Surgical treatment of spontaneous intracranial hypotension associated with a spinal arachnoid diverticulum

Case report

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The authors present the case of a 22-year-old woman who developed spontaneous intracranial hypotension as a result of a cerebrospinal fluid leak from a thoracic extradural arachnoid diverticulum. The patient was successfully treated by ligation of the diverticulum. The entity of spontaneous intracranial hypotension and its relevance to neurosurgery are discussed.

KEY WORDS • headache • intracranial hypotension • intracranial pressure • spinal arachnoid cyst

Postural headache secondary to low intracranial pressure is a well-known entity in neurosurgical practice. It is most commonly encountered following lumbar puncture for cerebrospinal fluid (CSF) examination or myelography. Postural headache may also be experienced following craniotomy, spinal surgery, or cranioplastic trauma when the dura has been violated, in association with oto- or rhinorrhea, or as a result of CSF overdrainage in patients with ventriculoperitoneal shunts. Uncommonly, it occurs as a consequence of medical conditions such as severe dehydration, diabetic coma, or uremia. In some cases no precipitating event is apparent and the intracranial hypotension is believed to have developed spontaneously. The syndrome of spontaneous intracranial hypotension was first described in 1938 by the German neurosurgeon Schaltenbrand.23

The underlying cause of spontaneous intracranial hypotension is rarely established and treatment is therefore usually nonspecific. In the present paper, we describe a patient with spontaneous intracranial hypotension due to a CSF leak from a spinal arachnoid diverticulum; the condition was successfully treated by ligation of the diverticulum.

Case Report

This 22-year-old woman presented with a 6-week history of persistent generalized headaches that had developed over a 12-hour period. Typically, the headache would start shortly after she assumed the erect position but she felt perfectly well lying down, confining her to bed. The headache was usually accompanied by nausea and vomiting. Valsalva maneuvers exacerbated the headache. She felt somewhat better during the 3-day period of her menses. Oral and parenteral analgesic agents and minor tranquilizers did not provide relief. She had noted mild discomfort in the right lower thoracic region since the onset of the headaches. There was no history of trauma, cutaneous eruption, diabetes, infection, or easy bruising. Her medical history was significant for previous episodes of common migraine and corrective surgery for retrusion. The family history was unremarkable.

Examination. The patient was of a tall and lanky build (height 176 cm, weight 50 kg). A high-arched palate, thin skin, and increased laxity of the finger joints were noted. The remainder of the general physical and neurological examination was normal, as were the results of a detailed ophthalmological examination and echocardiography.

A computerized tomography (CT) scan of the head was normal. Lumbar puncture in the lateral decubitus position revealed an opening pressure of 40 mm H₂O. On CSF examination, the cell count and glucose and protein levels were normal. Gram stain revealed no organisms, and bacterial and fungal cultures were nega-
Spontaneous intracranial hypotension

FIG. 1. Radionucleotide cisternograms at 24 hours after injection showing a focus of increased activity of tracer in the right lower thoracic region (curved arrows). A marker is seen at the L-3 level (straight arrow) in one panel (left).

FIG. 2. Postmyelography computerized tomography scan showing the presence of a large right extradural diverticulum extending through the nerve root foramen at the T-11 level (arrow). The density of contrast material within the diverticulum is not that of the intrathecal space, suggesting a slow filling rate.

Radionucleotide cisternography showed early accumulation within the bladder, less than expected activity over the convexities, as well as a focus of increased activity of tracer in the right lower thoracic region (Fig. 1). Myelography with subsequent CT disclosed the presence of a large right T-11 extradural arachnoid diverticulum extending through the nerve root foramen corresponding to the location of the abnormal radionucleotide activity (Fig. 2). In light of the patient's age, the unrelenting and debilitating nature of the postural headaches, and the well-demonstrated radiographic abnormalities, we recommended surgical intervention.

Operation. A right T11–12 hemilaminectomy was performed; upon removal of the ligamentum flavum, a thin arachnoid diverticulum was encountered extending laterally with a broad-based neck originating at the common dural sac. There was obvious disruption of the dura at this level through which the diverticulum appeared to have herniated. The T-11 nerve root was found to be enveloped by the arachnoid diverticulum. The fragility and broad-based neck of the diverticulum precluded ligation, and the neck was clipped with a straight Sugita aneurysm clip. The diverticulum was then aspirated with a syringe and there was no evidence of ongoing CSF leakage. Vascular clips were placed across the nerve root on either side of the diverticulum, which was then resected. Pathological examination revealed that the wall of the diverticulum consisted of arachnoid.

Postoperative Course. The patient recovered and was ambulatory on the 2nd day postoperatively. She has remained free of headaches during the 1-year period of follow-up review.

Discussion

Characteristics and Diagnosis of Low CSF Pressure Headache

The International Headache Society defines low CSF pressure headache as one that "occurs or worsens less than 15 minutes after assuming the upright position, and disappears or improves less than 30 minutes after resuming the recumbent position." By definition, spontaneous intracranial hypotension is not associated with any significant trauma; an innocuous precipitating factor, such as sneezing, coitus, or a minor fall, however, is frequently reported. Women are more commonly affected than men and the majority of patients are in the third or fourth decade of life. The onset of headache may be acute or gradual. Associated symptoms are common and include nausea, vomiting, tinnitus, vertigo, photophobia, and phonophobia. In approximately 10% of cases, subdural hematomas or hygromas have been detected. These subdural fluid collections are not considered to be the cause of the orthostatic headache but are more likely the result of rupture of bridging veins due to the decrease in CSF volume and downward displacement of the brain. Similarly, subdural hematomas have been described following lumbar puncture and myelography.

The diagnosis of a low-pressure headache is confirmed by a lumbar puncture in the lateral decubitus position demonstrating a CSF pressure of less than 60 mm H2O. Not infrequently, a "dry tap" is initially encountered and CSF can only be obtained with a Valsalva maneuver, raising the patient's head, or with gentle aspiration using a syringe. A sucking noise has occasionally been described as the stylet is withdrawn and air apparently enters the subarachnoid space, indicating subatmospheric pressure. This course of events may result in spontaneous pneumocephalus. On examination, CSF is often normal but may show mild elevation of total protein content or an increased cell count. These CSF abnormalities may be due to meningeal hyperemia resulting from the low CSF pressure. Interestingly, several cases of diffuse meningeal enhancement on magnetic resonance (MR) imaging have recently been described in low CSF pressure states.
Pathogenesis of Spontaneous Intracranial Hypotension

In 1938, Schaltenbrand proposed three mechanisms by which spontaneous intracranial hypotension may be explained: 1) diminished CSF production; 2) CSF hyperabsorption; and 3) CSF leakage. Although decreased CSF production is an attractive hypothesis, at the present time CSF production is not measured directly and has not been established as a cause of spontaneous intracranial hypotension. The introduction of radionuclide cisternography has shed some light on the pathogenesis of spontaneous intracranial hypotension. In most cases the radionuclide scan has characteristically shown early accumulation of radionuclide in the bladder, indicating a rapid uptake of the tracer in the bloodstream. This abnormal pattern, however, does not necessarily differentiate between a CSF leak and CSF hyperabsorption. In some cases a leak along the spinal axis has clearly been identified, while in others it has not, either because of rapid absorption of the tracer or because the leak is not contained and/or resolution of the scan is not adequate to demonstrate leakage.

The underlying defect causing a CSF leak usually remains obscure. Although small tears in nerve root sleeves or spinal epidural or Tarlov's cysts have frequently been mentioned as the likely cause of spontaneous intracranial hypotension, an anatomical substrate has only rarely been identified. Nosik reported a patient in whom myelography revealed leakage of contrast material through a nerve root tear. Rando and Fishman described a case with dilated nerve root sleeves on MR examination at the level of a CSF leak demonstrated by radionuclide cisternography. In a second patient studied with myelography, these same authors demonstrated leakage of contrast material into a contained spinal extradural space, believed to represent an epidural cyst, but surgical exploration was not pursued. In our patient spontaneous intracranial hypotension was caused by a CSF leak arising from a fragile spinal extradural arachnoid diverticulum. Spinal extradural meningeal diverticula are uncommon lesions that are usually located at the mid to lower thoracic levels. Their etiology is unclear but it is possible that they are purely congenital in nature or that they result from herniation of the arachnoid through a dural defect, as was observed in our patient. These meningeal outpouchings may become so large that they expand into the thoracic cavity (lateral intrathoracic meningocele), abdomen, or pelvis. Dural ectasia and meningeal diverticula have been described in patients with heritable connective-tissue disorders, such as Marfan's syndrome, neurofibromatosis, Ehlers-Danlos syndrome. In such cases, it is likely that the underlying abnormality of the extracellular matrix resulted in a structural weakness of the spinal meninges, predisposing the patient to the development of meningeal diverticula.

In our patient a generalized connective-tissue disease was strongly suspected, but she did not fulfill the criteria of any of the known connective-tissue disorders. Interestingly, Stern and Raftopoulos described patients with marked dural ectasia associated with Marfan's syndrome who appear to have suffered from low CSF pressure headaches at some time during their clinical course.

Treatment of Spontaneous Intracranial Hypotension

Spontaneous intracranial hypotension is often a self-limiting disease, responding well to bed rest and a generous intake of oral or parenteral fluid and salt. In refractory cases, an epidural blood patch may be of benefit, even if a CSF leak has not been demonstrated. Other treatment modalities that may prove beneficial include a short course of corticosteroids or epidural or intrathecal saline infusion. If these conservative measures fail and a dural tear or other defect has been demonstrated to be the underlying cause of the CSF leak, surgical repair of the defect should be considered.

References


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738
Spontaneous intracranial hypotension


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