

Papilledema secondary to parietal dermoid - epidermoid cyst with superior sagittal sinus compression

Áine Ní Mhéalóid, MBBCh, BAO, MRCSI; We Fong Siah, MBBCh, BAO, MRCPI

Department of Ophthalmology, Mater Misericordiae University Hospital, Dublin, Ireland

Abstract

Dermoid and epidermoid cysts are benign slow growing tumors that arise from ectodermal tissue. In this case report, a 32-year-old gentleman presented with a right posterior parietal dermoid - epidermoid cysts resulting in superior sagittal sinus compression. Clinical examination disclosed enlarged blindspot formation in both eyes and bilateral papilledema. He was treated with a low-pressure shunt, resulting in resolution of the papilledema and his symptoms. In a search of the peer-reviewed medical literature (using MEDLINE and cross-referenced literature), this report may be the second to report papilledema secondary to an intracranial dermoid or epidermoid cyst with superior sagittal sinus compression.

Introduction

Papilledema is a specific type of swelling of the optic nerve head, or disc edema, secondary to raised intracranial pressure (ICP), often secondary to intracranial tumor, central nervous system inflammation, or idiopathic intracranial hypertension. Dermoid and epidermoid cysts are slow growing tumors that account for only 1.5% of all brain tumors; they are typically extra-axial lesions, and may occur anywhere in the neuraxis.¹ They are benign

developmental abnormalities that arise from ectodermal tissue.

Superior sagittal sinus (SSS) thrombosis is a well-recognized cause of raised ICP.² However, intracranial tumors can also give rise to non-thrombotic compressive occlusion of dural venous sinuses resulting in papilloedema.² In this case report, a 32-year-old gentleman presented with non-thrombotic SSS compression from a right posterior parietal extra-axial dermoid - epidermoid cyst causing bilateral papilledema.

Case Report

A 32-year-old gentleman presented with a one-week history of transient visual obscurations (TVOs) associated with postural change. He denied any headache, nausea, or vomiting. He did however admit to regularly taking a sports supplement containing a methamphetamine-like compound. On examination, visual acuity was 20/20 in each eye, color vision was normal, and there were no other neurological focal signs. His body mass index was within the normal range. Fundoscopy revealed bilateral frank papilledema, more prominent on the left side (Figure 1). Goldmann visual fields disclosed bilateral enlarged blindspots (Figure 2). Computed tomography imaging of the brain and orbits did not show any abnormality.

Correspondence:

Aine Ni Mhealoid, MBBCh, BAO, MRCSI
Eye Department, Mater Misericordiae Hospital
3 Tudor Grove
Ashbourne, Co Meath, Ireland
E-mail: ainenimhealoid@rcsi.ie
Phone: (353) 86-360-1361

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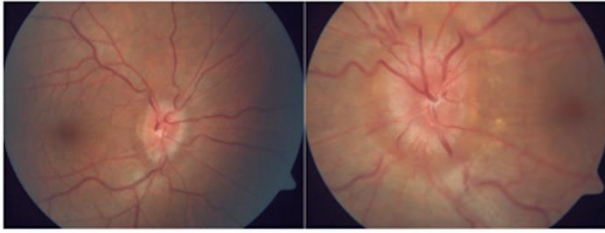


Figure 1
Fundoscopy disclosed bilateral disc edema, in the left eye more than right.

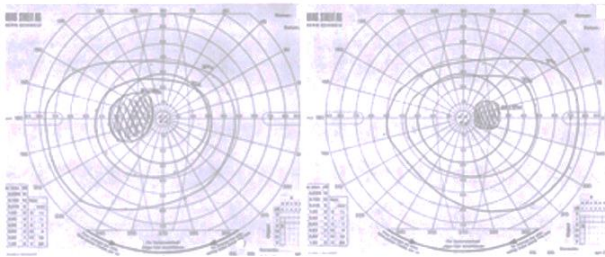


Figure 2
Goldmann visual fields disclosed bilateral enlarged blind spots.

Magnetic resonance imaging and magnetic resonance venography of the brain showed a right posterior parietal extra-axial mass, 1.6 cm in maximal diameter, compressing on the SSS suggestive of a dermoid or epidermoid cyst (Figure 3). A posterior SSS filling defect measuring 2.1 cm x 1.7 cm x 0.7 cm was evident. Lumbar puncture (LP) was performed with an opening pressure of 40 cmH₂O and a closing pressure of 12 cmH₂O. A thrombophilia screen was normal.

The patient was transferred to a neurosurgical center, where he was treated with oral acetazolamide 500 mg once daily. This led to resolution of his TVOs. However, he subsequently developed a headache; a repeat LP was performed with an opening pressure of 6 cmH₂O. A diagnosis of low-pressure headaches was made, and the dose of acetazolamide was reduced to 250 mg once daily. Stereotactic surgery was deemed too risky and shunting of the SSS was also out-ruled due to the possibility of the patient developing chronic low-



Figure 3
Axial T2 weighted magnetic resonance imaging (left) revealed a hyperintense lesion of the posterior parietal lobe. Sagittal T1 weighted post-contrast restricted-diffusion magnetic resonance imaging (right) revealed a mildly enhancing posterior parietal lobe lesion.

pressure headaches. Five weeks later, the patient's TVOs returned and he decided without medical consultation to increase his dose of acetazolamide to 500 mg. Clinical examination disclosed persistent papilloedema and a repeat Goldmann visual field showed persistent enlarged blind spots. A repeat LP was performed with an opening pressure of 32 cmH₂O. A low-pressure shunt was inserted by the neurosurgical team, which led to resolution of the papilledema and his symptoms.

Discussion

TVOs are a well-known symptom of increased ICP and papilledema.³ These visual disturbances can be explained by two mechanisms: optic disc edema causing ischemia of the optic disc⁴ or the effect of a space occupying lesion causing intermittent occipital lobe ischemia as a result of posterior cerebral artery compression against the tentorial margin.³ SSS thrombosis is a well-recognized cause of raised ICP. However, non-thrombotic occlusion of the dural venous sinuses can also give rise to raised ICP¹ often with the clinically distinctive feature of a lack of focal neurological signs.⁵ This phenomenon was first noted by Plant, et al. who described two cases of occipital

skull tumors causing occlusion of the SSS leading to raised ICP.¹

Dermoid and epidermoid cysts represent approximately 1.5% of all intracranial masses.^{6,7} Epidermoid cysts are comprised of stratified squamous epithelial tissue surrounding cystic fluid,⁶ which are derived from ectopic inclusions during the closure of the neural tube between the third and fifth weeks of fetal development.⁸ Dermoid cysts are closely related tumors of similar origin⁹ that contain varying amounts of ectoderm derivatives including apocrine, sweat and sebaceous glands as well as hair follicles, squamous epithelium and occasionally teeth.¹⁰

Only one other case of papilledema from an intracranial dermoid or epidermoid cyst has been described thus far in the peer-reviewed literature (identified through a search of the peer-reviewed medical literature using MEDLINE and cross-referenced literature). Maloca, et al. described the case of a 26-year-old gentleman with visual loss, headaches induced by exercise, photophobia, and concentric visual field defects due to a massive intracranial epidermoid cyst. Subsequent decompression and systemic steroid therapy did not prevent deterioration in the patient's visual acuity.¹¹

Meningiomas may occasionally produce increased ICP by occluding a venous sinus leading to persistence of papilledema and continuation of visual loss if measures are not taken to resect the tumor and to improve venous drainage. Metastatic lesions must also be considered as causes of compression, the site of which is usually found at the terminal portion of the SSS and the torcular Herophili.¹

Spontaneous rupture of epidermoid and dermoid cysts is uncommon.¹⁰ Clinical presentations of such an occurrence may include headache, seizure, aseptic meningitis, hydrocephalus, vasospasm, cerebral ischemia, neuropsychiatric symptoms, olfactory delusion, hemiparesis, visual loss, facial numbness and diplopia. Ruptured cysts require surgical removal with extensive irrigation of the subarachnoid space

during the surgery.⁷ In this case, surgical resection was deemed to carry an unacceptable risk, as sparing of the superficial veins of the cerebrum entering the SSS is necessary in order to prevent brain swelling and venous infarction.¹² However, the development of adequate anastomotic channels between the SSS and elements of the cerebral venous system proximal to the site of obstruction may facilitate the safe removal of a portion of the SSS in future.¹ Stereotactic surgery was also out-ruled as a therapeutic option in this case. Cerebral venous outflow obstruction due to extrinsic compression can be treated with venous sinus stenting.¹³ Reconstruction of the venous lumen with endovascular stents may be effective in lowering elevated cerebrospinal fluid pressure but may not be successful in all patients due to secondary narrowing of the sinus lumen, which may be reversed by shunt surgery procedures.¹⁴

Dermoid and epidermoid tumors should be considered in the differential diagnosis of papilloedema.¹¹ The risk of cyst rupture either spontaneously or intra-operatively must also be taken into consideration. Over time, anastomotic venous channels may develop resulting in adequate drainage of the SSS if it has been slowly occluded by a tumor. Subsequent removal of a portion of the SSS may be carried out safely.²

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