▪ Headache Medicine 2022, Supplement ▪ p-ISSN 2178-7468, e-ISSN 2763-6178

Headache Medicine

▪ DOI: 10.48208/HeadacheMed.2022.Supplement.49

Spontaneous intracranial hypotension syndrome presented with sixth cranial nerve palsy: a case report

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Introduction

Spontaneous intracranial hypotension (SIH) is a secondary cause of headache often misdiagnosed and underdiagnosed disorder. The main presentation is orthostatic headache. An uncommon symptom is cranial nerve palsy. We described a case of SIH that presented to our service with sixth cranial nerve palsy.

Case Report

Female, 39 years old, no relevant medical past or history of trauma, awoke due intense holocranial headache, associated with vomiting and photophobia, worsening in orthostasis. In the general emergency department, she was release with prescription of analgesics and topiramate, without resolution. After a week, the patient reported diplopia and was referred to our service, a tertiary center. In admission, she kept an intense orthostatic headache and in neurologic examination she presented right sixth nerve palsy, no other deficits. Laboratorial tests were normal, including metabolic and infectious marks. Cerebrospinal fluid (CSF) opening pressure was low (50 mmH2O) and analysis showed elevated protein levels (158 mg/dl). MRI showed diffuse pachymeningeal enhancement and venous sinus distension, mainly in superior sagittal venous sinus. CT myelogram detected a spinal CSF leak in transition C1-C2. Initially, performed hydration and bed rest, while patient with partial improvement of headache. An epidural blood patch was then performed using 15 ml of autologous blood with compatible contrast and lidocaine. There was complete resolution of pain after the procedure and four weeks later complete resolution of right sixth nerve palsy.

Discussion

According to International Classification of Headaches Disorders, third edition (ICHD-3), the headache attributed to SIH is descripted as a headache has developed in temporal relation to low CSF pressure (<60 mm H2O) and/or evidence of CSF leakage on imaging.

A key aspect of SIH is that headache may not be orthostatic, although orthostatic headache is the most common symptom, present in 97.7% of cases, according to a systematic review with 1,694 patients. Another's symptoms are nausea/vomiting, neck pain, tinnitus and dizziness. In some cases, there are photophobia and other visual symptoms. Diplopia was described, in same review, in only 3.9% of cases. Less commonly reported, cranial nerve palsy was described in few cases. In our case, the patient showed an isolate sixth nerve palsy, and the probable mechanism of palsy is traction in nerve course, due to low CSF pressure.

The main findings of SIH in brain MRI are pachymeningeal enchantment, brain sagging, venous distension sign and pituitary engorgement. Spine imaging are important because the demonstration of a leak can confirm the diagnosis and may guide treatment. The CSF leaks are grouped into 3 mechanisms: meningeal diverticula, ventral dural tears and CSF-venous fistulas. CT myelogram is the golden standard for detection of CSF leaks, mainly in high-flow leaks. MRI is also used in detection of spinal leaks, but due use of intravenous contrast, CSF-venous fistulas are not found. Other options are dynamic images studies using fluoroscopy and MRI with intrathecal gadolinium but are not accessible in daily practice.

Conservative treatment (hydration and rest) was effective in 28% of cases, while epidural blood patches resolved symptoms in 64% in first intervention. A review of 738 patients no showed serious adverse effect after epidural blood patch. In our case, the patient present complete improvement after blood patch.

Conclusion

We report a case with an atypical presentation of spontaneous intracranial hypotension, with diplopia and sixth cranial nerve palsy. Furthermore, our case illustrates the effectiveness of blood patch treatment for SIH, a reversible cause of secondary headache.

Keywords: CSF leak, Epidural blood patch, Spontaneous intracranial hypotension.