Case report

Cortical blindness associated with autonomic dysreflexia in a man with tetraplegia: A rare but serious complication

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Context: To describe a case of a 44-year-old man with complete C4 tetraplegia who developed transient cortical blindness in the subacute setting following episodes of autonomic dysreflexia.

Findings: Transient cortical blindness the day after surgery for appendicitis that had resulted in severe autonomic dysreflexia (AD) requiring aggressive blood pressure management. Imaging showed no evidence of acute stroke, but did show vasospasm in the occipital lobes. Vision improved over the next couple of months.

Conclusion/clinical relevance: This case illustrates a possible profound vasomotor phenomenon (cortical blindness) associated with AD and its symptomatic treatment. Early recognition of AD and treatment of its underlying cause cannot be overemphasized.

Keywords: Autonomic dysreflexia, Spinal cord injuries, Tetraplegia, Blindness, cortical, Appendicitis

Introduction

Autonomic dysreflexia (AD), a well-known complication following cervical and high thoracic spinal cord injuries (SCI), results in a paroxysmal reflex sympathetic response to noxious stimuli. While often self-limited, severe cases can result in significant long-term morbidity and mortality. Studies have shown the incidence of AD to range from 25 to 70% in patients with SCI at or above the T6 level.^{1,2} Signs and symptoms resulting from profound autonomic responses are varied. They commonly include headache, flushing, and sweating above the level of injury, piloerection, anxiety, and other symptoms.³ If left untreated, the hypertensive crisis from AD can result in seizures, strokes, coma, and death.⁴ Rapid recognition and appropriate treatment is critical to avoid catastrophic consequences. Removal of the noxious stimuli results in resolution of the autonomic response.

A 44-year-old man with C4 complete tetraplegia American Spinal Injury Association Impairment Classification A (AIS A) developed cortical blindness associated with vasospasm following a prolonged period of frequent and recurrent episodes of AD

requiring aggressive symptomatic treatment. This case illustrates a profound vasomotor phenomenon concomitant with this rare but potentially catastrophic complication.

Case report

A 44-year-old white man with a history of traumatic SCI with C4 tetraplegia AIS A, was admitted to the hospital for severe hypertension associated with AD. The initial injury was the result of a diving accident about 13 years prior to admission. The patient had no history of peripheral arterial disease or cardiac disease, and episodes of AD in the several years prior to this episode were mild and of brief duration. Previous episodes had been associated with bladder, distension, bowel distension, or urinary tract infection (UTI). During the first few years following his SCI, he had increased frequency of AD and has been maintained on a calcium channel antagonist. He had a history of type II diabetes mellitus that was well controlled on oral medications.

Initially, the patient was brought to the emergency department (ED) with symptoms consistent with his previous AD episodes and thought to have a recurrence of a UTI that had been treated the week before. While in the ED, blood pressures ranged from 109/61 to 189/108 mmHg; he was admitted to the general medicine

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service. Admission laboratory values were not indicative of an UTI. Evaluation of other common causes of AD (i.e. bladder/bowel distension, constipation, pressure, ulcers, etc.) failed to identify a cause for his condition. During the first several days of his admission, he continued to experience multiple daily episodes of symptomatic AD with headaches, flushing, and hypertension. Systolic blood pressure recordings were commonly >200 mmHg, which prompted aggressive management with multiple medications, including nicardipine drip, nitro paste, and hydralazine. Improvement in systolic blood pressures was significant, sometimes exceeding a 100 mmHg drop. Finally, after an extensive evaluation over several days, and despite absence of fever, nausea, or physical examination findings of intra-abdominal pathology, an abdominal computerized tomography (CT) scan was performed. Acute appendicitis was diagnosed requiring laparoscopic surgery. This was performed without complication on hospital day 10. AD resolved following surgery and he remained normotensive for the remainder of his hospital stay.

Although the AD and associated hypertension resolved, the day after surgery he developed acute bilateral vision loss associated with lightheadedness and dysarthria. This lasted for approximately 2 hours and then improved, but did not completely resolve. Two more episodes of shorter duration of vision loss with similar symptoms occurred over the next 8 hours. Neuroimaging (MRI, magnetic resonance imaging/MRA, magnetic resonance angiogram) showed no evidence for acute stroke, but did show evidence of vasospasm in both the posterior and anterior circulation

Figure 1 Brain MRA – basilar and posterior circulation with vascular attenuation consistent with vasospasm.

Figure 2 Brain MRA – frontal anterior circulation with decreased vasculature flow consistent with vasospasm.

(Figs 1 and 2). CT angiogram was also performed, which confirmed the existence of the vasoconstriction.

While symptoms had improved at the time of discharge, he continued to have diminished visual acuity for months after the incident. At multiple follow-ups over the next few years, he reported no more episodes of transient blindness. However, an MRI performed almost 2 years later showed multiple focal signal abnormalities that had not been seen in earlier scans, which were consistent with remote infarctions involving multiple areas of the brain including the bilateral occipital lobes.

Discussion

In this patient, profound vasomotor instability with significant blood pressure variability over many days from AD (and its treatment) due to appendicitis was followed by this rare but serious co-morbidity. The cortical blindness appeared to be the direct result of vasospasm within the occipital lobe. With symptomatic treatment of the hypertension and subsequent treatment for the appendicitis, the patient's labile blood pressure improved significantly. Unfortunately, he developed acute visual disturbances the day following the resolution of the AD episodes. The patient was noted to be normotensive both during the episodes of vision loss, as well as during the subsequent imaging procedures and the remainder of hospitalization.

Although causation of the vasospasm cannot be determined with certainty, it is indeed interesting to speculate as to what events following the resolution of AD may have resulted in the vasospasm, occipital ischemia, and cortical blindness. Possibilities include

occipital ischemia resulting from the onset of vasospasm following correction of the recurrent AD and labile blood pressures, continued vasomotor instability associated with AD on a subacute level, or perhaps some other unknown mechanism. Causes other than vasospasm for the occipital ischemia and cortical blindness are less likely. Imaging findings were consistent with vasospasm and did not show an acute infarction consistent with thrombus. Cardiac monitoring throughout hospitalization showed no evidence of cardiac arrhythmias, which could have resulted in cerebral hypoperfusion or an embolic source. Furthermore, being normotensive around the time of the cortical blindness without aggressive pharmacological management makes medication-induced hypoperfusion and ischemia unlikely. While the blindness did improve in the short term, he continued to have visual deficits for months. Fortunately, the blindness did eventually resolve.

Review of the medical literature indicates that documented vasoconstriction during AD, as well as cortical blindness associated with AD, is quite uncommon. Reversible vasoconstriction associated with AD was previously described by Edvardsson and Persson.⁵ They described a patient with recurrent thunderclap headaches associated with AD. Initial imaging was negative, but cerebral angiogram did show multiple intracerebral vasculature changes. That patient was diagnosed with reversible cerebral vasoconstriction syndrome associated with AD and was treated for this. Similar to our case, subsequent MR imaging showed ischemic intracerebral changes.

We were able to identify one case that reported episodes of cortical blindness attributed to AD. Chaves and Gee⁶ reported a case of a 55-year-old woman who developed short-term cortical blindness due to a coughing fit. MRI findings showed hyper-intensities of the bilateral occipital lobes and left post-central gyrus. Her symptoms resolved quickly and an MRI 1 month later showed resolution of the lesions. This present case was different in that the patient continued to have symptoms for months with slow resolution of symptoms. This occurred despite no evidence of an acute stroke on MRI/MRA. The only change was vasospasm. Also, this patient's blindness occurred in the subacute stage after the resolution of the AD episode.

Although documented vasospasm with ischemia is rare from AD, there have been reports in the literature of transient, stroke-like symptoms in patients with AD.

Colachis and Fugate⁷ reported a patient who developed transient aphasia after an AD episode. A full stroke work-up was negative. The patient experienced associated seizure-like activity. They ultimately concluded that the seizures were the result of the increased sympathetic activity, while the aphasia was attributed to cerebral vascular insufficiency. This mechanism appears similar to that in this case; vasospasm is postulated to have caused decreased cerebral flow to the occipital region, leading to the transient cortical blindness.

The present case adds to the literature another uncommon but serious long-term complication associated with AD. It is important to recognize that in rare instances, serious sequelae of AD can appear in the subacute stage following the removal of noxious stimuli. This case also further emphasizes the need to look for more uncommon causes of AD and to obtain early imaging of insensate regions of the body when the more common causes of AD are eliminated.

Conclusion

Profound vasomotor changes associated with AD and its treatment can result in serious and life-threatening complications. This present case of cortical blindness associated with vasospasm following a prolonged period of frequent and recurrent episodes of AD with aggressive blood pressure management further illustrates profound vasomotor phenomenon associated with AD and its symptomatic treatment. The importance of early recognition and treatment of this potentially life-threatening condition cannot be over emphasized.

References

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