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CLINICAL VIGNETTE

Missed Diagnosis: A case of Spontaneous intracranial hypotension presenting as headache

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Case Report

A 43-year-old female physical therapist was seen in the emergency department (ED) with a 1-day history of severe headache. She described the headache as bilateral and constant. A head CT and lumbar puncture were performed which were unremarkable and she was reassured and sent home on pain medications. The patient returned 2 days later to the same ED with persistent symptoms unrelieved with pain medications but now with associated dizziness. She described the headache as constant, with radiation of the pain from the upper neck to the crown of her head, spreading over her head. The pain was less severe when she is lying down, but worsened upon sitting or standing. She described nausea and vomiting. Her pain did not increase with urination, defecation, or valsalva. The patient was admitted, started on narcotic analgesics and antiemetics. The patient's symptoms intensified and she complained of nausea, vomiting, and photophobia. Her physical examination was unremarkable. Orthostatic vitals revealed mild orthostasis. A lumbar puncture was performed without an opening pressure, demonstrated clear colorless CSF without RBCs or WBCs, protein = 64, glucose = 27. A brain MRI demonstrated a 3 by 4 mm left cavernous carotid aneurysm. Neurosurgery recommended conservative management of her aneurysm. The patient also had a MRI of her cervical and lumbar spines which demonstrated a CSF leak seen on the left side from L1 to L3. An epidural blood patch was performed and the patient's symptoms completely resolved within 2 days.

Discussion

Spontaneous intracranial hypotension (SIH)

syndrome is usually associated with recent lumbar puncture; trauma; meningeal dysplasia; spontaneous leakage from 1 or more spinal nerve root sleeves, particularly in the thoracic and lumbar areas; and valsalva maneuver during excessive weightlifting¹. SIH syndrome was first described in 1938 by Schaltenbrand², who considered three causes: CSF leak, reduced CSF production, and increased CSF absorption. Orthostatic headache that improves rapidly in the recumbent position is a characteristic of SIH syndrome. Most patients have a benign course, but some patients can have serious complications like subdural hematoma due to tearing of bridging veins³.

The headache is a consequence of the low CSF pressure producing displacement of pain-sensitive structures. Associated symptoms, including tinnitus and vertigo, and subdural fluid collections are presumably from hydrostatic changes among intracranial fluid compartments that occur at low CSF pressures⁴. Other symptoms and physical findings may include horizontal diplopia related to unilateral sixth nerve palsy, postural posterior neck pain, change in hearing, transient right facial numbness and weakness, and transient right upper-limb radicular symptoms in C-5 and C-6 root distributions⁵.

The spinal fluid is usually clear, but some of the lumbar punctures show xanthochromic fluid and CSF protein elevation. A primarily lymphocytic pleocytosis occurs in some. CSF opening pressure is low but not in all patients. Some patients with symptomatic CSF leaks may have CSF opening pressures that are consistently within normal limits⁵.

MRI images may show a descended brain, taking the start of the sylvan aqueduct and the location of the cerebellar amygdalae as points of reference; diminished size of the subarachnoidal cisterns and occasionally of the cerebral ventricles; meningeal enhancement from increased uptake of the contrast solution; subdural hygromas and hematomas; and pituitary enlargement. Para spinal fluid and dilated epidural veins may be observed⁶.

Patients with SIH generally respond favorably to conservative management⁷. In a study involving 90 patients, all patients were treated by nonsurgical conservative management, such as absolute bed rest, intravenous hydration and repetitive epidural blood patch (5 patients). The mean duration of follow up was 51.4 months (range, 15-80 months). Among 13 patients included in this study, only one patient developed recurrent SIH, and the other patients improved from orthostatic headache. Although 7 of 13 patients had complete resolution of headache at a minimum of 2 years follow-up, 4 patients had mild headache and 2 patients continued to have moderate headache at the final examination⁸. It is critical that periodic follow-up examinations be performed and a more effective treatment modality developed to achieve complete resolution of SIH⁸. Early diagnosis of SIH correlates with better outcome, further suggesting that patients with a new headache that may worsen on standing or sitting should undergo MRI with contrast to expedite a possible SIH diagnosis, even if the pain is relatively mild⁹.

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