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Spontaneous Intracranial Hypotension without Orthostatic Headache

Ortostatik Baş Ağrısı Olmaksızın Spontan İntrakraniyal Hipotansiyon

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ÖZET

Altıncı sinir parezisi ile başvuran, ortostatik baş ağrısı tanımlamayan ve spontan intrakraniyal hipotansiyon saptanan 2 hastada, altıncı sinir parezisi tedavisiz düzelirken, manyetik rezonans görüntülemelerinde subdural hematom saptanmıştır. Ortostatik baş ağrısının olmaması ve altıncı sinir parezisi düzelirken, subdural hematom gelişmesi gibi özellikleri nedeniyle olgular bildirilmeye değer bulunmuştur.

Anahtar Kelimeler: İntrakraniyal hipotansiyon, spontan, baş ağrısı, altıncı sinir parezisi, subdural, hematom.

ABSTRACT

Spontaneous Intracranial Hypotension without Orthostatic Headache

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We report 2 cases of spontaneous intracranial hypotension that presented with unilateral abducens nerve palsy, without orthostatic headache. While sixth nerve palsies improved without any intervention, subdural hematoma was detected with magnetic resonance imaging. We conclude that headache may be absent in spontaneous intracranial hypotension and spontaneous improvement of sixth nerve palsy can occur, even after the development of a subdural hematoma.

Key Words: Intracranial hypotension, spontaneous, headache, abducens nerve, subdural, hematoma.

INTRODUCTION

Spontaneous intracranial hypotension (SIH) is characterized by postural headache and low opening pressure at the lumbar puncture, without an obvious cause. The common clinical features of SIH are orthostatic headache, neck or interscapular pain, nausea, emesis, change in hearing, facial numbness, and upper limb radicular symptoms. Headaches, although typically orthostatic, may occur when seated or recumbent and evolve into chronic daily headaches (1-3). The non-orthostatic feature or absence of headache is very rare (4-6).

Neuro-visual findings in SIH may include diplopia due to sixth nerve paresis, transient visual obscurations, blurred vision, visual field defects, photophobia, and nystagmus (7). Cranial magnetic resonance imaging (MRI) abnormalities include diffuse pachymeningeal gadolinium enhancement, evidence of descent of the brain, decrease in the size of the ventricles, engorgement of cerebral venous sinuses, and enlargement of the pituitary gland. Spine MRI may show extra arachnoid fluid, meningeal diverticula, meningeal enhancement, or engorgement of the epidural venous plexus. MRI may also define the level or site of the leak (8). Most patients respond well to supportive therapy that includes increased fluid intake, bed rest, and oral or intravenous (IV) caffeine intake. An epidural patch and, rarely, surgical intervention may be used in refractory cases. Berroit et al. suggest early use of an epidural patch, without lumbar puncture, for a majority of patients with SIH and typical orthostatic headache (9). A blood patch works by sealing holes in the dura from the epidural side, leading to restoration of normal subarachnoid pressure.

Small subdural collections without mass effect can be seen with cranial MRI. Although this condition is thought to be benign, acute deterioration of a patient's clinical status may occur secondary to a large subdural hematoma (SDH), requiring urgent neurosurgical intervention (10). The cause of such hematomas is presumably rupturing of the bridging veins as the cerebrospinal fluid (CSF) volume decreases and the brain sags, pulling away from the dura (8).

CASES

Case 1

A 42-year-old female presented with diplopia on right gaze on November 11, 2001. She complained of a mild, intermittent headache with decreasing intensity for the past 15 days, which was not associated with body position. On examination she had a complete abduction deficit on the right side, but no other abnormal findings. Cranial MRI revealed diffuse meningeal enhancement and a subtle subdural collection in the left frontotemporal area (Figure 1A). The patient was investigated for causes of meningeal infiltration-namely sarcoidosis, carcinoma, or infection. No abnormality was observed in her hemogram or blood chemistry. ACE was normal. CSF opening pressure was not recorded. There were 20 cells in the CSF, all lymphocytes; protein was 101 mg/dL, glucose was 52 mg/dL, and polymerase chain reaction (PCR) for tuberculosis and serology for Borrelia were negative. No treatment was given. The patient began to improve within 1 month. She was asymptomatic and had full ductions during follow-up MRI on May 30, 2002, which showed expansion of the left frontotemporal SDH, in addition to diffuse meningeal enhancement (Figure 1B). Surgery was performed and the patient was in good health at the time of her last follow-up examination in May 2005. The last computerized tomography (CT) performed in July 2002 was normal.

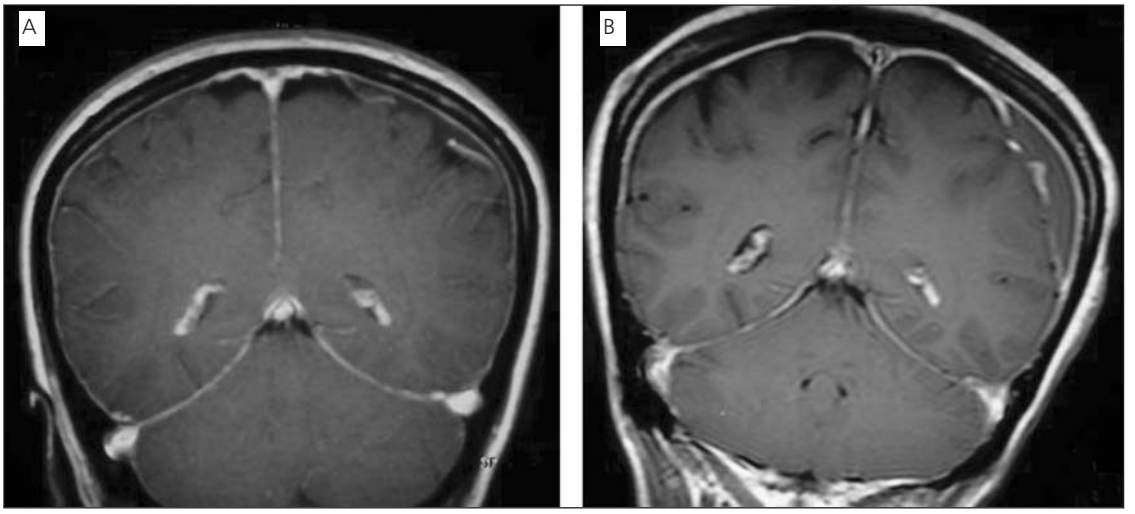

Figure 1. (Case 1) **A.** Diffuse meningeal enhancement (November 2001). **B.** Left frontotemporal subdural hematoma (May 2002).

Case 2

A 61-year-old male presented with double vision, nausea, and vomiting in August 2004. He had a history of mild neck pain without orthostatic features for 1 week. He also suffered a minor head injury 3 months prior to the onset of his symptoms. Past medical history included prostatectomy for benign prostate hypertrophy. Neurological examination revealed a complete abduction deficit on the right side, but no other abnormal findings. MRI showed diffuse meningeal enhancement (Figure 2A). Meningeal infiltration or intracranial hypotension was considered in the differential diagnosis. CSF opening pressure was 60 mmH_2O. Protein was 183 mg/dL and there were no cells in the CSF. PCR for tuberculosis was negative. CBC, ESR, ACE, and tumor markers were within normal limits. Blood chemistry did not reveal any abnormality, except high levels of cholesterol, triglycerides, and LDL. His diplopia improved and he had full motion in the eye 1 month after the onset of symptoms. MRI performed in October 2004 revealed meningeal enhancement and a small bilateral subdural collection (Figure 2B). Cervical, thoracic, and lumbar spinal MRI failed to reveal a CSF leak. The patient was put on bed rest and analgesic with caffeine was given. One month later there was an increase in the subdural collection (Figure 2C). He underwent surgery on December 14, 2004 for bilateral SDH. MRI revealed complete resolution of SDH and he was asymptomatic at his last follow-up in September 2005 (Figure 2D).

DISCUSSION

The 2 presented cases had diplopia due to unilateral abducens nerve palsy. Cranial MRI demonstrated diffuse pachymeningeal enhancement with gadolinium, leading to the diagnosis of spontaneous intracranial hypotension. CSF opening pressure was low in case 2, but was not recorded in case 1. While sixth nerve palsy improved without any intervention, follow-up MRI revealed the development of SDH. Surgical treatment was performed in both patients. During follow-up periods of 9 months in patient 2 and 3.5 years in patient 1, the patients remained symptom free.

Headache was not a prominent symptom in either patient during the course of the disease. The diagnostic criteria for intracranial hypotension, according to The International Headache Society, include headache that worsens within 15 minutes after sitting or standing, and imp-

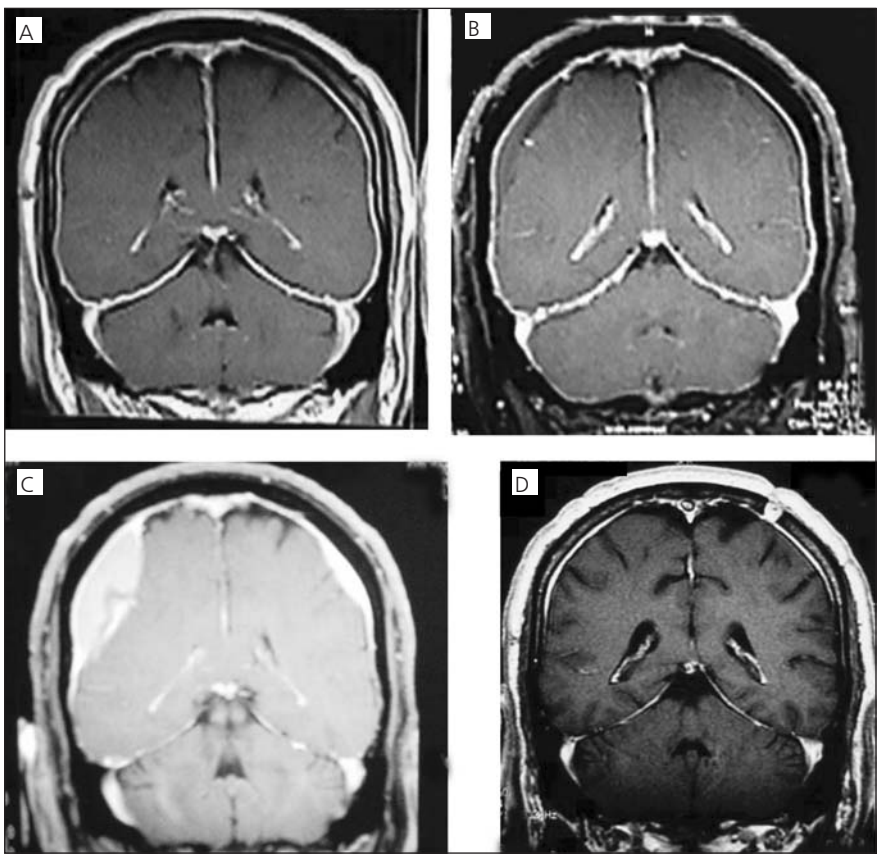

Figure 2. (Case 2) **A.** Diffuse meningeal enhancement (August 2004). **B.** Small bilateral subdural collection (October 2004). **C.** Increase in subdural collection (December 2004). **D.** Complete resolution of the subdural hematoma (April 2005).

rovement within 15 minutes after lying down (11). There are few reports describing non-orthostatic features or the absence of headache in patients with SIH (4,5). Mokri et al. reported 3 patients with intracranial hypotension-2 with overdraining CSF shunts and 1 with a proven CSF leak, but without headache. They suggested that a balance may occur between decreased CSF volume and buoyancy of the brain, and that the brain may not sink enough to produce headaches (6).

Another interesting feature of the presented cases is the improvement in sixth nerve palsy after the development of SDH, which has not been previously documented. It is possible that the subdural blood created a mass effect sufficient to restore intracranial pressure, but progression of SDH and enhancement of meninges would suggest that ICP was still low. It is also possible that SDH had expanded before the pressure had spontaneously normalized, in which case the improvement of sixth nerve palsy was simply a naturally occurring event, rather than an effect of SDH. As we didn't know where the CSF leaks were located, we also considered the possibility that coexisting subdurals covered the leaks and that SDH may have acted as a subdural patche in our patients. Although it is unlikely that subdural blood in the cranium could find its way into the spinal subdural space, there are 2 case reports that support this view-1 had cranial SDH and the other had a ruptured intracranial aneurysm associated with spinal SDH (12,13). It is suggested that spinal SDH may be related to redistribution of blood from the supratentorial subdural space.

In conclusion, diplopia may be the only presenting symptom in SIH and orthostatic headache may not be a part of the clinical picture. Spontaneous improvement of sixth nerve palsy can occur, even after the development of SDH.

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