Acta Orthop. Belg., 2017, **84**, 73-77

ORIGINAL STUDY

Spontaneous posterior shoulder dislocation as the first symptom of a Charcot arthropathy

Jacques Hernigou, Sofiane Boulares, Olivier Delahaut

From the CHU Charleroi, A. Vésale hospital - Orthopaedic and Traumatology Surgery, Montigny-le-Tilleul

Neuroarthropathy or Charcot disease is a destructive joint pathology. Upper limb involvement is unusual and often due to syringomyelia.
We reported a patient with spontaneous posterior right shoulder dislocation. Thereafter, she presented a quick joint destruction evoking a Charcot shoulder disease after excluding infectious and rheumatologic diseases. Explorations of this Charcot disease lead to the discovery and treatment of syringomyelia. A conservative management of her shoulder neuro-arthropathy has been proposed to the patient.
Shoulder involvement in Charcot disease is unusual: only 5% of cases. 18 cases have been reported in English literature. Two theories try to explain Charcot disease: neuro-vascular and neuro-traumatic. There is no efficient treatment for Charcot shoulder disease but it can be prevented by surgical treatment of syringomyelia.
Charcot shoulder is a rare and morbid disease for which conservative treatment is recommended because of disappointing results of arthroplasty and arthrodesis.

Keywords: Charcot disease; shoulder; arthrolysis; Arnold Chiari; syringomyelia.

INTRODUCTION

Neuro-arthropathy is a joint destruction disease due to neurological deficit. Charcot has described first it in 1868. Neurological damage of proprioception and pain-thermal sensory would be responsible of the articular destruction. Diabetes, tabes dorsalis and alcoholism are usually involved for lower limb while syringomyelia is the most involved for upper limb *(5,15)*. This disease is relatively unknown and rarely described at the upper limb. We reported the case of a patient with a right spontaneous posterior shoulder dislocation as the first sign of a Charcot disease due to the emergence of a syringomyelia.

CASE

A 48 years old patient presented for a diminution of range of motion of her right shoulder for 15 days. Her antecedents are: diaphysis humeral fracture with radial nerve injury and persistent paresis at 2/5; weaned smoking; C5-C6 posterior arthrodesis; type I Arnold Chiari disease without syringomyelia. She is also complaining of "crunches", that appeared recently, during the mobilization of her shoulder. There are no traumatisms or seizures reported. Clinical examination revealed a deformed shoulder

No benefits or funds were received in support of this study.
The authors report no conflict of interests.

■ Jacques Hernigou, MD.
■ Sofiane Boulares, MD.
■ Olivier Delahaut, MD.
CHU Charleroi, A. Vésale hospital - Orthopaedic and traumatology surgery.
Correspondence : J. Hernigou, Avenue de l'Université, 88 1050 Ixelles, Belgium. Tel.: 0033661169481
E-mail : Jacques.hernigou@gmail.com

A new surgery is performed for equipment and bone sequestration ablation. Biology was normal (no inflammation) and bacterial tests were negative (multiple samples in surgery room). Follow-up is marked by an increase of shoulder destruction on radiographs (Fig. 2). Rheumatologic consultation has eliminated an autoimmune or inflammatory pathology.

We performed a cerebral MRI and an MRI of the cervical spine. MRIs reveal an aggravation of her Arnold Chiari disease and apparition of a multi level syringomyelia from C1 to Th1 (Fig. 3).

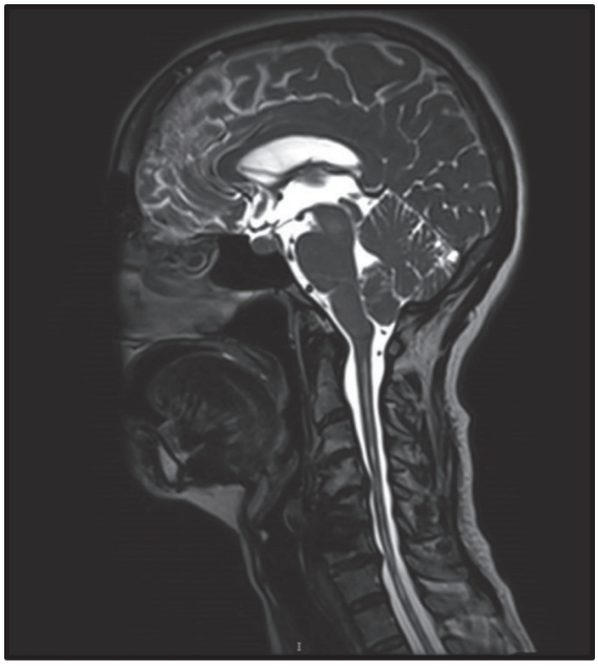

Fig. 3. — Cervical spine T2 MRI with syringomyelia C2-T1

Quick shoulder destruction, negative infectious and rheumatologic explorations, and apparition of a syringomyelia are suggestive for right neuro-arthropathy shoulder or Charcot disease. A conservative management of the shoulder has been proposed to the patient and she recently has been operated of her syringomyelia.

DISCUSSION

Incidence of this pathology is rare and difficult to estimate. According to Butala, 5% of Charcot disease involved shoulder *(2)*. In his series of patients with syringomyelia due to Arnold Chiari disease (795 patients in 9 years), Deng observed only 11 Charcot diseases with 3 to the shoulder. He also found that Charcot disease due to syringomyelia represents only 0.66% of arthropathy *(5)*. Hatzis reported 6 cases between 1969 and 1997 *(9)*. 18 "case-report" have been reported in English literature between 1992 and 2015 (Table I). 11 women and 7 men involved. Mean age of diagnosis was 49 years (min 36 - max 62). 76% of the patient had only one joint achieved. 88% of patients presented a syringomyelia and for 71% of them, syringomyelia was unknown before neuro-arthropathy of the shoulder. At least 35% of patients were operated of their syringomyelia. 50% of the patients had a conservative management of their Charcot disease, 17% of them had an arthrodesis. For others patient, treatment of neuro-arthropathy was not reported *(1-15)*.

In his series, Deng found that neurological symptoms were often unnoticed. In almost all cases, neuro-arthropathy was the motivation of consultations. Then, explorations for the neuro-arthropathy revealed neurological deficiency and syringomyelia *(5)*. We recommend a cervical spine MRI for all arthropathies of upper limb (furthermore shoulders) to research a syringomyelia that would be responsible of 25% of Charcot diseases *(2)*.

Physiopathology of Charcot disease is still unappreciated and discussed. Initially two hypotheses were opposed: the French theory described by Charcot and Mitchell and the Dutch theory *(15)*. More recently two new theories have appeared: the neuro-traumatic one and the neuro-vascular one. In the first theory, neurological deficiency inhibits muscular reflexes that warrant articular stability. Increased articular range of motion to unusual values and repeated trauma, caused by lost of stability, lead to destruction of joint involved. In the second theory, sensitive neurological deficiency decreased neurological reflexes around articular joint that lead to hyperemia and increased osteoclastic activity. According to Yanik, these two mechanisms are both present in the pathogenesis but not at the same time. The neuro-vascular one would be the first one while the neuro-traumatic one would intervene secondary in joint destruction *(15)*.

and an increased volume with a limitation of the mobility: abduction and flexion 90°; external rotation 0°; internal rotation 90°. Patient has no pain.

Radiographs have revealed a posterior shoulder dislocation with a reverse Hill Sachs lesion (Fig. 1). Patient is operated in emergency for the reduction of her dislocation and a "remplissage procedure" of her reverse Hill Sachs lesion. During immediate postoperative follow-up, patient's shoulder is immobilized, elbow beside the body with an arm sling and she doesn't present any complication. Two months after surgery, she consults to emergencies for a right shoulder pain and increased volume. Radiographs reveal right shoulder destruction (Fig.2).

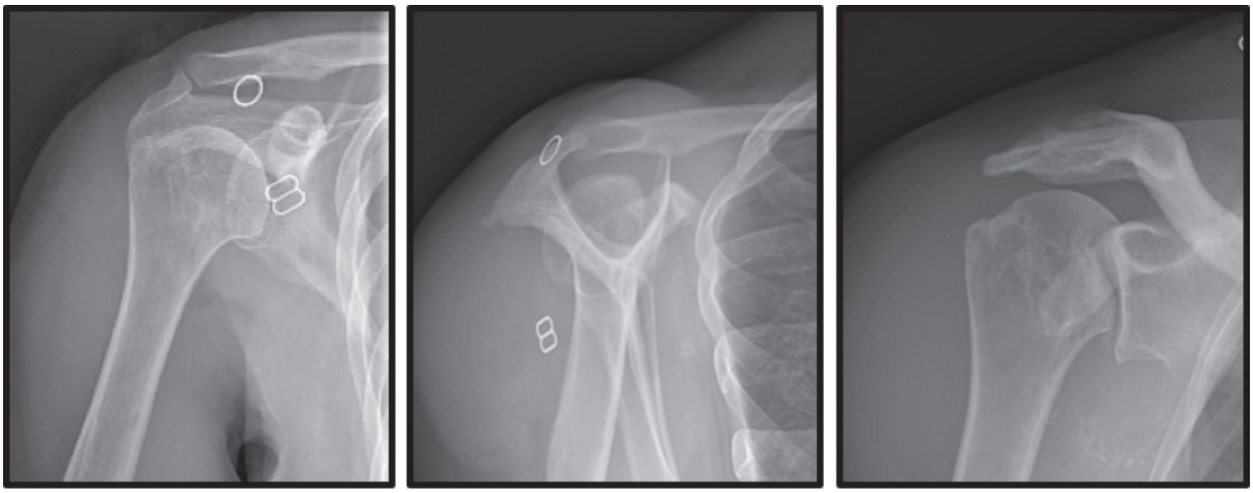

Fig. 1. — On the left: radiograph before posterior dislocation. On center and the right: right posterior shoulder dislocation

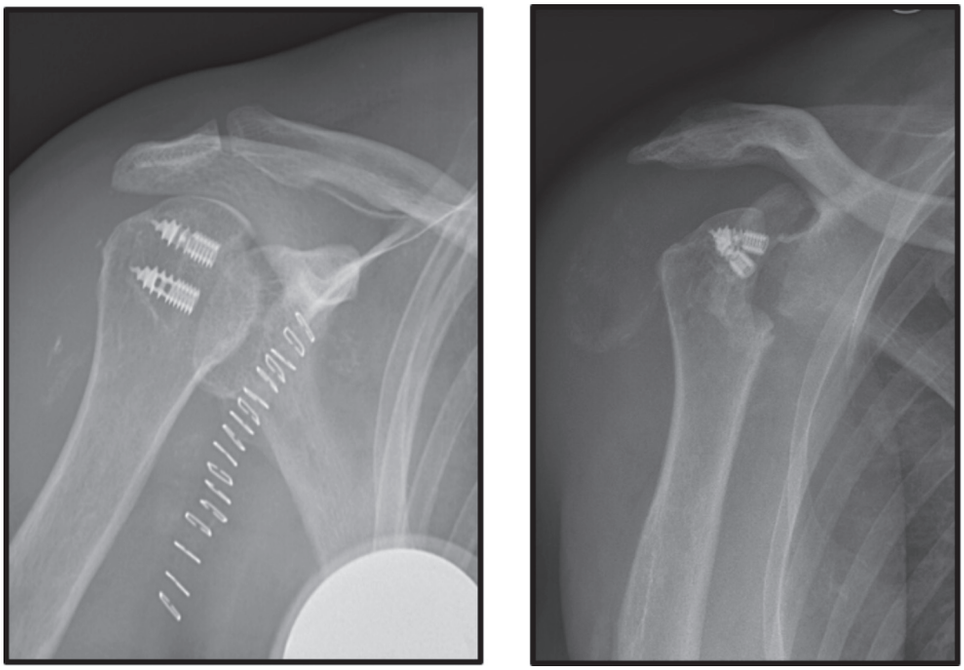

Fig. 2. — Quick shoulder joint destruction. On the left, two months after the first surgery and on the right after second surgery

Table I. — Shoulder Charcot disease in English litterature

Authors	Date	Age (y)	Sex	Begining of symptoms	Other joint involved	A. Chiari	Syringo-myelia	Syringo-myelia already known	Surgical treatment of syringo-myelia	Treatment of neuro-arthropathy
R.R. Richards [3]	1992	54	W	-	No	Type I	C1-T1	No	Yes	Conservative
R.R. Richards [3]	1992	51	W	-	No	No	C1-T1	No	-	Conservative
R.R. Richards [3]	1992	61	W	-	No	No	C2-T6	No	-	Conservative
Burcu Yanik [1]	2003	43	M	6 months	No	Type I	C1-T2	No	Yes	Arthrodesis
A.B. Cullen [13]	2005	36	M	3 months	No	-	-	-	Yes	-
Garg RK [4]	2008	42	M	-	No	Type I	C3-T12	No	-	Arthrodesis
B. Nacir [5]	2010	54	M	-	Elbow and wirst	Type I	C2-T4	No	-	-
G. Grahovac [6]	2011	62	W	6 months	No	Type I	C1-T5	No	Yes	Arthrodesis
R.B. Gaskins III [7]	2011	52	W	3 weeks	No	No	C2-T1	No	-	-
S. Kumar [8]	2011	38	W	4 years	No	Type I	C1-T5	No	-	Conservative
P. Ashok [9]	2012	62	M	-	Shoulder	No	C3-T3		-	Conservative
A. Alai [10]	2013	49	M	2 months	No	Type I	C1-T2	No	Yes	Conservative
X. Deng [2]	2013	48	W	2 years	Elbow and wirst	Type I	C1-T?	-	-	Conservative
X. Deng [2]	2013	57	W	-	No	-	-	-	-	-
X. Deng [2]	2013	44	W	-	No	-	-	-	Yes	-
H. Liu [11]	2014	44	M	1 year	No	Type I	C1-T4	No	-	-
R.R. Butala [12]	2014	53	W	3 years	Thumb	No	C2-C5	No	-	Conservative
P.P. Chakraborty [12]	2015	40	W	3 months	No	-	C2-T2	No	-	Conservative

M: Man; W: Woman

The Presentation of our patient is unusual because of her spontaneous posterior shoulder dislocation. That happens often during seizures or high-energy traumatism. None of them were identified in patient history. In Charcot shoulder disease, spontaneous posterior shoulder dislocation can be observed but at a late stage of joint destruction.

Treatment of Charcot shoulder disease is poor. Conservative management is recommended. Other treatments like total arthroplasty and arthrodesis allow disappointing results. These bad results would be due to neuro-traumatic and neuro vascular deficiency leading to increases implants stress and osteopenia causing peri-prosthetic fractures and

early loosening *(2)*.

However, according to Deng, treatment of pathology leading to neuropathy (surgical decompression of foramen magnum in Arnold Chiari Type I with syringomyelia) would allow to prevent Charcot disease due to syringomyelia *(5)*.

CONCLUSION

Charcot shoulder is a rare disease with high morbidity and no efficient treatment. Only the treatment of disease that leads to neurological deficiency allows limiting consequences of Charcot disease or preventing it.

REFERENCES

1. **Alai A, Reddy CG, Amrami KK, et al.** Charcot arthropathy of the shoulder associated with typical and atypical findings. Clin. Anat 2013; 26: 1017-1023.
2. **Butala RR, Arora M, Rao AA, et al.** A rare case of ipsilateral shoulder and thumb CMC joint neuropathic arthropathy. J. Surg. Case Reports 2014.
3. **Chakraborty PP, Datta S, Ray S, et al.** Unilateral neuropathic arthropathy of the shoulder secondary to syringomyelia: Diagnostic challenges. World J. Clin. cases 2015; 3: 1017-1020.
4. **Cullen AB, Ofluoglu O, Donthineni R.** Neuropathic arthropathy of the shoulder (Charcot shoulder). MedGenMed 2005; 7: 29.
5. **Deng X, Wu L, Yang C, et al.** Neuropathic arthropathy caused by syringomyelia. J. Neurosurg. Spine 2013; 18: 303-309.
6. **Garg RK, Kar AM.** Charcot shoulder in syringomyelia. Intern. Med. J 2008; 38: 868-869.
7. **Gaskins RB, Miller BJ, Scarborough MT.** Charcot arthropathy of shoulder: a case report. Orthop. Surg 2011; 3: 268-270.
8. **Grahovac G, Vilendecic M, Srdoc D.** Charcot shoulder caused by Chiari type i malformation with syringomyelia with six-year follow-up. Wien. Klin. Wochenschr 2011; 123: 512-514.
9. **Hatzis N, Kaar TK, Wirth MA, et al.** Neuropathic arthropathy of the shoulder. JBJS Am 1998; 80: 1314–1319.
10. **Kumar S, Sharma V, Kumar S, et al.** Imaging Findings in Chiari Malformation with Syringomyelia in a Case of Charcot Shoulder. J. Clin. Imaging Sci 2011; 1: 20-23.
11. **Liu H, Wang Y, Yang Z, et al.** A case report of Charcot arthropathy caused by syringomyelia and Chiari malformation complicated with scoliosis. BMC Res. Notes 2014; 7: 277-280.
12. **Nacir B, Arslan Cebeci S, Cetinkaya E, et al.** Neuropathic arthropathy progressing with multiple joint involvement in the upper extremity due to syringomyelia and type I Arnold-Chiari malformation. Rheumatol. Int 2010; 30: 979-983.
13. **Panagariya A, Sharma AK.** Bilateral Charcot arthropathy of shoulder secondary to syringomyelia: An unusual case report. Ann. Indian Acad. Neurol 2012; 15: 202-204.
14. **Richards RR, Delaney J.** Syringomyelia presenting as shoulder instability. J. Shoulder Elbow Surg 1992; 1: 155-161.
15. **Yanik B, Tuncer S, Seçkin B.** Neuropathic arthropathy caused by Arnold-Chiari malformation with syringomyelia. Rheumatol. Int 2004; 24: 238-241.