

Bilateral neurogenic hip arthropathy

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Abstract

Neurogenic arthropathy is a rare joint disorder, characterized by rapid progression and marked destruction of articular surfaces (known as Charcot's joint) with only mild to moderate pain. Most cases are related to diabetic neuropathy, but they may complicate the course of other neurogenic clinical conditions such as neurosyphilis (tabes dorsalis), syringomyelia, myelomeningocele, Peroneal Muscular Atrophy, spine or peripheral nerve injury, alcoholism and avitaminosis. Loss of superficial sensation and proprioception plays a pivotal role in development of arthropathy because it affects the joint's normal protective reflexes and leads to joint instability, degeneration and destruction. The authors describe an unusual case of bilateral neurogenic hip arthropathy in a 61-year old women who developed this condition in the course of multilevel lumbar spondylosis with spinal and nerve root compression. The patient underwent a successful bilateral hip replacement and remains almost asymptomatic contrary to literature reports that suggest high risk of complications after the operative treatment of Charcot's joints.

INTRODUCTION

The natural history of the hip osteoarthritis can be summarized as a long-standing degenerative process that leads to joint destruction and variable symptoms, usually pain and restricted movements. Sometimes, however, one sees a patient with rapid progression that results in a severe disability over a short period of time (Flik and Vargas, 2000). Those cases of secondary osteoarthritis are mostly related to hip trauma, metabolic or endocrine disorders, and rheumatological conditions (Hasegawa *et al.* 1997). Only rarely the arthropathy is caused by neurogenic disorders.

The first description of neurogenic arthropathy was made in 1868 by Jean-Martin Charcot, who described a characteristic pattern of joint destruction in patients with tabes dorsalis. Nowadays, most cases of *Charcot joints* are related to diabetic

Abbreviations :

ROM - Range of motion
MRI - Magnetic Resonance Imaging
NSAID - Non-steroidal anti-inflammatory drugs
VTE - Venous thromboembolism

neuropathy, but they may also occur in the course of other neurogenic clinical conditions such as syringomyelia, myelomeningocele, Peroneal Muscular Atrophy (Charcot-Marie-Tooth disease), spine injury, peripheral nerve injury, alcoholism and avitaminosis (Sprenger and Foley, 1982) The key factor in the pathogenesis of joint destruction is the loss of peripheral sensation and proprioception. This results in impaired joint stability (both static and dynamic) and poor fine motor control, that, in turn, leads to repetitive microtrauma and progressive degeneration and joint destruction (O'Connor *et al.* 1985).

The authors describe a case of a 61-year-old woman with bilateral hip arthropathy attributed to neural deficits caused by lumbar spondylosis with associated disc herniation.

CASE DESCRIPTION

In February 2007, a 61-year old women was referred for hip surgery at our department. We noted unusually quick progression of hip disorder and her history was remarkable for spine surgery due to discopathy and subsequent neural deficits, which allowed us to establish the diagnosis of bilateral neurogenic arthropathy.

The patient had a long-standing history of low back pain that was initially treated in the primary care center and later, for almost 4 years, at an Orthopedic outpatient clinic. The physical examination performed there revealed positive straight leg raising test on the left at 60 degrees of flexion, hip ROM was normal. Lower extremity motor strength was normal and symmetric. Peripheral sensation was not disturbed.

Due to progression of pain a MRI study was obtained in 2003. The study revealed marked degenerative changes within the vertebral column, particularly involving L3, L4, L5 vertebral bodies with irregular terminal plates. There was a mild peripheral disc bulging at the level of L2–L3 without nerve root compression. At the L3–L4 level there was a mild disc bulging and osteophytes which resulted in stenosis of the spinal canal and nerve root canal on the left. At the L4–L5 level there was a marked postero-lateral disc protrusion (to the left) with narrowing of the spinal canal and compression of the dural sac and the left nerve root. A postero-lateral disc protrusion with dural sac compression was also noted at the L5–S1 level. The sagittal diameter of the spinal canal was decreased in the lower lumbar spine but normal at the level of conus medullaris. A plain radiograph of pelvis, which was made as a part of differential diagnosis, demonstrated normal outlines of both femoral heads with some subchondral sclerosis of acetabula and narrowing of the joint space (Figure 1).

The patient did not consent for the proposed spine surgery. Therefore she was treated conservatively with NSAIDs, myorelaxants and physical therapy. Despite treatment, her condition worsened. Due to progression of pain and development of motor and sensory deficits (involving mostly the right thigh) another MRI was obtained in 2005. The study revealed progression of degenerative bony changes and multilevel spinal compromise as well as nerve root compression between L2 and S1 vertebrae. (Figure 2). The patient was subsequently referred for operative treatment. On admission to the Department of Neurosurgery she was only able to walk in a walker, with severe pain of the lumbosacral



Fig. 1. A plain radiograph of pelvis in 2003y.

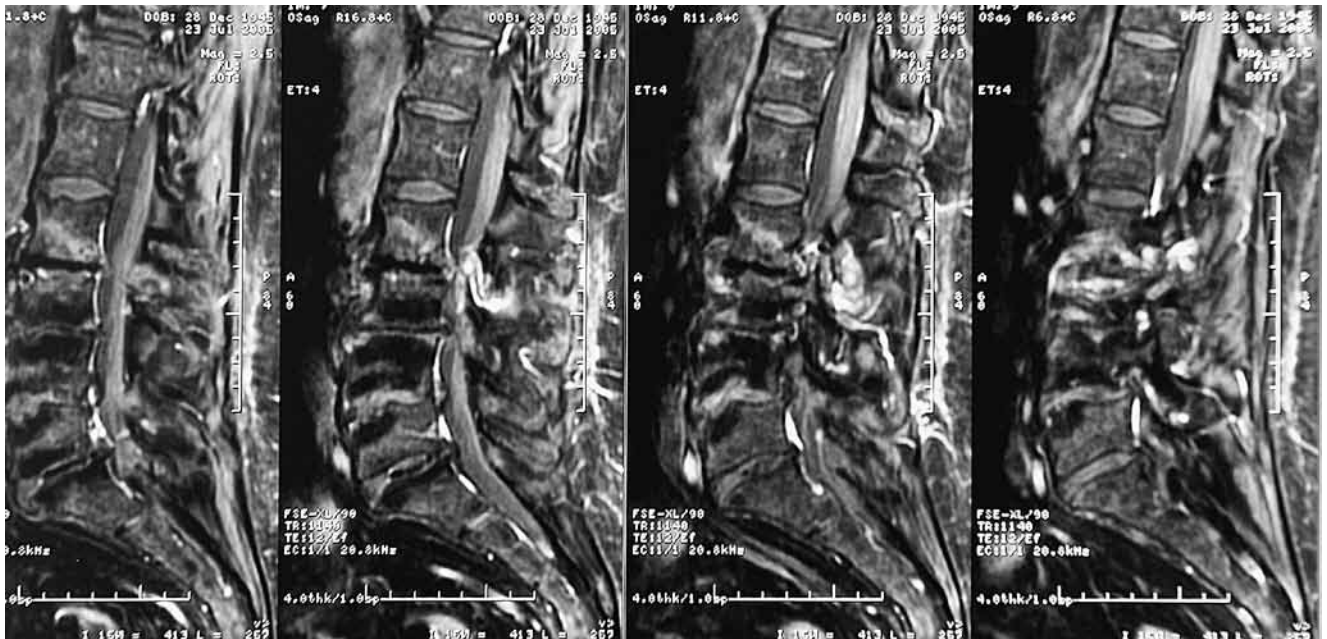


Fig. 2. Progression of degenerative bony changes and multilevel spinal compromise.

area and motor deficits of lower extremities (2/5 in the Lovett score).

After the scheduled spinal decompression the back pain subsided and motor function gradually improved. However, during the postoperative rehabilitation program a new problem became evident: an increasing bilateral hip pain, more pronounced on the left side, without significant limitation of joint range of motion. Due to pain issues a radiograph of pelvis was obtained,

which demonstrated destruction of the articular surfaces, rarefaction within the proximal femora and subchondral sclerosis. (Figure 3) The radiographic appearance was suggestive of septic or rheumatoid arthritis, therefore a set of laboratory test and additional radiographic studies was ordered. It revealed normal level of acute-phase proteins, slightly elevated uric acid, positive Waaler-Rose test for Rheumatoid Factor and negative anti-CCP antibody test. Radiographs of



Fig. 3. A plain radiograph of pelvis demonstrate destruction of the articular surfaces.

both hands and feet showed no rheumatoid features. The clinical presentation and results of accessory studies allowed for exclusion of rheumatoid disorders as a cause of hip destruction and allowed us to establish the diagnosis of bilateral neurogenic hip arthropathy.

The patient underwent replacement of the right hip in February 2007 and left hip in August 2008. (Figure 4) In spite of antithrombotic medication, the postoperative course was complicated by marked lower limb swelling. Therefore, soon after the second hip replacement, she received Low Molecular Weight Heparins due to clinical presentation suggestive of venous thromboembolism (September 2008). Except for VTE, the follow-up was uneventful and did not reveal any additional hip pathology. There were no incidents of hip dislocation and radiographs taken at the last follow-up revealed normal implant positioning. At the present the patient reports only occasional pain. She is able to walk without aids, with slightly positive Trendelenburg sign on the left. There is only a slight decrease in hip muscle power (scored mostly 4 or 4+/5 in the Lovett score). Due to her conditions, the patient receives medication with Enarenal (enalaprilate), NSAIDs, namely Ketonal (ketoprofen), combined with Proton Pump Inhibitors (e.g. Omeprazole), Detralex (diosmin) and Tolperis (tolperisone).

DISCUSSION

Normal, well coordinated and efficient joint motion depends on good function of both the static stabilizers (which include articular surfaces, ligaments and joint capsule) and dynamic stabilizers, comprising the muscles acting across a given joint. The precise control of these stabilizers by the central nervous system requires sensory feedback, which is provided by various proprioceptors located in joint capsules, tendons, ligaments and skin. Some of them, such as Pacinian corpuscles, are stimulated throughout entire arc of motion and also detect changes in speed and direction of movement. Other receptors, such as Golgi organs and Ruffini corpuscles, are associated with sensing of the relative position of neighbouring parts of the body. Muscle spindles detect changes in muscle length, which is helpful in stabilizing a joint by contraction of muscles.

Disturbed proprioception caused by disrupted afferent nerve pathways combined with diminished superficial sensation, affects the joint's normal protective reflexes and leads to joint instability, degeneration and destruction. In this situation, hip disorder is secondary to neurologic problems. Therefore, it is called a neurogenic arthropathy (Styczyński *et al.* 2007). Clinical features of this condition include rapid progression of



Fig. 4. A plain radiograph of pelvis after bilateral total hip replacement.

degenerative changes and marked destruction of articular surfaces, with only mild to moderate pain (due to decreased superficial sensation (Martinet *et al.* 1999). Range of motion is surprisingly good contrary to the dramatic radiographical appearance of the affected hip (Thompson and Manive, 1988). On examination, the affected joint is usually unstable, one may also find mild tenderness and crepitations on movement. Progressive destruction of the articular surface, microfractures and increased bone desorption lead to multifocal osteochondral necrosis that result in multiple free intra-articular bodies that give the impression of “bag of bones” (Milgram *et al.* 2000).

The neurogenic arthropathy may have polyarticular manifestation but usually involves one or, less frequently, two joints. Tabetic neuropathy affects mostly knee and hip joints, whereas diabetic neuropathy results in destruction of the ankle joint and small articulations of the foot. Neuropathy related to syringomyelia affects joints of the upper extremity, i.e. elbow and shoulder.

The diagnosis of neurogenic arthropathy is based on establishing rapid progression of degenerative changes in the setting of diminished superficial and deep sensation caused by neurogenic disorders (Avimadje *et al.* 2000). Imaging studies are usually limited to plain radiography and perhaps computer tomography. (Figure 5) These studies usually reveal findings very similar to those found in primary osteoarthritis. However, joint destruction has faster progression and radiographic findings are out of proportion in relation to mild clinical symptoms (Batra *et al.* 2008).

The affected joints may develop infection, sometimes with only mild general symptoms (Unnanuntana and Waikakul, 2006). The differential diagnosis must include arthropathy caused by intraarticular steroid injections, avascular necrosis, as well as heterotopic calcifications (Orpen *et al.* 2003). The diminished sensation requires further work-up studies including neurologic opinion, as well as testing for infections such as syphilis or leprosy. Established diagnosis of diabetes mellitus does not justify lack of further testing, which may reveal other underlying causes, e.g. amyloidosis, injury or tumors of spine or peripheral nerves, and hereditary factors (Waguri-Nagaya *et al.* 2004).

Management of neurogenic arthropathy depends on localization of changes, degree of progression and the nature of the primary disorder. Of course, the underlying disorder must be treated regardless of the joint involvement. As for the affected joint(s), symptomatic treatment include periods of joint immobilization, partial weight-bearing and physical therapy. Surgical treatment does not restore the anatomy and usually involves removal of free intraarticular bodies, cheilotomy, and ultimately joint replacement (Robb *et al.* 1988). However, outcomes of joint replacement in patients with diabetic neuropathy may be worse than usual because of disturbed proprioception. There are studies that report an increased risk of complications following



Fig. 5. A computer tomography before surgery.

joint replacement, in particular hip dislocation and implant loosening (Baldini *et al.* 1985). Replacing the affected joint will not restore its function, unless the underlying neurologic disorder will be treated. It is still unclear why some sensory neuropathies are complicated by neurogenic arthropathies while others are not. Similarly, we do not know why different neurogenic disorders result in involvement of different joints. One possible theory is that in the presence of neuropathy a preexisting minor joint degeneration might rapidly progress into full arthropathy.

CONCLUSION

Charcot joints are relatively rare disorder, especially when compared to primary osteoarthritis. However, those rare cases may be difficult to diagnose and treat and often require multidisciplinary approach. Marked destruction of articular surfaces usually has to be addressed with joint replacement. However, this symptomatic management is often associated with high risk of complications unless the underlying disorders have been treated.

REFERENCES

- 1 Avimadje AM, Pellieux S, Goupille P, Zerkak D, Valat JP, Fouquet B (2000). Destructive hip disease complicating traumatic paraplegia. *Joint Bone Spine.* **67**: 334–6.
- 2 Baldini N, Sudanese A, Toni A (1985). Total prosthetic replacement in tabetic arthropathy of the hip joint. *Ital J Orthop Traumatol.* **11**: 193–7.
- 3 Batra S, Batra M, McMurtrie A, Sinha A (2008). Rapidly destructive osteoarthritis of the hip joint: a case series. *J Orthop Surg.* **3**: 3.

- 4 Flik K, Vargas JH 3rd. (2000). Rapidly destructive hip disease: a case report and review of the literature. *Am J Orthop.* **29**: 549–52.
- 5 Hasegawa Y, Matsuda T, Iwase T, Iwata H (1997). Rapidly destructive arthropathy of the hip in siblings. *Clin Orthop Relat Res.* **336**: 152–5.
- 6 Martinet P, M'Bappé P, Lebreton C, Heinzleff O, Sibony M, Papeix C, *et al.* (1999). Neuropathic arthropathy: a forgotten diagnosis? Two recent cases involving the hip. *Rev Rhum Engl Ed.* **66**: 284–7.
- 7 Milgram J, Jimenez M, Rao A (2000). Femoral head disintegration in a paraplegic patient: loose bodies in neuropathic joints. *Spinal Cord.* **38**: 112–6.
- 8 O'Connor BL, Palmoski MJ, Brandt KD (1985). Neurogenic acceleration of degenerative joint lesions. *J Bone Joint Surg Am.* **67**: 562–72.
- 9 Orpen N, Walker G, Fairlie N, Coghill S, Birch N (2003). Avascular necrosis of the femoral head after surgery for lumbar spinal stenosis. *Spine (Phila Pa 1976).* **28**: E364–7.
- 10 Robb JE, Rymaszewski LA, Reeves BF, Lacey CJ (1988). Total hip replacement in a Charcot joint: brief report. *J Bone Joint Surg Br.* **70**: 489.
- 11 Sprenger TR, Foley CJ (1982). Hip Replacement In Charcot Joint. *Clin Orthop Relat Research.* **165**: 191–4.
- 12 Styczyński T, Gasik R, Pyskło B (2007). Proprioception's clinical validity for the motor system. *Reumatologia* **45**: 404–6.
- 13 Thompson RC Jr, Manivel C (1988). Neuropathic arthropathy as a possible cause of failure of a whole joint allograft. A case report. *Clin Orthop Relat Res.* **234**: 124–8.
- 14 Unnanuntana A, Waikukul S (2006). Neuropathic arthropathy of the elbow: a report of two cases. *J Med Assoc Thai.* **89**: 533–40.
- 15 Waguri-Nagaya Y, Mizutani J, Kobayashi M, Otsuka T, Matsui N (2004). Neuropathic arthropathy caused by chondrosarcoma of the cervical spine. *Mod Rheumatol.* **14**: 160–3.