Abstract

A 49-year-old white male presented with a pseudo-subarachnoid hemorrhage and diffuse brain edema. Neuroimaging showed brain edema causing the unusual findings of a pseudo-subarachnoid hemorrhage and bilateral occipital lobe infarcts following herniation and compression of the posterior cerebral arteries. An enlarged corpus callosum was noted which led to a brain biopsy and a diagnosis of gliomatosis cerebri.

Keywords: gliomatosis cerebri; pseudo-subarachnoid hemorrhage

Case report

A 49-year-old previously healthy white man presented locally with new onset, moderately severe headaches for 2 to 3 months prior to admission. He had been seen for recently diagnosed systemic hypertension and his systolic blood pressure was in the 180 to 200 mm Hg range. He was started on oral amlodipine (Norvasc) and his blood pressure improved but his headaches persisted. He then developed worsening headaches, nausea, vomiting, diaphoresis, and tingling in his hands and feet. On 6/6/04, he presented to a local emergency room, with blood pressure of 183/112 mm Hg, where a computed tomogram (CT) of the head was interpreted to be consistent with a diffuse subarachnoid hemorrhage (Figure 1). He was then transferred to the University of Iowa Hospital.

On arrival at our institution, he was awake and alert with no neurological deficits. His past medical history was remarkable only for prior refractive surgery. His family and social history were non-contributory. Review of the outside head CT was consistent with diffuse cerebral edema with pseudo-subarachnoid hemorrhage. He underwent CT angiogram and catheter angiogram studies that did not reveal any etiology for subarachnoid hemorrhage. Infectious disease and neurology consultations were obtained.

The patient was transferred to our institution at 9:00 AM on 6/6/04. By 1:30 PM he had acutely developed bradycardia (30–40 bpm) and had become unresponsive. The pupils were fixed and dilated and he suffered several convulsions. He also had transient ventricular tachycardia. The patient was intubated, transferred to the neurosurgical intensive care unit and treated with dexamethasone. A right frontal ventriculostomy was placed with intracranial pressures noted to be in the 30–35 mmHg range. Cerebrospinal fluid (CSF) analysis showed an elevated CSF white blood count of 1040 cells/mm³, 10,920 red blood cells/mm³, protein was 8 mg/dL and glucose was 82 mg/dL. The cytology was negative and cultures showed no growth. Repeat CSF analysis several days later showed a white cell count of 3 cells/mm³ with normal glucose and protein, suggesting that the elevated white and red cell counts on the initial assessment were increased due to traumatic CSF sampling.

CT scan of the head showed bilateral occipital infarcts and diffuse brain edema. Serologic and CSF testing for Lyme disease, herpes simplex virus (HSV), and West Nile virus were all negative. The patient was treated empirically with intravenous vancomycin, ceftriaxone, and acyclovir.

Over several days his intracranial hypertension improved and he became awake and conversant. He was noted to have a severe visual field deficit OU. On 6/15/04, the neuroophthalmology exam at the bedside showed a visual acuity

This work was supported in part by an unrestricted grant from Research to Prevent Blindness, Inc., N.Y., N.Y.
Pseudo-subarachnoid hemorrhage and cortical visual impairment as the presenting sign of gliomatosis cerebri

of hand motions OD and counting fingers at 3 feet OS. Confrontation visual fields showed sparing of only his left superior quadrant of visual field OU with a bilateral homonymous hemianopsia. The pupils were isocoric, reactive OU, and there was no relative afferent pupillary defect. There was mild underaction of horizontal gaze bilaterally with about 20-prism diopter exotropia by Krimsky testing. Slit lamp examination showed a normal anterior segment OU. The intraocular pressure was 10 mm Hg OU. Ophthalmoscopy showed grade 2 to 3 optic disc edema OU. Magnetic resonance imaging (MRI) of the head showed bilateral occipital lobe ischemic infarctions, diffuse brain edema, and diffusely abnormal hyperintense T2 and FLAIR signal extending subcortically into the deep white matter and thickened appearing corpus callosum while sparing the subcortical white matter in gyri along the convexity and watershed distributions (Figure 2). A stereotactic brain biopsy was performed in the subcortical white matter adjacent to the splenium of the corpus callosum. Histopathologic examination of tissue samples showed mild increased cellularity with atypical astrocytes, infiltrating the neuropil without forming a mass or destroying underlying tracts, consistent with gliomatosis cerebri1 (Figure 3). The patient was discharged to a physical rehabilitation facility.

Discussion

Gliomatosis cerebri (GC) is a rare primary astrocytic tumor that diffusely infiltrates the brain with relatively preserved cerebral architecture. The treatment options are limited due to the diffusely infiltrative nature of the tumor causing the prognosis to be poor. Surgical resection is not generally possible and radiotherapy has not produced improved outcomes.2-4 This case is unusual for a number of reasons. First, the patient presented with symptoms suggestive of acute aneurysmal subarachnoid hemorrhage. His head CT scan was initially suggestive of a subarachnoid hemorrhage but was most consistent with a pseudo-subarachnoid hemorrhage. The differential diagnosis for increased attenuation of...
basal cisterns and subarachnoid spaces seen in pseudo-subarachnoid hemorrhage on CT scan includes: 1) pyogenic leptomeningitis, 2) intrathecally administered contrast medium, 3) leakage of high-dose IV contrast medium, and 4) diffuse cerebral edema. The proposed mechanisms for pseudo-subarachnoid hemorrhage include: displacement of the normally hypoattenuated CSF and engorgement of the superficial venous structures secondary to diffuse brain edema causing an elevation of the intracranial pressure. The subarachnoid space fills with the meninges and distended blood vessels.5–7 Second, this patient developed cortical blindness from transtentorial herniation caused by the diffuse brain edema. This resulted in compression of the posterior cerebral arteries bilaterally on the tentorium. Although this finding has been reported previously, it is an unusual finding in gliomatosis cerebri.5–11 Weinberg et al. reported three cases in whom posterior fossa decompression was performed for life-threatening tonsillar herniation.3

Third, the MRI finding of the enlarged corpus callosum led to a diagnostic brain biopsy of gliomatosis cerebri.12–14 Del Carpio-O’Donovan reported the radiographic findings of gliomatosis cerebri in 22 patients and 12 had corpus callosum involvement.2 Essig et al. reported callosal infiltration in 6 of 7 cases.12 Finally, although unproven, this patient presented with new onset and difficult to control systemic hypertension and then acutely herniated on the day of transfer. It is possible that the systemic hypertension was related to the Cushing response from increased intracranial pressure. The Cushing response is believed to be the result of dysfunction in the reticular activating system leading to systemic hypertension from sympathetic mediated vasoconstriction followed by bradycardia, respiratory irregularity, and sometimes death.15

References