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Clinical case / Cas clinique

Kienböck's disease and cerebral palsy case report

Maladie de Kienbock et paralysie cérébrale à propos d'un cas

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Abstract

Introduction. – Pain is the main problem in patients suffering from cerebral palsy, particularly in adults. The upper limbs are affected in 25% of cases. Here, we report the case of a patient with Kienböck's disease.
Method. – Clinical case and literature review. A 28-year-old man suffering from dystonic quadriplegia consulted for progressively worsening pain in the right wrist. Kienböck's disease was diagnosed and conservative treatment with botulinum toxin in the flexor carpi radialis recommended. A good result was obtained with a decrease in pain. This result was still present two years later.
Discussion. – Although few references are made to it in literature, Kienböck's disease in cerebral palsy is probably underestimated. Maintenance of the wrist in a permanent flexed position and muscular hypertonia may be risk factors. Knowledge of this particular clinical picture will enable it to be detected promptly and thus enable conservative treatment to be organised with a maximum chance of therapeutic success, preventing the need for surgery.

Keywords: Cerebral palsy; Kienböck's disease; Pain; Botulinum toxin

Résumé

Introduction. – La douleur est le principal motif de consultation des personnes atteintes de paralysie cérébrale, notamment dans la population adulte. L'atteinte des membres supérieurs est retrouvée dans un quart des cas. Nous rapportons ici le cas d'un patient souffrant d'une ostéonécrose du lunatum connue sous le nom de maladie de Kienböck.
Méthode. – Présentation d'un cas clinique et revue de la littérature.
Observation. – Un patient de 28 ans présentant une tétraparésie dystonique consulte pour une douleur de poignet de rythme mécanique et d'aggravation progressive. Après le diagnostic de maladie de Kienböck de stade 2 de Lichtman, un traitement conservateur par immobilisation avec traitement associé de la spasticité du fléchisseur radial du carpe par toxine botulique permet d'obtenir une nette amélioration, avec diminution de la douleur de 80 % selon le patient. Le bénéfice thérapeutique persiste à deux ans, avec des injections itératives de toxine botulique. Sur le plan radiologique, aucune modification n'est observée à deux ans.
Discussion. – La maladie de Kienböck chez les personnes atteintes de paralysie cérébrale est peu décrite dans la littérature mais probablement sous-estimée. L'attitude permanente en flexion du poignet et l'hypertonie musculaire pourrait être des facteurs de risques. La connaissance de ce tableau clinique particulier doit permettre de l'évoquer précocement et de mettre ainsi en place un traitement conservateur avec un maximum de chance de succès thérapeutique et ainsi d'éviter un geste chirurgical.

Mots clés : Paralysie cérébrale ; Douleur ; Maladie de Kienböck ; Toxine botulique

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1. English version

Pain is the main health problem reported by adult sufferers of cerebral palsy. Localisation in the upper limbs is reported in 25% of cases, with a frequent context of overuse [2,4,15]. Kienböck's disease is an ischaemic necrosis of the lunate bone, posing the same etiological problem as other idiopathic necroses: none of the many explanations set forth in literature has yet provided definite proof of its value [11,13]. Among these, hyperflexion of the wrist resulting from spasticity of the flexors was detected in sufferers of neurological ailments [1]. Spasticity and dystonia are constants in cerebral palsy, the current term for cerebral motor disability; it could therefore be assumed that Kienböck's disease is frequently found in this context. Even if two series report rates of 2.7% and 10% in patients presenting with cerebral palsy [5,12], the number of cases described in literature remains extremely slight. Here, we will report on an observation of a young cerebral palsy sufferer. We will then compile another summary of the information provided in literature from this case.

1.1. Observation

M.D., a male of 28 years suffering from dystonic quadriplegia in the context of cerebral palsy, with bilateral flexion of the wrists from a clinical viewpoint in particular. He consulted with a progressive pain in his right wrist, with tumefaction of the carpus. M.D. is right-handed, entirely autonomous, with no previous asymmetry noted on the articular amplitudes of the wrists with a 40° bilateral extension limit. The pain developed after 15th May 2006, was strictly unilateral and became permanent leading to an X-ray in September 2006. This led to the diagnosis of Lichtman stage 2 osteonecrosis of the lunate bone, which would be confirmed by CT scan with a fragmentary aspect of the lunate bone (Figs. 1 and 2). Negative ulnar variance was also noted (Fig. 1) in the standard radiological assessment. A surgical procedure to shorten the radius was recommended on the therapeutic level due to the inequality of the length of the two bones in the forearm. However, fear of increasing the handicap led to conservative treatment being proposed in the first instance. An injection of botulinum toxin into the flexor carpi radialis was proposed in conjunction with immobilisation of the wrist using a splint. Toxin A was injected on 13th December: 100 U Botox in 1 ml via tracking by stimulation in the flexor carpi radialis. Distinct improvement was noted after 15 days, with a distinct decrease in pain which only persisted when extended, but M.D. no longer took analgesics. He did not complain of loss of strength nor loss of function. The injections were repeated every three months with persistence of the benefit; the splint was no longer worn continuously. After two years, M.D. no longer took analgesics. He estimated that there had been a 70% improvement and enjoyed normal functioning of the right wrist again. He wore his splint during his packaging job at a sheltered workshop. An X-ray showed images of osteonecrosis without lunate bone collapse.

1.2. Discussion

Kienböck's disease is a rare pathology which most frequently affects young men of between 20 and 40 years of age.

Pain is the main reason for consultation in sufferers of cerebral palsy [2,4,15]. The vertebral column and lower limbs are the most frequent locations. It is reported that the upper limbs are affected in 25% of patients [4,15]. The most frequently encountered problems are articular overwork syndromes, in particular in conjunction with the usage of walking sticks or wheelchairs, and pain related to spasticity and dystonia. Few studies have been devoted to the incidence of Kienböck's disease in cerebral palsy. This is 2.7% according to Joji et al. [5]. He actually found six cases in a systematic

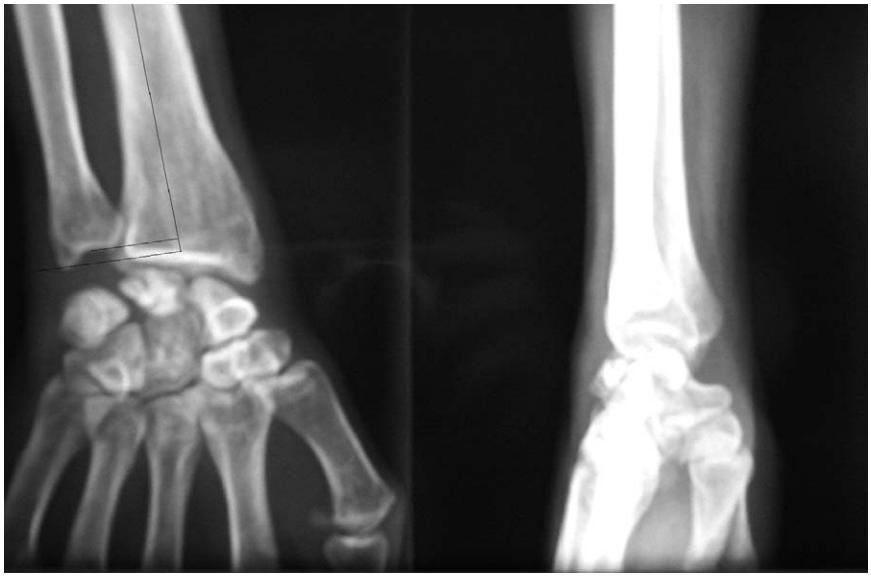

Fig. 1. Standard radiography, frontal and lateral view: osteonecrosis of the lunate bone, negative ulnar variance.