

# Myxopapillary ependymoma of cauda equina presented with communicating hydrocephalus and papilloedema: A case report

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### SUMMARY

**We reported a case of cauda equina myxopapillary ependymoma in a patient who presented with atypical history of progressive blurring of vision. Ophthalmology examination revealed relative afferent pupillary defect, binasal hemianopia and papilloedema. This case report serves as a reminder that the intraspinal tumour could be a cause of papilloedema, despite rare, should be considered in a hydrocephalus patient who presented with no intracranial pathology and minimal spinal symptoms.**

### INTRODUCTION

Communicating hydrocephalus secondary to intraspinal tumour is rare.<sup>1</sup> The diagnosis is challenging as blurring of vision might be the initial presentation. Symptoms pertaining to the spinal lesion may be minimal.

### CASE REPORT

A 33-year-old man presented to our district eye clinic with reduced left eye vision for two weeks. He gave history of intermittent bilateral blurring of vision of three months duration, each time lasted seconds to few minutes which subsequently recovered fully. He denied headache, nausea or vomiting. He did not experience loss of weight or appetite, fever, chronic cough or pulmonary tuberculosis contact. He was a known case of controlled hypertension. On examination visual acuity was 6/9 in the right and counting finger close to face in the left. His examination revealed relative afferent pupillary defect in the left eye, binasal hemianopia and papilloedema (Figure 1). The neurological examination was normal but on further questioning, patient gave history of non-specific low backache for six months. Otherwise there was no sciatica pain or autonomic dysfunction. Power of both lower limbs was full with normal gait, and no sensory change over the perineum and legs. Magnetic Resonance Imaging (MRI) of the brain showed dilated ventricular system with no obvious intracranial space occupying lesion (Figure 2). Multiple lumbar punctures were attempted but failed due to dry taps. MRI of spine eventually revealed cauda equine tumour with encasement of the nerve

roots within with fairly extensive leptomeningeal enhancement of the rest of the cord (Figure 2). Insertion of intracranial pressure monitor was performed with clear colourless cerebral spinal fluid (CSF) drained. Ventricular CSF contained 0.607g/l of protein, with normal glucose and cell count, culture and sensitivity did not grow any organism. Infective screening for tuberculosis, syphilis, hepatitis B and C, and retroviral, as well as demyelinating disease and autoimmune diseases as the causes of papillitis were all negative. Right ventriculoperitoneal shunt was inserted in view of persistent high intracranial pressure (ICP) monitoring range 20 to 30 mmHg and worsening of vision. Subsequently, L5 laminectomy and tumour biopsy only were performed in view of nerve roots encasement by the tumour. Grossly, the tumour was soft and pinkish with moderate to high vascularity. Histopathologic examination showed myxopapillary ependymoma, WHO grade 1. Immunohistochemistry was done. The neoplastic cells expressed glial fibrillary acidic protein (GFAP), CD 99 and epithelial membrane antigen (EMA). The S100 stain was negative. Ki67 proliferation index is low. One month later, however, left vision acuity did not improve. Papilloedema resolved and both eyes optic atrophy were noted. No new neurological signs of observed. Repeated MRI of the spine one-year post-operative revealed no progression of tumour.

### DISCUSSION

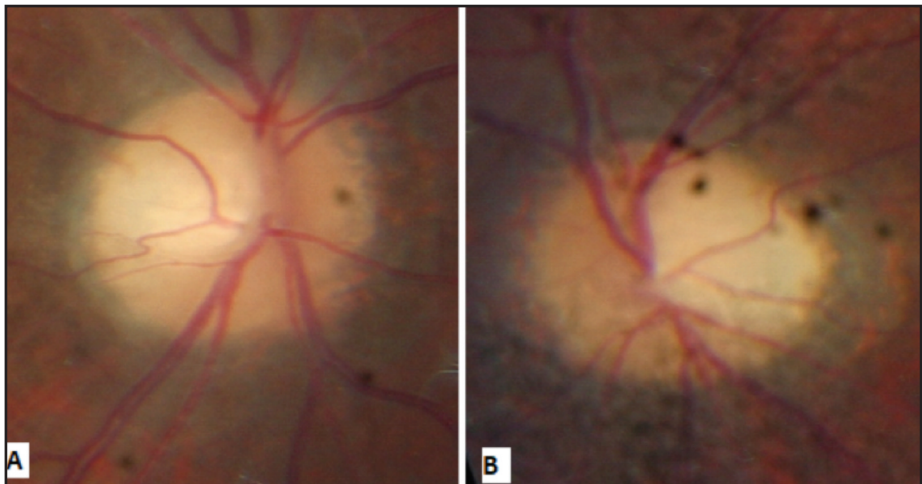
Ependymomas in adults are rare. Myxopapillary ependymomas are generally seen in the lumbosacral spine or cauda equine cauda.<sup>2</sup>

Myxopapillary ependymoma appear to be slow growing tumour, with a mean patient age at presentation of 35.5 years<sup>2</sup> which corresponded to our case. Chronic back pain is the commonest presenting symptoms, with or without sciatica. However, our patient presented with rare binasal hemianopia. This could be due to lateral displacement of the parts of both optic nerves and uncrossed fibres at chiasma against the internal carotid arteries by a distended third ventricle in the internal hydrocephalus.<sup>3</sup>

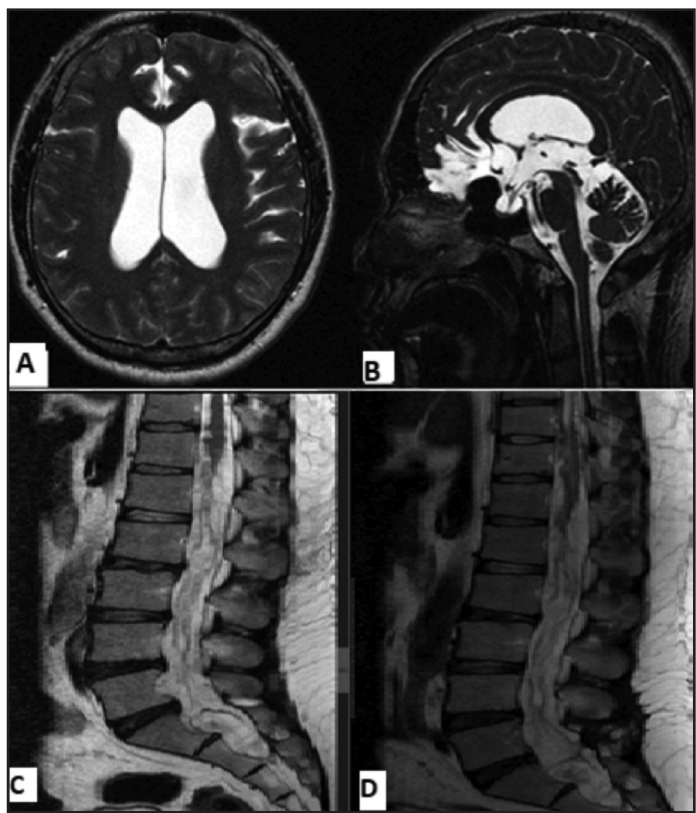
*This article was accepted: 24 May 2019*

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**Fig. 1:** Pre-operative fundoscopy showed bilateral (A – right side, B – left side) optic discs with peripapillary greyish halo, and obscuration of border vessel with pale temporal disc. The multiple black spots on (B) were artefact.



**Fig. 2:** A: Pre-operative axial view of brain MRI in T2 sequence showed, dilated ventricular system; B: Pre-operative sagittal view of MRI brain showed ballooning of 3rd ventricle, with enlargement of supra-sellar and pre-pontine cisterns; C: Pre-operative MRI of spine, T2 sequence sagittal view, revealed extensive cauda equina tumour; D: Post-operative one year, T2 sequence sagittal view showed no worsening of tumour.

Raised CSF protein content has been observed in myxopapillary ependymoma. Large protein molecules may mechanically clog the pores of the semipermeable membrane of the arachnoid, delay CSF absorption and in turn producing hydrocephalus.<sup>1</sup> Hayreh, demonstrated that mechanical phenomenon in the raised intracranial pressure (ICP) causing the formation of papilloedema.<sup>4</sup> Increased ICP is

transmitted from the subarachnoid space via the optic nerve sheath to cause axoplasmic flow stasis, disc swelling, retinal venous stasis and subsequent fluid leakage.

The papilloedema patients will experience transient visual obscuration which was seen in our case. The symptom is due to transient ischaemia or distortion at nodes Ranvier's

myelin. Chronic papilloedema causes progressive and permanent nerve fibre loss, optic atrophy and leading to vision loss.

In summary, an intraspinal tumour could be a cause of papilloedema, despite rare, should be considered in a hydrocephalus patient who presented with minimal spinal symptoms. In the absence of intracranial pathology, spinal MRI should be performed to locate the possibility of spinal lesion origin.

#### **ACKNOWLEDGEMENT**

The authors would like to thank the Director General of Health Malaysia for his permission to publish this article.

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