CASE REPORT

Encephalopathy, Chiasmal Compression, Ophthalmoplegia, and Diabetes Insipidus in Pituitary Apoplexy

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ABSTRACT

Pituitary apoplexy with haemorrhage is a potentially life-threatening condition, and a rare cause of third nerve palsies. The range of vision loss and ophthalmoplegia seen in cases of apoplexy reflects the variability of cranial structures compressed by mass effect. The pathophysiology of extraocular muscle limitation and facial paraesthesia occurs with compression of the cavernous sinus, which contains cranial nerves III, IV, VI, and the ophthalmic branch of V. Blood supply to adjacent structures may be also compromised, causing additional loss of function. This case report of a patient with diabetes insipidus and a third nerve palsy illustrates the anatomic basis of the presenting signs of pituitary apoplexy, and the necessity for prompt neuroimaging if it is suspected.

Keywords: Central diabetes insipidus, pituitary apoplexy, pupil-involving third nerve palsy

CASE REPORT

A 50-year-old African American male with a history of hypertension presented with a 4-day history of progressive, painful vision loss in the right eye. The vision loss was described by the patient as “closing in superiorly and inferiorly” and progressed to no light perception in the right eye. The patient also noticed a droopy right eyelid and an outward drift of the right eye. The corresponding vision in the left eye had loss of the temporal visual field. Systemically, the patient complained of a right-sided headache, nausea, vomiting, diarrhoea, extreme thirst, and polyuria. His wife described a personality change over the preceding week. The patient denied any recent trauma or previous episodes of vision loss.

On initial examination, visual acuity was no light perception in the right eye and 20/200 in the left eye. Both pupils were equal in size, but a relative afferent pupillary defect in the right eye was noted. External

examination showed a ptosis and exotropia of the right eye. Confrontational visual field testing of the left eye showed complete temporal hemianopsia. The remainder of the ophthalmic examination was within normal limits. Specifically, optic nerve examination showed healthy nerves bilaterally without pallor or papilloedema. The constellation of symptoms was suspicious for an intracranial process, and a magnetic resonance imaging (MRI) was ordered. Imaging showed a reduced ventricular size and a $2.7 \times 1.7 \times 1.5$ cm pituitary adenoma displacing the optic chiasm superiorly on the right with partial extension to the left side. Figure 1 A magnetic resonance angiography (MRA) was performed to further characterize the lesion prior to neurosurgical intervention, and it showed a left side deviation of the pituitary stalk with apoplexy and haemorrhage measuring $1.9 \times 1.9 \times 1.2$ cm. The MRA also revealed 50% compression of the right internal carotid artery due to mass effect and compression of the right cavernous sinus.

Received 4 June 2014; revised 3 July 2014; accepted 3 July 2014; published online 15 September 2014

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FIGURE 1 T2 and T1 axial, coronal, and sagittal images showing a $2.2 \times 1.7 \times 1.5$ cm Sellar mass that demonstrates mild enhancement with contrast. There is mass effect on the right cavernous sinus, with compression of the cavernous right internal carotid artery, and mild elevation of the optic chiasm.

Initially, serum sodium was decreased to 130 mEq/L on presentation due to hypovolaemia secondary to the diarrhoea. After a 2-L normal saline bolus, his creatinine returned to baseline from 3.30 mg/dL. The acute change in mental status change resolved as the sodium normalized; however, the sodium continued to rise to 145 mEq/L. Further endocrinologic work-up was consistent with the diagnosis diabetes insipidus, as the urine specific gravity was 1.005 and urine osmolality was 200 mOsm/kg. Serum osmolality was elevated at 309 mOsm/kg. Concurrently, the patient was managed with Decadron 4 mg every 6 hours for intracranial swelling and acute adrenal crisis. The patient then underwent neurosurgical intervention via a transphenoidal approach to decompress the adenoma. Pathology revealed fragments of necrotic tissue with inflammation suggestive of a pituitary tumour, although no viable tissue was obtained for definitive diagnosis. Subsequent follow-up confirmed complete resolution of the cranial nerve palsies, although dense superior and inferior arcuate visual field defects persisted in the right eye. A 24-4 Humphrey visual field (Figure 2) showed a small superior central island corresponding to a best-correct visual acuity was 20/40 in the inferior field. An early superior arcuate defect was also noted

in the left eye. Figure 2 The patient is doing well clinically and is currently managed with hormone therapy for panhypopituitarism.

DISCUSSION

Pituitary apoplexy with haemorrhage is a potentially life-threatening condition, and a rare cause of third nerve palsies.^{1,14,16} The range of vision loss and ophthalmoplegia seen in cases of apoplexy reflects which cranial nerves are compressed by mass effect.^{15,16} The pathophysiology of extraocular muscle limitation and facial paresthesia is due to a compressive effect within the cavernous sinus, which contains cranial nerves III, IV, VI, and the ophthalmic branch of V.^{1,15} Superior compression of the optic chiasm can lead to bitemporal hemianopia.¹

Pituitary apoplexy, in a patient with a known or unknown adenoma, can be precipitated during pregnancy, postpartum, following surgery, and during treatment with bromocriptine or anticoagulation.⁴⁻⁹ It is often idiopathic, and clinically mimics rupture of a Rathke cyst, craniopharyngioma, or central nervous system lymphoma.^{10,11} Case reports of apoplexy occurring without a mass lesion, such as

FIGURE 2 Humphrey visual fields status post resection of pituitary mass showing a central island OD and a superior arcuate defect OS.

after cardiac surgery and postpartum, offer several pathophysiologic mechanisms of action, including hypotension with haemorrhage after infarction or microemboli.^{12,15} Several proposed etiologies for the cause of the cranial nerve palsies exist, including direct tumour expansion into the cavernous sinus or indirect compression of the nerves through the cavernous sinus wall, impingement of the oculomotor trigone at the posterior clinoid process, and infarction of the nerves.¹² Although most commonly causing unilateral findings, cases of pituitary apoplexy causing bilateral cavernous sinus compression have been reported.¹³

Clinically, most patients present with headache emanating from behind the forehead and orbit.^{1,11} This is secondary to haemorrhage extending into the dura surrounding the pituitary, causing meningeal irritation and distention.¹⁶ Less common presenting symptoms include restricted visual fields, nausea, vomiting, diplopia, decreased visual acuity, altered mental status, and hemiplegia.¹¹ Medical treatment involves observation of neurologic status along with intravenous steroids and hormone replacement.¹¹ Disruption of the hypothalamic-pituitary axis by mass effect can range from isolated anterior pituitary hormones, diabetes insipidus with posterior pituitary compression, to panhypopituitarism.^{2-3,5-8,10,11,17} Of these, diabetes insipidus can cause high output of dilute urine (5200 mOsm/kg) with a specific gravity of less than 1.005.⁷ Subsequently, the serum osmolality can rise to over 300 mOsm/kg, with resultant hypernatraemia.^{6,17} The level of electrolyte imbalance can induce a mild change in mental status to coma.¹⁷ Iatrogenic administration of desmopressin induces urine concentration and normalizes serum osmolality.¹⁷

Surgical decompression is warranted if visual acuity and fields are compromised or are rapidly diminished, or if there is altered mental status.^{1,4} Most

commonly, the mass is removed using a transphenoidal approach.^{1,4}

It is estimated that 60–80% of pituitary apoplexy cases occur subclinically, which illustrates the atypical and fulminant presentation in this case.¹ Symptomatic apoplexy in surgically resected pituitary adenomas has been estimated to be between 0.6% and 9.1%.⁹ Given the rarity, it reinforces that a high level of suspicion and emergent neuroimaging are critical to make the diagnosis. The key to a good clinical outcome in cases of pituitary apoplexy is timely intervention to correct the life-threatening addisonian crisis and prevent further visual compromise.

Declaration of interest: The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

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