Grand Rounds

A 16-year-old girl with bilateral optic disc swelling

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History

A 16-year-old girl with a 6 months' history of moderately severe headaches was referred to Wolverhampton and Midland counties Eye Infirmary (WMEI) for ophthalmological evaluation after her optician had noted swollen discs; she had seen the optician at the request of her general practitioner to rule out refractive error as the cause of headaches. She described the headaches as generalized, though more severe in the mornings. They did not worsen with coughing, sneezing, or physical activity and were unrelated to posture. She did not experience tinnitus or any whooshing auditory sensations. She denied nausea or vomiting and reported no visual symptoms.

According to her mother, she had gained about 10 kg in the previous 6 months. Her menstrual cycles were largely regular; she had neither dysmenorrhea nor menorrhagia. She presented to the hospital emergency department 7 months previously with right iliac fossa pain, which was attributed to constipation. She was discharged following dietary advice and laxatives, which relieved her symptoms.

Examination

Corrected visual acuity was 6/4 in both eyes. No color vision deficit could be elicited. Anterior segments were unremarkable in both eyes. Pupillary reactions were normal, with no relative afferent defect. Dilated fundus examination showed bilateral optic disc swelling (Figure 1) with absent spontaneous venous pulsations, although the retinal vasculature appeared normal. Goldmann visual fields (Figure 2) showed an enlarged blind spot in both eyes. B-scan ultrasonography showed no evidence

of optic nerve head drusen. Computed tomography (CT) of the head and orbits was normal, and lumbar puncture revealed a cerebrospinal fluid opening pressure of 13 cm H₂O (normal pediatric range, 11.5–28 cm H₂O).¹

General physical examination was normal, apart from a distended abdomen. At 76 kg, the patient was slightly overweight for her height (5'4") and her blood pressure was elevated, at 150/106 mm Hg. A pediatric referral was arranged. Ambulatory blood pressure monitoring by the pediatricians showed an average blood pressure of 156/110. A hypertensive work-up ensued.

Ancillary Testing

To exclude all causes of hypertension in the patient's age group, the pediatricians performed a detailed work-up, including urea and electrolytes, urinary cortisol measurements (9 AM and midnight), liver function tests, gamma glutamyl transferase, urine HCG, full blood count, erythrocyte sedimentation rate, C-reactive protein, thyroid function tests, and early morning urine test for microalbuminuria. All tests were normal.

A lumbar puncture showed clear and colorless cerebrospinal fluid, total protein count of 23 mg/100 mL (normal, 15–60 mg/100 mL), gamma globulin 11% of the total protein (normal, 3%–12% of total protein), glucose level of 76 mg/100 mL (normal, 50–80 mg/100mL), cell count of 0–5 white blood (all mononuclear) cells (normal, 0–5) with no red blood cells, and chloride level of 117 mEq/L (normal, 116–130 mEq/L). Microbiology of the cerebrospinal fluid was unremarkable, with no growth.

An electrocardiogram was normal.

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Figure 1. Fundus photographs of right (A) and left (B) eyes at presentation.

Figure 2. Results of Goldmann visual field testing of right (A) and left (B) eyes at presentation.

In view of the normal lab work-up, an ultrasound of the abdomen was requested to rule out gynecological or renal causes of hypertension. Ultrasonography of the abdomen showed a large abdominal cyst extending from the symphysis pubis to the level of xyphisternum (Figure 3). Magnetic resonance imaging of the abdomen showed a cyst arising from the pelvis and measuring $34 \times 29 \times 12$ cm. Within its right side border an additional septated area was seen, suggesting a small additional follicular cyst within a large ovarian cyst. No renal compression or hydronephrosis was noted.

Treatment

The abdominal cyst was surgically removed. Exploratory laparotomy revealed an ovarian cyst weighing approximately 12 kg. A right salpingo-oophorectomy was performed and histology revealed a serous cystadenoma. Following surgery, her hypertension resolved, and she maintained excellent vision. The disc swelling showed evidence of improvement, with faintly positive

spontaneous venous pulsations. Her visual fields improved in both eyes and have remained static (Figure 4). At 2 months follow-up, abdominal ultrasound revealed a left ovarian cyst, which is being monitored by serial ultrasound scans at four-month intervals since the last 2 years.

Differential Diagnosis

Bilateral optic disc swelling can result from a variety of inflammatory, infectious, toxic, metabolic, genetic, or vascular conditions, including hypertension.²

In hypertensive optic neuropathy, the mechanism of disc swelling is controversial, but ischemia of the optic nerve head is thought to play a role.³ However, it is usually unilateral or, if bilateral, is frequently associated with other signs of hypertensive retinopathy, such as retinal hemorrhages and exudates and cotton wool spots. There can also be signs of hypertensive choroidopathy with exudative retinal detachments.⁴

Figure 3. Ultrasonography of the abdomen showing a large abdominal cyst extending from symphysis pubis to the level of xyphisternum, approximately 25 cm in transverse, 40–60 cm in longest axis and 20 cm deep (all at deepest and widest points): in relation to the uterus (UT) (A), the bladder (LADDER) (B), and the left ovary (LT OV) (C). Content is generally clear and anechoic, with a suggestion of a trace of debris at its dependant aspect.

Optic nerve head drusen can cause bilateral optic disc swelling, but spontaneous venous pulsation is present in about 80% of patients, and it is not associated with headaches. B-scan of the optic discs is usually diagnostic, but a lack of calcification in children and adolescents can sometimes render this investigation unreliable. In

such cases the optic discs have a pink appearance, and the edges are rarely completely obscured.

Idiopathic intracranial hypertension can cause papilledema with associated headaches. It usually affects young, overweight women. Patients frequently report worsening of the headaches with activities that increase intracranial pressure, such as coughing or sneezing. They can have visual obscuration, tinnitus, and auditory sensations. CT scan may be normal, although there may be small or slitlike ventricles and an “empty sella sign.” Cerebrospinal fluid pressure is raised on lumbar puncture.

An intracranial space-occupying lesion will usually be detectable on head CT or will be associated with evidence of hydrocephalus. Hypertension is not a feature of these conditions.

Diagnosis

On presentation, the patient had had headaches for 6 months, bilateral disc swelling with enlarged blind spots, and was moderately hypertensive. She did not have any other signs of hypertensive retinopathy or of raised intracranial pressure. CT scan of the head and orbits was normal. Abdominal imaging showed a large ovarian cyst, but no hydronephrosis. She was diagnosed with hypertensive optic neuropathy.

This case is unusual because the intra-abdominal cyst seems to have caused hypertension and bilateral disc swelling in the absence of common fundoscopic signs of hypertensive retinopathy. Whereas polycystic ovarian syndrome is known to cause hypertension through hemodynamic alterations and variations in peripheral vascular resistance, it is uncommon for bilateral disc swelling to be a presenting feature.^{5,6} Moreover, the presence of a large solitary ovarian cyst does not fulfill the criteria of PCOS diagnosis.⁷ We hypothesize that the size of the ovarian cyst may have had a role in compressing the renal veins, thus causing an associated hypertension and bilateral optic neuropathy.

Bilateral optic disc swelling secondary to malignant hypertension has been reported in children, although this is usually accompanied by, for example, retinal hemorrhages, exudates, or cotton wool spots.⁸

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Figure 4. Results of Goldmann visual field testing of right eye (A) and left eye (B) after removal of cyst.

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