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Original article

Cervical laminectomy and micro resection of the posterior venous plexus in Hirayama disease

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ABSTRACT

Introduction. – Hirayama disease is a rare cervical myelopathy predominantly affecting young adults and mainly found in Asia. It results in a pure motor distal lesion of the upper limbs with slow progression. Dynamic magnetic resonance imaging (MRI), which allows the diagnosis to be made, shows a typical appearance of anterior compression of the cervical spinal cord associated with enlargement of the posterior epidural spaces due to a dilated venous plexus. Surgery is considered when conservative treatment has failed. However, the type of surgery is not well standardized in this compressive myelopathy.
Methods. – We report on three patients with Hirayama disease operated using an original method: cervical decompressive laminectomy and coagulation of the posterior epidural plexus without fixation. The clinical, radiological and surgical data of these three patients were analyzed. Each patient underwent postoperative MR imaging.
Results. – The mean age at diagnosis was 18.6 years (16–20 years) with a history of progressive symptoms lasting 1 to 4 years before treatment. Follow-up was 21 to 66 months after surgery. Neurological and electrophysiological improvement was noted in two patients; the third had stabilized. Postoperative MRI confirmed normalization of flexion imaging on MRI. None of the three patients complained of disabling neck pain.
Conclusion. – Posterior cervical decompression with coagulation of epidural venous plexus is a technique that seems effective in Hirayama disease in young subjects. It effectively treats patients by avoiding permanent cervical fixation.

1. Introduction

Hirayama disease was described in 1959 by a Japanese neurologist and is a well-known cause of lower cervical myelopathy in Asian countries [1]. A Japanese survey found a prevalence estimated at 1/30,000 people [2]. Several cases or small series of patients have been reported in Europe and America [3–7] but the incidence remains unknown in non-Asian countries. The disease preferentially affects young men between 15 and 20 years of age and is characterized by muscle weakness and atrophy in the distal portion of one or both upper limbs [8]. Installation is progressive and painless, without long tract signs. Presentation is most often unilateral at the beginning but bilateral involvement occurs during the progression of the disease in up to one third of patients [2]. It mainly affects the metameric territories of C7-T1. The brachioradial muscle, with C6 innervation, is classically spared, which results in a clinical presentation of oblique atrophy [9]. The course of the disease is characterized by gradual worsening of symptoms until stabilization occurs after several years of progression [8]. While it is a nonlethal disease, its clinical consequences can be extremely disabling for patients [8]. Several differential diagnoses among motor neuron diseases must be considered, such as amyotrophic lateral sclerosis (rare in young subjects), multifocal motor neuropathy and spinal muscular atrophy. Electroneuromyography studies (ENMG) can be helpful in supporting the diagnosis, since a low ulnar/medial compound muscle action potential (CMAP) ratio in the C7 to T1 territories is suggestive of Hirayama disease [10]. Magnetic resonance imaging (MRI) can reveal atypical signs such as cord atrophy or intramedullary hyperintensity on T2-weighted images [11]. Finally, the diagnosis

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is confirmed if flexion MRI shows spinal cord compression by posterior dilated venous plexus and forward shift of the dura [11].

Surgical treatment is generally considered for advanced disease [9]. The choice of surgical technique is, however, debated. We report three European cases of rapidly progressing Hirayama disease treated by an original technique consisting of laminectomy and microcoagulation of the posterior epidural venous plexus.

2. Materials and methods

We present a retrospective series of three patients treated in the same center for Hirayama disease. The diagnosis was confirmed after complete neurological assessment. All patients were operated by the same surgeon. Clinical follow-up was done by the senior author and two assistants qualified in the review of clinical and radiological data related to spinal surgery.

2.1. Surgical technique

The patient was positioned prone, attached to a Mayfield headrest in slight cervical flexion. After making an incision on the midline and detaching the muscles, cervical laminectomy was performed carefully and extended over the stenosed levels (defined by cervical dynamic flexion MRI). A large dilated venous plexus was found in all cases in the posterior epidural space and was gradually and gently coagulated, with progress being followed under the microscope, until the posterior part of the dura mater was fully exposed. Blood loss was closely monitored. Depending on the decompression quality, duraplasty was carried out in addition to laminectomy. Once the cervical decompression was completed, hemostasis was progressively achieved using hemostatic gauze and bipolar coagulation. No fixation was used and the incision was closed carefully.

2.2. Evaluation of outcomes

Clinical evaluation was performed through pre- and postoperative motor testing using the standard neurological classification of spinal cord injury (ASIA Score). Patients with a clinical improvement of one point on two or more metameres were considered as improved. At the last follow-up, cervical neck pain and disability were evaluated on a Visual Analog Scale (VAS) and the Neck Disability Index (NDI), respectively. Radiological evaluation focused on the quality of the spinal cord decompression based on the reappearance of cerebrospinal fluid (CSF) around the spinal cord in flexion MRI 1 year after surgery. We also looked for signs of instability on dynamic MRI sequences. Electrophysiological evaluation used the median/ulnar CMAP ratios and the sum of the ulnar and median CMAP between pre- and postoperative measurements.

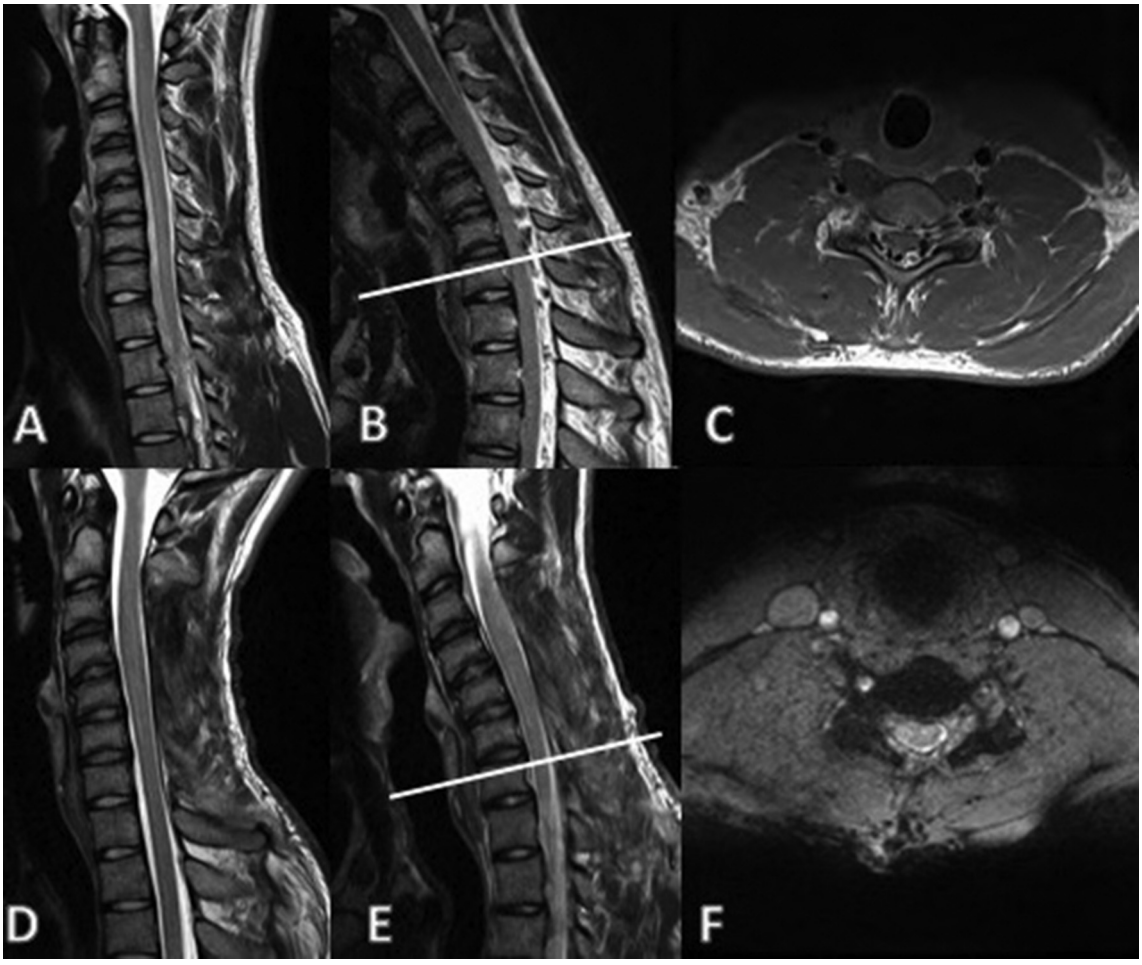

Fig. 1. A 16-year-old man complaining of bilateral upper limb motor deficit. On examination, he has C7-C8-T1 amyotrophy predominant on the right side. Spinal MRI does not show any obvious abnormality (A). Dynamic MRI reveals canal stenosis due to large dilated epidural plexus resulting in impingement of the spinal cord at C3 to C7 (B), which was confirmed by axial sequences (C). C3 to C7 laminectomy was performed resulting in almost complete neurological recovery. Four-month postoperative MRI confirms the reappearance of CSF in neutral flexion (D), and absence of cord stenosis in dynamic views (D). Axial sequences did not show any recurrence of dilated plexus (E).

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