NEUROPATHIC ARTHROPATHY OF THE HAND ASSOCIATED WITH CERVICAL SYRINGOMYELIA

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Abstract

Neuropathic osteoarthropathy is a rare chronic, degenerative arthropathy associated with decreased sensory innervation. Numerous causes of this arthropathy have been described, syringomyelia, fluid-filled intramedular cavity, being among them. Neuropathic osteoarthropathy associated with syringomyelia is usually mono/oligoarticular, asymmetrical, involving the elbow, shoulder, rarely the wrist.Skin and nails trophic changes sometimes may appear.

The present case is that of a female patient with asymmetrical oligoarthritis of right wrist and metacapophalangean joints with rapid reumathoid-like deformity of the hand.The diagnosis key was the radiologic aspect with both lytic and sclerotic lesions but also the thermal sensory impairment, minimized by the patient, longtime considered to be related to cervival spine osteoarthritis. The MRI examination of the cervical spine confirmed the fluid-filled cavity of the whole cervival region with secondary medular atrophy.

Keywords: neuropathic arthropathy, syringomyelia

CASE PRESENTATION

B.U., female waitress, aged 34, comes to the outpatient clinic for painful swelling of right wrist, 2nd and 3rd MCP joints with insidious onset, 2 months ago. She also.relatedmechanical pain of the right shoulder and relapsing episodes of mechanical low back pain.

Physical examination reveals grade I obesity, hypopigmented scars on the left forearm and wrist (after thermal burns), "camel's back" deformity of the right wrist, interosseous muscle atrophy, carpal deviation of the right metacarpophalageal joints (MCP), paintful swelling of 2nd and 3rd MCP joints,limited range of motion in the right wrist. The skin of the right hand was thickened with hyperhidrosis; left shoulder was also paintful with reduced mobility.

Lab examination showed minimal inflammatory syndrome (ESR = 38 mm/h, fibrinogen = 372 mg/dl, CRP = 9.7 mg/dl), negative rheumatoid factor, antiCCP autoantibodies, antinuclear autoantibodies, normal level of serum and urinary uric acid .

Xray examination of both hands revealed extensive resorbtion of right carpal bones with fragmentation, condensationof subchondral bone with fragmentation and intra-articular calcification with new heterotopic bone formation and subluxation.

Ultrasound examination of the right wrist revealed grade II synovitis with Doppler signal and multiple erosions of carpal bones,increased dimensions of right median nerve (17 mm).

The MRI examination of the right wrist revealed avasculat necrosis of the scaphoid, multiple erosions with positive STIR signal in all carpal bones, distal extremity of radius and ulna (Fig. 3).

Based on the previously mentioned investigations we could exclude rheumatoid arthritis (asymmetry, minimal inflammatory syndrome, negative RF and antiCCP autoantibodies), crystal arthropathies (normal serum and urinary uric acid, lack of

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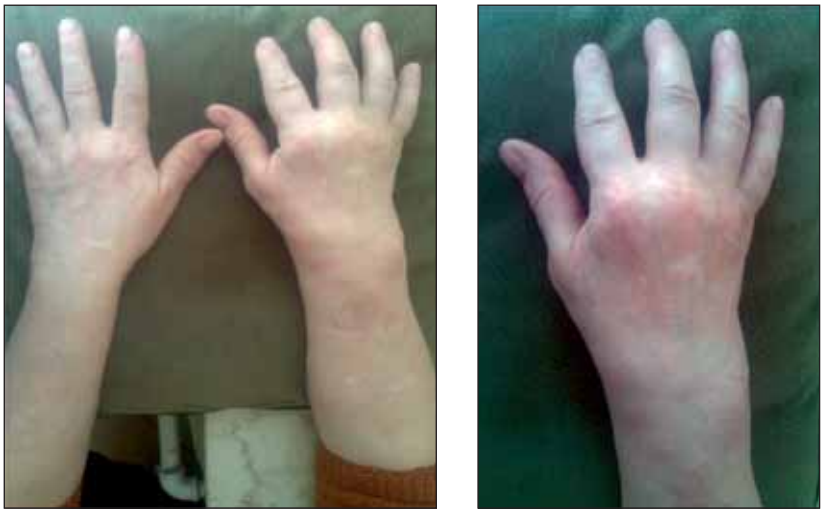

FIGURE 1. *Rheumatoid-like asymmetrical deformity of the right hand:"camel's back" deformity, interosseous muscle atrophy, carpal deviation of MCP, swelling of 2nd and 3rd MCP joints*

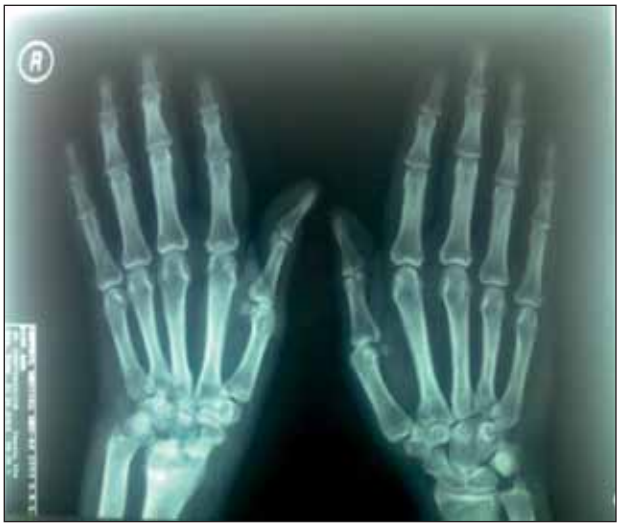

FIGURE 2. *Hand X-ray – advanced distructive changes of the radiocarpal joint, ulnar deviation, patchy osteoporosis*

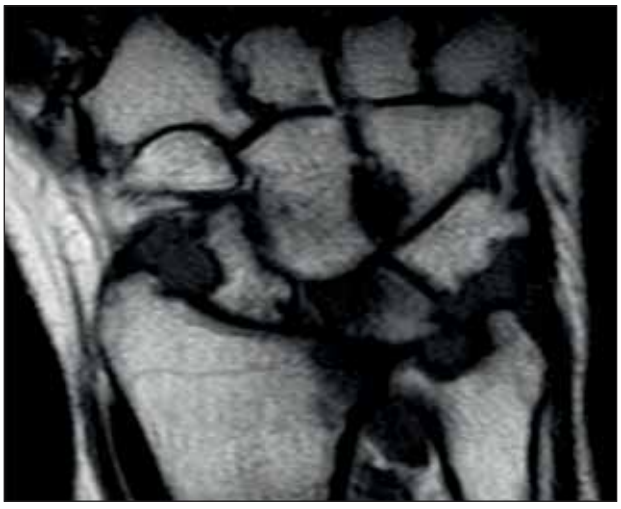

FIGURE 3. *Vascular necrosis of the scaphoid, erosions in carpal bones, distal extremity of radius and ulna*

typical ultrasound features, normal calcium and magnesium level, normal PTH), spondylarthritis (HLAB27 negative, without sacroiliitis on pelvis X-Ray), septic arthritis (insidious onset, no fever, normal blood leukocytes); the mottled appearance of the skin and hyperhidrosis might also suggest reflex sympatethic osteodystrophy, but would not explain erosions and the STIR signal on the MRI examination of the wrist.

Careful medical history of the patient mentioned some sensitivity abnormalities in the upper limbs with significantly diminished thermal perception, slightly diminished sensitivity to touch and paresthesias in the first three fingers of both hands; neurological symptoms started 10 years ago with gradual enhacement but they were related by the general practitioner to cervical osteoarthritis of spine.″An

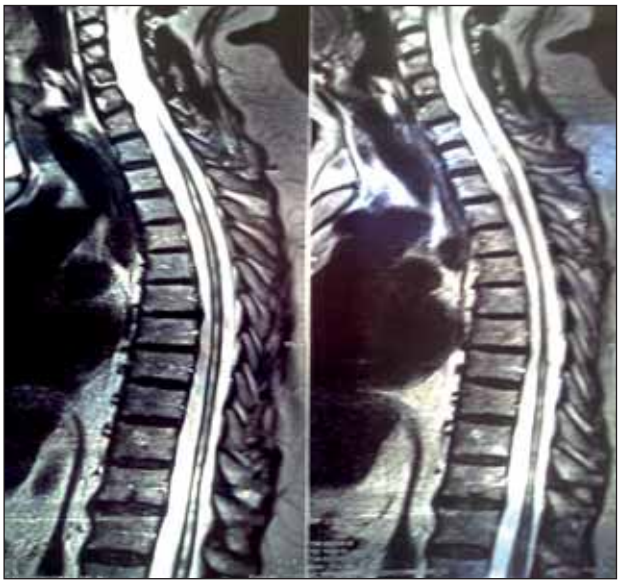

FIGURE 4. *MRI of cervical spine showing dilated cavity fluid-filled of the whole cervival segment with spinal atrophy*

electromyography was performed and the diagnosis of bilateral carpal tunnel syndrome was confirmed.

The MRI of the cervical spinerevealed degenerative changes (osteoarthritis at C5-C6 interapophyseal joint) and, most important, a dilated cavity with the same intensity as cerebrospinal fluid on T2-weighted imaging in the whole cervival segment with secondary medular atrophy. Considering all this, the final diagnosis was: "Neuropathic arthropathy of the right wrist, Cervical syringomielia. Bilateral carpal tunnel syndrome" .

Treatment included symptomatics (nonsteroidal anti-inflammatory drug), oral bisphosphonates. She was reffered to an orthopaedics unit for adapted orthesis and to the physical therapist.

DISCUSSIONS

Neuropathic arthropathy, also known as Charcot arthropathy is a progressive and distructive arthopathy secondary to a peripheral neuropathy (1,2). Any condition that causes sensory or autonomic neuropathy can lead to a Charcot joint. Charcot arthropathy occurs as a complication of diabetes (0.1-0.5%), syphilis (5-19% of the patients with tertiary syphilis), chronic alcoholism, leprosy, myelomengocele, spinal cord injury, syringomyelia, renal dialysis, congenital insensitivity to pain, chronic alcoholism. Up to 30% of the cases are idiopathic. The pathogenesis is quite complex, two major theory being discussesed:neurotraumatic theory and neurovascular theory(3).

The etiology determines the type of joint involvement (3-5). Untreated syphilis involves the hips and the knees, diabetes usually involves the foot and syringomielia upper limbs (5). The onset is usually insidious, very rare acute or subacute with mono or oligoarticular involvement, although some poliarticular cases have also been reported (5). Acute Charcot arthropathy almost always presents with signs of inflammation: swelling, an increase in local skin temperature, erythema, joint effusion, and bone resorption. Pain can occur in more than 75% of patients; however, the pain's severity is significantly less than would be expected based on the severity of the clinical and/or radiographic findings. Instability and loss of joint function also may be present. Passive movement of the joint may reveal a „loose bag of bones" (1,2). Approximately 40% of patients with acute Charcot arthropathy have concomitant ulceration, which complicates the diagnosis and raises concerns that osteomyelitis is present (4).

In early stages X-ray examination reveals soft tissue edema, then destruction of articular surfaces starts with opaque subchondral bones, joint debris, deformity, and dislocation. Neuropathic arthropathy can be classified into hypertrophic and atrophic types (6). Progressive joint effusion, fracture, fragmentation, and subluxation should raise the suspicion of neuroarthropathy. In the advanced stages, abnormal findings on radiographs include subchondral sclerosis, osteophytosis, subluxation, and soft-tissue swelling. Long-standing neuroarthropathy is characterized bycomplex disorganization of joints (1,5). Nuclear imaging, CT or MRI scanning are mainly use for early diagnosis or to exclude infection.

Synovial fluid examination usually shows a clear yellow effusion but sometimes the fluid can be bloody or cloudy due to lipid crystals (1,2). Synovial biopsy findings consist in considerable amounts of cartilaginous and osseous debris within the synovial membrane (termed detritic synovitis). Inflammatory syndrome is usually absent(1).

Differential diagnosis include osteonecrosis, calcium pyrophosphate dihydrate crystal deposition disease, psoriatic arthritis, osteoarthritis and osteoarticular tumors (1,2).

Treatment of Charcot arthropathy is primarily nonoperative. Treatment consists of 2 phases: an acute phase and a postacute phase. Management of the acute phase includes early diagnosis, NSAIDs, immobilization and reduction of stress in order to diminish joint destruction (1,2,7). When possible, treatment of the neurological condition is recommended but joint destruction is irreversible (1,7). Immobilization usually is accomplished by casting. Reduction of stress is accomplished by decreasing the amount of weight bearing on the affected extremity. While total non-weight bearing is ideal for treatment, patients are often not compliant with this treatment. Studies have shown that partial weight bearing with assistive devices (eg, crutches, walkers) also is acceptable without compromising healing time (6,7). Bisphosphonates have been shown to be effective for reducing bone turnover markers and skin temperature in some studies. Nevertheless, the long-term efficacy, specifically regarding the occurrence of deformities and ulcerations, remains to be demonstrated as no follow-up studies have been published (8). Patient's medical comorbidities, compliance, location and severity of deformity, presence or absence of infection, pain or instability are the

factors considered in the decision of surgical treatment. Due to the increased risk of wound infection and mechanical failure of fixation, surgery should be avoided during the active inflammatory stage (1,6).

SYRINGOMYELIA – GENERAL DATA

Syringomyelia is the development of a fluid-filled cavity or syrinx within the spinal cord. (10). Etiology of syringomyelia is often associated with craniovertebral junction abnormalities: bony abnormalities, soft-tissue masses (tumors, inflammatory masses), neural tissue abnormalities (cerebellar tonsils and vermis herniation, Chiari malformation), membranous abnormalities (arachnoid cysts), posthemorrhagic or postinflammatory membranes. Other etiologies not associated with craniovertebral abnormalities may include the following: arachnoid scarring related to spinal trauma , meningeal inflammation, subarachnoid space stenosis due to spinal neoplasm or vascular malformation or idiopathic (10,11). The most frequent localization is the cervical spine (11). Estimated prevalence of the disease is about 8.4 cases per 100,000 people. The disease usually appears insidious in the third or fourth decade of life (10,12).

Symptomatic presentation depends primarily on the location of the lesion within the neuraxis. Clinical picture can be modified by the associated neurological condition (11,12). Syrinx interrupts the decussating spinothalamic fibers that mediate pain and temperature sensibility, resulting in loss of thermal sensations, while light touch, vibration, and position senses are preserved (dissociated sensory loss). Pain and temperature sensation may be impaired in either one or both arms, or in a shawllike distribution across the shoulders and upper torso anteriorly and posteriorly (13,14). When the cavity enlarges to involve the posterior columns, position and vibration senses can be impaired.Syrinx extension into the anterior horns of the spinal cord damages motor neurons and causes diffuse muscle atrophy that begins in the hands and progresses proximally to include the forearms and shoulder girdles. Clawhand may develop (10-12). Other manifestation includefasciculaions, trophic changes of the skin, nails or hair (″turtle skin”, ″juicy hand″, edema, hyperhidrosis), Charcot arthropathy, Claude-Bernard-Horner syndrome or autonomic changes (10,11).

Syringomyelia associated neuropathic arthropathy can appear in up to 25% of the patients with syringomyelia, upper limbs being involved in 80% of the cases (15).

Syringomyelia associated neuropathic arthropathy is characterized by slow progression, usually monoarticular; the joints involved most frequently are the shoulders and elbows. Still, the disease isquite rare and often misdiagnosed, the largest case serie consists of 12 patients (17).

A variety of surgical treatments have been proposed for syringomyelia (suboccipital and cervical decompression, laminectomy and syringotomy, shunts, terminal ventriculostomy etc.) depending on the etiology and localisation. Surgical treatment is not mandatory for old patients, asymptomatic patients, lack of progression (10,14).

DISCUSSIONS

Insidious onset, rheumatoid-like deformity of the hand created first the confusion with rheumatoid arthritis, treatment with sulphasalazyne was strated with lack of efficacy What was striking at physical examination was hand edema and skin trophic changes (sclerodactily, mottled appearance, hyperhidrosis) that might had led the diagnosis to algoneurodystrophic syndrome. Lack of specific rheumatic laboratory tests and radiological signs of lytic and sclerotic lesions leaded to other investigations. A key element for the diagnosis wasasymmetrical thermal sensitivity impairment related by the patients for more then 10 years in both hands and forearms, but they were neglected and considered to be related to cervical spine osteoarthritis. Cervical spine MRI confirmed the suspicion of syringomielia of the whole cervical spine with medular atrophy.

The main particularity of the case is the neuropathic arthropaty of the hand, although it is known that usually Charcot involves the shoulder or the elbow. Another particularity is the fact that the patients minimized the sensitivity impairement leading to late diagnosis of both neurologic and rheumatic condition.

CONCLUSIONS

Neuropathic arthropathy associated with cervical syringomyelia is a rare condition.Careful medical history, physical examination and X-ray are the clues to proper diagnosis. Although there is no specific treatment, early diagnosis could halt the joint destruction.

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