and wrists found bilateral damage with inferior radio-ulnar articular synovitis, left radio-carpal, MCP and predominant PIP extensively on the left and tenosynovitis of almost all of the tendons of the palmar and dorsal sides (Fig. 1c and d). An echo-guided articular puncture did not find any signs of sepsis (no pyogenes or mycobacteria), a slightly inflammatory haemorrhagic fluid, without microcrystals. On the biological level, a moderate inflammatory syndrome, a rheumatoid factor and negative anti-CCP antibodies, slightly positive ANA at 200 and positive anti-SSa positive at 116 were noted. The rest of the assessment did not reveal anything specific. A diagnosis of chronic inflammatory rheumatism such as Sjögren’s syndrome was raised, with possible super-infection of the left wrist. Complementary surgical biopsies of the synovials were carried out, without any germ found, with a slightly inflammatory appearance of the synovial. Four months later, her left shoulder was red, mildly painful and she suffered from moderate functional disability. The puncture of the articular fluid found a haemorrhagic fluid, without microcrystals and few nucleated elements. All of the samples were sterile. Control X-rays of the left wrist (Fig. 1b) revealed a shock of the carpus with destruction and dislocation of the bones of the first row. Front X-rays of both shoulders were taken (Fig. 2).

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☆ Here is the answer to the case “A highly damaged shoulder” previously published in the No. 1/2013. As a reminder we publish again the entire case with the response following.
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Neuropathic arthropathy

Figure 1. a: frontal view of the hands and wrists; b: control frontal view at month 4 of the hands and wrists; c: MRI of both hands Coronal T1; d: MRI of both hands Coronal T1 with fat saturation (Fat Sat) after injection of gadolinium.

Figure 2. Comparative frontal view of both shoulders 6 months after the initial treatment.

What is your diagnosis?

After reading the case report, what diagnosis would you choose from the following proposals:
- chronic autoimmune arthritis (chronic inflammatory rheumatism);
- infectious tubercular arthritis;
- neuropathic arthropathy;
- microcrystalline arthropathy.

Diagnosis

Destructive neuropathic arthropathy (syringomyelic).

Discussion

The X-ray assessment, in view of the picture of arthropathy of the left shoulder, reveals a full lysis of the left head of the humerus with the presence of peripheral osseous debris and gleno-humeral dislocation, without damage to the glenoid or acromioclavicular side and without contralateral anomaly (Figs. 3–4). Although the damage to the wrists may initially indicate the diagnosis of chronic autoimmune arthritis, the lesions of the left shoulder do not support this. The moderately painful, asymmetric and very fast nature of the destruction with complete fragmentation of the head of the humerus contrast with the respect of the glenum, the not very distinct biological inflammatory syndrome and the low positivity of the auto-antibodies. The multiple sterile samples and the asymmetric nature of the articular destruction do not indicate infectious arthritis. The absence of microcrystals makes the diagnosis of microcrystalline arthropathy unlikely. However, this series of arguments indicates neuropathic arthropathy: the spinal MRI (Fig. 5) reveals a
Figure 3. a: frontal view of the hands and wrists. Infiltration of the soft parts of the left wrist; b: control X-ray at month 4: shock of the carpus with distinct destruction of the first row of the left carpus. MRI of both hands; c: coronal T1: major bone oedema of the carpus, the bottom of the metacarpals and the lower end of the left ulna and radius; no contralateral bone oedema; d: coronal T1 with fat saturation (Fat Sat) after injection of gadolinium: articular synovites and bilateral tenosynovites predominating on the left carpus.

Figure 4. Comparative frontal view of both shoulders 6 months after the initial treatment. Complete lysis of the left head of the humerus with bone fragments, global respect of the glenoid opposite and contralateral integrity.

syringomyelic fluid cavity in T2 hypersignal, T1 hypersignal without contrast enhancement after the injection of gadolinium. It extends from C1 to T9, filiform until C6, then wide upstream reaching 11 mm in diameter, laminating the medullary cord, without anomaly of the cranio-cervical junction. The final diagnosis is that of destructive neuropathic arthropathy of the shoulder on the syringomyelic cavity, possibly following tubercular meningitis.

Neurogenic arthropathy of the shoulder is a rare disease [1] corresponding to chronic destructive arthritis. There is often a divergence between the low pain and the major bone destruction, generally evolved at the diagnostic stage, with a quickly evolving picture [1–3]. The functional disability varies. Neurological symptomatology is almost always associated (reflexo-tendinous or sensory anomalies, weakness or clonus of the limbs), although the symptoms of neuropathic arthropathy may mask those of the syringomyelia [1,4,5]. The radiography and MRI provide arguments in favour of neuropathic arthropathy and allow the bone destruction to be assessed before deciding on therapy. The osteoarticular modifications are related to the repeated mechanical traumatisms of a joint that have become insensitive to pain and leading to its destruction, or according to the neurovascular theory, to stimulation of the vascular reflexes by the autonomous nervous system, with activation of bone resorption by the osteoclasts [2,5].

The aetiologies of neuropathic arthropathy of the shoulder are varied and include syringomyelia, diabetes, syphilitic atrophy, chronic alcoholism and more rarely, malformation of the cranio-cervical junction (Arnold-Chiari) [1–5].
Figure 5. Panmedullar MRI. Demonstration of an extensive syringomyeletic cavity from C1 to T9 in hypersignal T2, hyposignal T1 and without contrast after injection of gadolinium. Sagittal sections with T2 weighting: a: cervical; b: thoracic; c: sagittal cervical section T1 weighted; d: sagittal thoracic section T1 weighted Fat Sat after injection of gadolinium.

Neuopathic arthropathy is found in 25% of all syringomyelias, of which 80% are located in the arm, the shoulder and elbow being the joints most affected, most often in a unilateral manner [4,5]. The time before the occurrence after the formation of the syringomyelic cavity varies. The evolution depends on the aetiology, the rapidity of the diagnosis and the care. The treatment most often involves management with measures to prevent repeated traumas [2,4,5]. Surgery is used to treat evolved arthropathies, with major bone lysis (arthrodesis, arthroplasty). In the case of post-syringomyelia neuropathic arthropathy, the evolution of the bone destruction is stopped by the neurosurgical decompression of the marrow, without which the orthopaedic treatment is of no use [4].

Conclusion

Neurogenic arthropathy of the shoulder is a rare disease with various aetiologies. Syringomyelia is foremost among them. It should be considered when faced with a picture of rapid destruction contrasting with functional disability and moderate pain. Imaging (simple X-rays, MRI) is indispensable as a complement for the biological assessment in determining the diagnosis, to allow for the monitoring and determine the aetiology. The main treatment of post-syringomyelia neurogenic arthropathy of the shoulder consists of medullar neurosurgical decompression. Orthopaedic treatment should only be considered once the bone destruction process has been halted.
Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

References


