Unilateral optic disc swelling associated with idiopathic hypertrophic pachymeningitis: a rare cause for a rare clinical finding

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DESCRIPTION

A woman aged 28 years presented with a 1-year history of left-sided headache. The headache was continuous and interfering with activities of daily living. She did not have vomiting or visual obscurations. She developed left-sided sixth nerve palsy and facial numbness and was referred to us. On clinical evaluation, she had left-sided sensorineural deafness. Fundus examination showed optic disc swelling on the left side and a normal right eye (figure 1A). MRI brain with contrast showed features of hypertrophic pachymeningitis predominantly affecting the left side involving the tentorium and cerebral cortex (figure 1B, E) and encasing the cavernous sinus (figure 1C, D). Optic nerve sheath dilation was evident on the left side (figure 1F, G). Blood counts, serum biochemical tests and erythrocyte sedimentation rate were normal. The serum was negative for rheumatoid factor, antinuclear antibodies and antidouble-stranded DNA. The serum VDRL test and hepatitis B surface antigen were negative. Work up for neurosarcoidosis including chest CT scan, serum calcium and ACE were negative. A CT scan of the abdomen did not reveal evidence for a neoplastic or a chronic inflammatory disease. The cerebrospinal fluid (CSF) pressure measured at lumbar puncture was normal at 12 cm H₂O. The CSF showed mild lymphocytic pleocytosis, normal biochemical values and was negative for malignant cells. Microbiological evaluation of the CSF for cryptococcal antigen and VDRL was negative. The

CSF culture for fungi and acid-fast bacilli was also negative. Dural biopsy showed meningeal thickening and non-specific chronic inflammation and lacked features of granuloma or vasculitis. The biopsy specimen was negative for acid-fast bacilli and fungal stains. A diagnosis of idiopathic hypertrophic pachymeningitis (IHPM) was made based on the neuroradiological findings of thickened dura, histopathological findings of non-specific inflammation and exclusion of known causes of chronic inflammation. She was initiated on oral prednisone 1 mg/kg/day. At 3 months follow-up, she was headache-free and the optic disc swelling had resolved. She is being maintained on azathioprine and low-dose steroids.

Papilloedema refers to optic disc swelling in the presence of raised intracranial pressure (ICP).¹ In the absence of raised ICP, it is referred to as disc oedema. Unilateral optic disc oedema in the setting of normal ICP as seen in our case is caused by ischaemia (eg, anterior ischaemic optic neuropathy, retinal vein occlusion) and demyelination (eg, multiple sclerosis) of the optic nerve and diabetic papillopathy.² It can also be due to direct compression of the optic nerve due to intracranial lesions like sphenoidal meningioma, optic-nerve glioma and pituitary adenoma where the disc swelling is ipsilateral to the lesion. In our patient, IHPM was causing the compression resulting in ipsilateral disc swelling and multiple cranial nerve palsies. To the best of our knowledge, this inflammatory lesion is

Figure 1 Fundus examination showed left-sided disc oedema. (A) MRI brain postcontrast axial (B, C) coronal (D) and sagittal (E) images showed the pachymeningeal thickening involving the parieto-occipital convexity and tentorial leaflets (arrows in B and E) and cavernous region (arrows in C and D). MRI brain axial (F) and coronal (G) T2 image showed left optic nerve sheath dilation (arrows).

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not reported to be associated with unilateral disc oedema. Our case reiterates the importance of brain imaging in all patients with unilateral optic disc swelling. Inflammatory lesions like

Learning points

- ▶ Careful evaluation of optic fundus on both sides should be performed in every case of headache.
- ▶ The presence of unilateral optic disc swelling also warrants further evaluation with neuroimaging.
- ▶ Medically manageable ipsilateral intracranial inflammatory lesions like idiopathic hypertrophic pachymeningitis can also present as unilateral optic disc swelling.

IHPM which can be medically managed should also be considered along with other tumours as mentioned above while evaluating a patient with this clinical presentation.

Contributors RSI conceived the idea and designed the paper. SP reported the eye finding and helped in finalising the draft. MR helped in interpretation of the eye finding and contributed in the reporting.

Competing interests None declared.

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REFERENCES

- 1 Huna-Baron R, Landau K, Rosenberg M, *et al*. Unilateral swollen disc due to increased intracranial pressure. *Neurology* 2001;56:1588–90.
- 2 Van Stavern GP. Optic disc edema. *Semin Neurol* 2007;27:233–43.

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