Primary Pseudotumor Cerebri Syndrome in a Young Obese African Woman

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A 22 year old, obese (body mass index 41) black African woman presented at Princess Marina Hospital, Gaborone, Botswana with a 2 week history of repeated episodes of diffuse headache exacerbated by the Valsalva manoeuver and accompanied by light intolerance, dizziness, neck pain and blurred vision. The day before presenting she had had an episode of visual loss. At presentation the patient was oriented and her blood pressure, pulse, respiratory rate and temperature were normal. She had no neck stiffness; reflexes, tone, sensation and motility were normal. However, limited abduction and no light perception of the right eye were noted. Hematology and biochemistry examinations were within normal limits. Antinuclear antibodies were negative.

Fundoscopy revealed bilateral papilledema with flame hemorrhages, and engorgement of the veins and exudate bilaterally, i.e., features of acute papilledema [Figure 1]. A brain computerized tomography scan and magnetic resonance imaging [Figure 2] showed no abnormality, and a cerebrospinal fluid study only revealed high CSF pressure. A diagnosis of primary pseudotumor cerebri syndrome (idiopathic intracranial hypertension) was made.

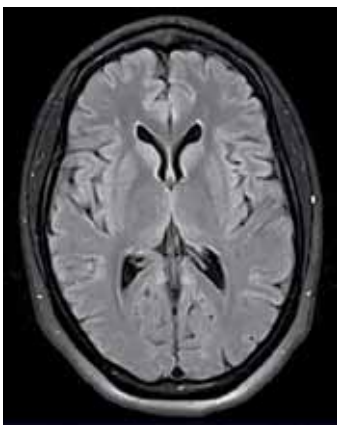
The patient was treated with tapering doses of prednisolone (initially 100 mg daily), amitriptyline (25 mg daily), propranolol (80 mg daily) and acetazolamide (1 g daily). Her condition improved and 2 months later the papilledema was resolving, although exudate was still present in both maculae and the vision of the right eye was poor.

Idiopathic intracranial hypertension, or primary pseudotumor cerebri syndrome, is a rare neurologic disorder of unknown etiology characterized by chronically increased intracranial pressure and symptoms and signs of a space-occupying intracranial mass (headache, transient visual obscurations, papilledema) in the absence of a mass or ventricular obstruction [1-3]. It generally presents in women of childbearing age and has no racial predilection. Obesity and recent weight gain occur more frequently in these patients [4]. Obesity might contribute to the pathogenesis by increasing the intraabdominal pressure and the cardiac filling pressure and therefore impeding the venous return from the

CSF = cerebrospinal fluid

Figure 1. Fundoscopy showing papilledema with flame hemorrhages, engorgement of the veins and exudates: **[A]** right eye, **[B]** left eye

Figure 2. Brain MRI showing no abnormalities



brain and increasing the intracranial pressure, or through neuroendocrine effects on the mineralocorticoid receptor which is abundant in choroid plexus epithelial cells that regulate CSF production.

Treatment for primary pseudotumor cerebri syndrome is intended to reduce intracranial pressure and thus prevent visual loss and headaches. Medical treatment includes a low sodium weight-reduction diet, carbonic anhydrase inhibitors, steroids, digoxin and loop diuretics. Surgical treatment (nerve sheath fenestration, lumboperitoneal shunt, ventriculoperitoneal shunt, or subtemporal decompression) is necessary for patients unresponsive to therapy [5].

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