Meningocele-induced Positional Syncope and Retinal Hemorrhage

Ivo Bekavac and John I. Halloran

Summary: Meningocele is recognized as a rare, usually asymptomatic condition not associated with acute neurologic symptoms. We herein describe the case of a patient with a longstanding history of a lower back “mass” and recurrent syncope who became acutely unresponsive and developed bilateral retinal hemorrhages when she was placed in the supine position to undergo carotid sonography. MR imaging revealed a large, dorsal lumbar meningocele. The episode likely was caused by acutely increased intracranial pressure caused by displacement of CSF from the meningocele intracranially.

A simple dorsal meningocele is a protrusion of CSF and meninges into the subcutaneous tissue through a spinal defect. The overlying skin is usually intact (1). A complex meningocele represents a meningocele that is associated with other significant spinal anomalies (2). To the best of our knowledge, syncope and retinal hemorrhage are not reported manifestations of dorsal meningoceles.

Case Report

A 78-year-old right-handed woman had fallen onto her lower back without head injury or loss of consciousness at age 20 years. Shortly thereafter, she noticed a soft mass in her lower back that continued to grow until the age of 50 years. Six months after the fall, she experienced her first syncopal episode. She soon discovered that applying pressure to the mass caused a pressure sensation and ringing in her ears, blurred vision, and made her “see stars.”

Since then, she had experienced syncopal episodes approximately every 2 years, and each episode had been associated with inadvertent compression of the meningocele. She experiences transient headaches without any additional complaints after each of these syncopal episodes.

The patient had no symptoms of lumbosacral radiculopathy or lower back pain. These episodes were not associated with seizure activity or incontinence. The symptoms were present as long as the patient was in the supine or sitting position and disappeared without permanent deficit after she assumed a lateral decubitus or standing position.

The patient had been avoiding any pressure against the affected area, sleeping on her side, and using a special pillow when driving or sitting. The symptoms had been similar in severity until the episode described in this case report. The patient had undergone neurosurgical evaluation in 1981, and surgery was not recommended.

In January of 2002, the patient underwent carotid duplex sonography. The sonography technologist, despite the patient’s admission regarding her condition, placed the patient in the supine position for the examination. The patient immediately lost consciousness and was unresponsive for several minutes. She regained consciousness after her husband lifted her upright. Immediately after the unresponsive episode, she was confused, had a headache, and lost vision in her left eye. No incontinence, weakness, or numbness occurred. Blood pressure and heart rate were not assessed.

Neurologic and ophthalmologic examinations revealed visual acuity of 2/200 in the left eye and intraretinal hematoma bilaterally, more pronounced on the left. After 1 month, her vision improved to 20/40 in the left eye with no residual retinal hemorrhage in either eye.

A physical examination revealed a large, easily compressible, nontender midsceral mass. The remaining results of the neurologic examination were normal.

MR imaging of the lumbar spine was performed to further evaluate the mass. Axial and sagittal view T1- and T2-weighted images were obtained with the patient in the lateral decubitus position. The examination revealed a very large, lobulated meningocele, the largest component of which measured 10.7 × 10.2 × 11.0 cm (Fig 1). The meningocele originates from the caudal aspect of the sacrum and projects into the left side of the buttock. A tethered cord was present. Findings of MR imaging of the brain and carotid duplex sonography were unremarkable.

Laboratory studies, including electrolytes, white blood cell count, creatine kinase, and troponin, were normal. Serum glucose was 198. EKG results were normal. These laboratory studies were ordered by the emergency department physician as part of a workup for syncope. Subsequently, the patient underwent a second neurosurgical evaluation and elected to defer surgery.

Discussion

Meningocele is a rare, usually asymptomatic, spinal anomaly. In this case, compression of the meningocele produced acute onset of clinical findings. We suggest that these findings are due to acutely increased intracranial pressure from displacement of extra-spinal CSF into the intracranial cavity.

The total average intracranial volume is 1700 mL. Brain, blood, and CSF are the components that occupy the intracranial cavity. Brain tissue makes up approximately 1200 mL of the total intracranial volume, CSF from 70 to 160 mL, and blood approximately 150 mL. The spinal subarachnoid CSF volume is 10 to 30 mL (3).

Weed proposed that the total intracranial volume of CSF, blood, and brain are relatively constant and changes in the volume of one are compensated by changes in others (4). Brain tissue is relatively noncompressible (5). Therefore, changes in CSF volume...
are compensated by changes in blood volume. When either volume exceeds the compensatory ability of the other, intracranial pressure will increase.

Normally, cerebral blood flow is constant with a cerebral perfusion pressure between 50 and 150 mm Hg or a mean arterial blood pressure of 60 to 160 mm Hg. When intracranial pressure rises above 40 to 50 mm Hg, cerebral perfusion pressure and cerebral blood flow may be diminished to a level that can cause loss of consciousness (3).

In this case, extra-spinal CSF (approximate total meningocele volume was 1200 mL) was displaced into the intraspinal and intracranial subarachnoid space. Even if only a fraction of the CSF contained within the meningocele was displaced intracranially, it may have led to a several-fold increase in the intracranial pressure with associated arrest of cerebral blood flow.

This case has important teaching points. First, not all cases of increased intracranial pressure (and its attendant signs and symptoms) are intracranial. Most research on the effects of increased intracranial pressure has used a model involving an intracranial mass or CSF flow obstruction as a cause for the elevated intracranial pressure. These are the causes that most often come to mind for physicians evaluating patients with symptoms of increased intracranial pressure. The cause for the elevated intracranial pressure in this case was extracranial and nonobstructive.

Second, the supine position of the patient at onset of syncope is very unusual. If this history is available, the radiologist may suggest a search for an extracranial source for the symptoms if such a search has not yet been conducted.

The short period of increased intracranial pressure required for the loss of consciousness (instantaneous) and retinal hemorrages (several minutes) is very unusual. It is likely due to the acuteness of the change and/or the markedly elevated level of the intracranial pressure. These signs and symptoms may represent clinical manifestations of dorsal meningocele previously under-recognized and should be kept in the differential when evaluating patients with atypical histories of syncope or "increased intracranial pressure."

References

INTRODUCTION
Every treatment or intervention, no matter how seemingly benign, carries with it some risks. Every caregiver should give consideration to the risk-benefit ratio before embarking on any therapy. Unnecessary treatments or those with negligible benefit, despite theoretically low risks, may occasionally raise the risk-benefit ratio to unacceptable levels. We report a case in which a treatment that may not have been indicated led to a dangerous complication and an expensive and prolonged treatment and recovery.

CASE REPORT
A 49-year-old, right-handed man underwent his first chiropractic treatment for chronic low back pain. Although there was no history of neck pain, he had neck manipulation as part of "chiropractic adjustment." Within half an hour after the treatment, he experienced right-sided neck pain with no additional symptoms. The pain was mild but persisted for several days. However, on the fifth day, he awoke with excruciating right neck pain spreading to his right ear and eye. He returned to his chiropractor and received a second neck manipulation, which increased the pain. He then went to an emergency room for evaluation. On examination, he reported blurred vision in his right eye in addition to his right neck pain. The only pertinent finding was hypertension, and he was started on atenolol. A computerized tomography study of the brain was normal, but, because of the worsening pain, he was referred to a neurologist. Other than the pain and blurred vision, he voiced no additional symptoms. The patient denied any weakness, numbness, double vision, slurred speech, or difficulty walking or any problem with coordination. Past medical history was remarkable for smoking (45 pack-years) and removal of melanoma 20 years earlier with no signs of recurrence. There was no history suggestive for underlying collagen vascular disease.

Physical examination revealed a blood pressure of 145/105 mm Hg and regular heart rate of 70 beats/min. There was no ocular or carotid bruit. The cardiac examination was normal, without murmur. Neurological examination did reveal right Horner's syndrome, but the rest of the examination was normal. An emergent magnetic resonance imaging (MRI) of the brain and neck, as well as a magnetic resonance angiography (MRA) of intra- and extracranial circulation was performed. There was a distal right internal carotid artery dissection. The rest of the MRI results were normal, including diffusion weighted images.

The patient was started on intravenous heparin. Various treatment options including anticoagulation, neurointervention, and antiplatelet treatment were discussed with the patient, and he decided to start antiplatelet therapy (clopidogrel and aspirin).

An extensive workup was obtained including a complete metabolic panel, coagulation studies, lipid profile, complete blood count, and homocysteine level. All the results were normal. The patient was discharged with a residual right Horner's syndrome but otherwise in good condition. One month later, he had only minimal anisocoria on the right and an MRA showed some improvement of the carotid artery stenosis.

Two months later, a follow-up MRA revealed the formation of an aneurysm just proximal to the stenosis. Conventional angiography confirmed the MRA finding (Figure 1). The patient subsequently had a successful stenting of the aneurysm. One month later, the patient remained asymptomatic; there was no radiographic evidence of restenosis, and clopidogrel was discontinued.

DISCUSSION
Arterial dissections result when the hemorrhage from an intimal tear or rupture of the vasa vasorum creates a false lumen.1,2 The incidence of spontaneous cervicocephalic dissection is 2.6 to 2.9 cases per 100,000 individuals.1,3 Internal carotid artery dissections are more common than vertebral dissections, representing approximately 66% of the reported cases.1,3 Although chiropractic manipulation is frequently mentioned as a cause of an internal carotid artery dissection, the medical literature does not support this cause-effect relationship.4 However, there is evidence that chiropractic manipulation is an independent risk factor for vertebral artery dissection.5 Despite this, the temporal proximity of onset and the symptom constellation strongly suggest the chiropractic treatment as the etiology of this patient's carotid artery dissection.
The clinical history is pivotal to the diagnosis of dissection, particularly for patients without stroke risk factors. Signs and symptoms of cerebral ischemia or a positive Horner’s syndrome with a history of neck manipulation or trauma are particularly suggestive. Neurological symptoms and signs may result from a mass effect of the dissecting aneurysm or cerebral ischemic damage because of thromboembolism or hypoperfusion in the distal arterial territory. In addition to the history and examination, vascular studies are essential in establishing the final diagnosis of dissection. These include conventional angiography, MRI and MRA, computed tomographic angiography, and ultrasonography.

Although the equipment and techniques for diagnosing cervicocephalic arterial dissection have evolved significantly in recent years, the question of the most appropriate treatment modality for any particular case remains open. Evaluation of the therapeutic modalities is further complicated by the variability in the clinical spectrum of arterial dissection and resultant complications. Treatment options include intraarterial and intravenous thrombolysis, anticoagulation, antplatelet agents, angioplasty, stenting, and surgery, each carrying its own risks. Unfortunately, the current treatment recommendations are based on small, uncontrolled series, case reports, and anecdotal experiences. The quality of evidence for these therapeutic approaches is restricted to grade C evidence, consisting of class four (nonrandomized, historical controls) studies and class five (case series, no controls). Despite this, it is interesting that, after the initial event, the majority of patients do well, either because of or despite treatment.

CONCLUSION
This case report raises two important issues. First, even seemingly benign treatments or manipulations have risks. Chiropractic neck manipulations can be dangerous, even life threatening. As with any other treatment, neck manipulations should be restricted to those cases in which there is a clear indication. The neurologic and radiologic literatures both report a number of vertebral artery and carotid artery injuries as a consequence of neck manipulation. Second, despite the number of carotid artery dissections reported, there seems to be no consensus regarding the best treatment approach.

REFERENCES