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PAPILLEDEMA WITH SIXTH NERVE PALSY AND BILATERAL OCULAR HYPERTENSION IN PATIENT WITH POSTERIOR FOSSA TUMOR

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ABSTRACT

Background: Papilledema is bilateral optic disc swelling due to high intracranial pressure. Papilledema cannot occur in the absence of high intracranial pressure, but high intracranial pressure can occur in the absence of papilledema. Increased intracranial pressure can causes an increase in intraocular pressure due to increase of ophthalmic venous pressure.

Case Presentation: A 11-years-old woman presented with blurry and double vision with difficulty moving the eyeballs, especially when looking outward 2 weeks after ventriculoperitoneal shunt (VP shunt) removal surgery. The patient has a history of brain tumor with anatomical pathology results pilocytic astrocytoma in posterior fossa since 2015. Ophthalmology examination revealed bilateral papilledema with visual acuity was 1/60 and <1/60, increased intraocular pressure, sixth nerve palsy. After VP shunt surgery, the intracranial pressure was slowly reduced and returned to normal, the visual acuity was getting better to 6/21 in the right eye, but in the left eye was still 1/60.

Conclusion: Management of papilledema, either medically or surgically are according to the underlying pathological process and ocular findings. In patients with an underlying mass lesion, removal of the mass should be performed. A lumboperitoneal shunt or a ventriculoperitoneal shunt can be used to improve cerebrospinal fluid (CSF) flow. If papilledema develops rapidly, this will worsen the prognosis.

Keywords: papilledema, increased intracranial pressure, sixth nerve palsy

BACKGROUND

Optic disc edema refers to ophthalmoscopic swelling of the optic disc with a concomitant increase in fluid in or around the axon. Many terms have been used to describe optic disc edema including swollen optic disc, disc edema, papilledema, and papillitis. The most common terms to describe this phenomenon are optic disc edema and swollen optic disc, whereas other terms imply the cause of optic disc swelling. For example, papilledema is a term for optic disc edema caused by increased intracranial pressure.[1,2]

By definition, papilledema cannot occur in the absence of high intracranial pressure, but high intracranial pressure can occur in the absence of papilledema. Papilledema is often bilateral and symmetrical. Possible conditions causing elevated intracranial pressure and papilledema include intracerebral mass lesions, brain hemorrhage, head trauma, meningitis, hydrocephalus, spinal cord lesions, impaired brain sinus drainage, cranial anomalies, and idiopathic intracranial hypertension (IIH). Regardless of the cause, visual loss is a dreaded morbidity of papilledema, and the main mechanism of optic nerve damage is secondary intraneuronal ischemia.[1-3]

The main mechanism of visual loss is probably due to stasis ofaxoplasmic flow. A high increase in intracranial pressure results in an increase in cerebrospinal fluid pressure around the optic nerve, which disrupts the normal gradient between intraocular pressure and retrolaminar pressure, leading to high tissue pressure within the nerve. Increased tissue pressure within nerves disrupts metabolic processes that mediate axoplasmic flow.[1,3,4]

The diagnosis of papilledema was established based on neuroophthalmological examination, fundoscopy, and radiology. Treatment is directed at correcting the underlying cause. Management depends on the severity of the disease, ranging from conservative therapy, medication, and surgery.[1,3]

CASE PRESENTION

A 11-years-old girl, was consulted from the paediatric neurology department after post-removal VP shunt due to exposed shunton December 27th, 2021. Two weeks after VP shunt removal, the patient complained of blurry and double vision when she looked with both eyes. The patient also complains difficulty moving the eyeballs, especially when looking outward and pain in botheyes and intermittent headache without nausea, vomiting, nor limb weakness.The patient denied the presence of pain when moving the eyeball.

The patient has a history of brain tumor since 2015 and has had VP Shunt insertion surgery in March 2015 due to increased intracranial pressure and has had Craniotomy for tumour Resection (CTR) surgery in April 2015 with the anatomical pathology results were: pilocytic astrocytoma (WHO grade I) in the right cerebellum (posterior fossa), but there was still residual tumour in the right cerebellum (posterior fossa). In December 2nd, 2022, the patient had had VP shunt removal surgery due to shunt expose, and 2 weeks after surgery signs and symptoms of increased intracranial pressure was appeared.

Upon ophthalmologic examination, her best corrected visual acuity (BCVA) of both eyes were 1/60 on the right eye and < 1/60 on the left eye consecutively. Intraocular pressure was increased on both eyes, 25.8 mmHg on the right eye and 30.4 mmHg on the left eye. Other anterior segment examination within normal limits with no relative afferent pupillary defect (RAPD) was detected.

Ocular motility examination showed a restriction to lateral gaze in both eyes and after the forced duction test there was no resistance. Fundus examination showed edematous optic disc on both eyes with partial obscuration, telengiectasis, and tortous vessel in the retina (Figure 1). Head CT scan on first day of VP shunt removal showed a fossa posterior mass in the right hemi cerebellum and arachnoid cyst in the left temporal that appeared as a hypodense lesion, with 3,2 x 2 cm in size but signs of hydrocephalus have not beenseen(Figure 2).

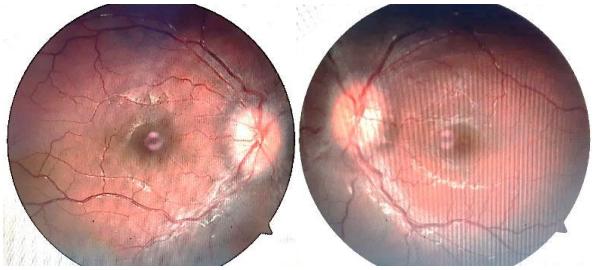


Figure 1. Funduscopic examination of right eye shown edematous optic disc on both eyes with partial obscuration, telengiectasis, and tortous vessel in the retina.

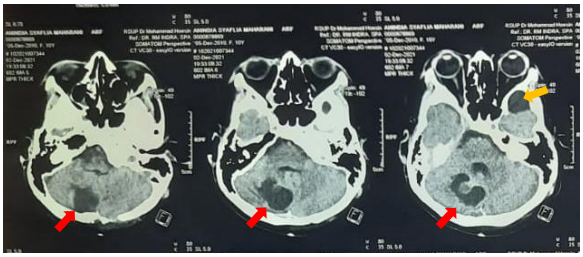


Figure 2. A head CT-scan first day after VP shunt removal. In axial plane, a fossa posterior mass was appeared in right hemi cerebellum (red arrow) and arachnoid cyst in left temporal (yellow arrow)

Patient was diagnosed as bilateral papilledema with bilateral sixth nerve palsy and bilateral ocular hypertension et causa increased intracranial pressure due to posterior fossa tumor. The patient was given 2 types of intraocular pressure lowering drugs from 3 different groups, nonselective beta blocker, carbonic

anhydrase, manitol and was planned to have a VP shunt re-insertion surgery by the neurosurgeon.

Four weeks post-operative the patient hadno longer complained of double vision and headache. On ophthalmologic examination, the visual acuity in the right eye had progressed to 6/21 but in the left eye the vision was still 1/60. Ocular motility examination showed no abnormality on both eyes and intraocular pressure was normal on both eyes.

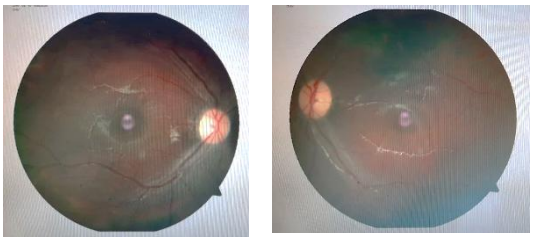
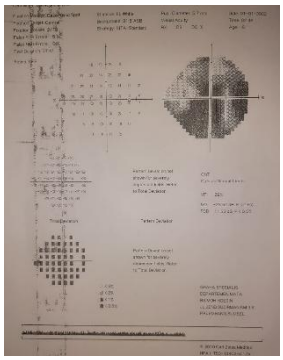
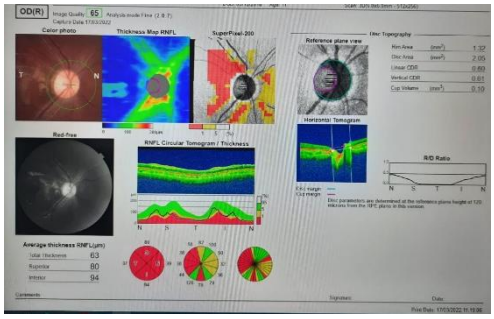


Figure 3. Funduscopic examination shown temporal pallor of the optic disc on both eyes



(a)



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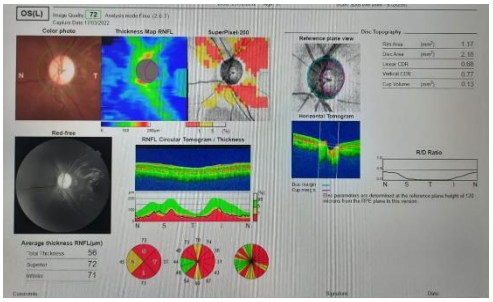

(c)

Figure 4. A 24-2 Humphrey visual Field test (a) revealed wide visual field defect. Optic disc OCT scan (b) right eye and (c) left eye revealed thinning RNFL thickness.

Fundus examination revealed temporal pallor of the optic disc in both eyes (Figure 3). Humphrey visual field (HVF) was performed in her right eye and found a wide visual field defect, especially in inferior segment. On Optical coherence tomography (OCT) examination of both eyes (Figure 4), it was found that there was thinning of the retinal nerve fiber layer (RNFL) in the optic disc of both eyes.

DISCUSSION

An increase in intracerebral volume from a mass lesion may produce high ICP leading to papilledema, but compensatory mechanisms may preclude the development of papilledema in chronic cases. In one series, papilledema was found in only 28% of patients aged 0–90 years with a history of brain tumor presenting to an emergency department, but the sensitivity and reliability of detection of papilledema in this setting may be low. In contrast, several larger neurosurgical series found papilledema in up to 60%–80% of patients with cerebral tumors.[1]

Infratentorial mass lesions, which may obstruct the ventricular outflow at the relatively narrow Sylvian aqueduct are more likely to produce papilledema than supratentorial mass lesions. Brain tumors in children are more commonly located in the posterior fossa, and thus present more frequently with papilledema.[1]

Papilledema presents as a bilateral phenomenon and can develop within hours to weeks after an increase in intracranial pressure occurs. Decreased visual acuity to permanent blindness can occur if the etiology of increased intracranial pressure is not treated.[5,6,7] The study of Hayreh et al. explained that disc swelling in papilledema is the result of a mechanical phenomenon. The cerebrospinal fluid in the intracranial subarachnoid space has a continuous relationship with the subarachnoid space in the orbit. Pressure can be transmitted to the subarachnoid space in the orbit when there is an increase in intracranial pressure. This results in distension of the optic nerve sheath in the subarachnoid space of the orbit.[7,8]

Distension of the optic nerve sheath will impede lymphatic flow in the dura layer resulting in obstruction of backflow from the CSF. This condition will stimulate the proliferation of meningothelial cells in the innermost layer of the arachnoid and pia thereby further narrowing the subarachnoid space and producing compartment syndrome. This is in direct compression against the axon.[7,9] Passi et al said this can trigger obstruction of blood vessel flow resulting in distension of veins and capillaries as well as increased capillary permeability and accumulation of extracellular fluid. This results in swelling of the optic disc. Elevated intracranial pressure (tumor or pseudotumor) can also cause nerve palsy due to downward compression of the brainstem stretching the subarachnoid segment of nerve VI between the exit point of the brainstem and the dural adhesions, this is usually accompanied by headache and papilledema as occurs in this patient.[10]

This patient also had an increase in intraocular pressure in both eyes. Zhen li et al in their study described direct pressure transmission through the cerebrospinal fluid surrounding the optic nerve sheath at the point where the optic nerve enters the orbit. Most of the blood in the optic vein returns to the intracranial cavernous sinus through the superior orbital fissure. Increased intracranial pressure causes an increase in ophthalmic venous pressure, which is transmitted directly to the ocular fluid, thereby increasing intraocular pressure.[11,12]

Management of increased intracranial pressure aims to reduce cerebrospinal fluid and treat complaints, signs and symptoms of increased intracranial pressure. Treatment is carried out according to the patient and the course of the disease, which can be done by observation, medical or surgical. The patient was given 2 types of intraocular pressure lowering drugs from 2 different groups, namely the non-selective beta blocker group and the carbonic anhydrase group, both of which have a mechanism of action by reducing the production of aqueous humor. It is known that the increase in intraocular pressure in these patients is caused by an increase in intracranial pressure which causes an increase in ophthalmic venous pressure, which will be transmitted directly to the ocular fluid, thereby increasing IOP. Therefore, the selection of the two classes of antiglaucoma drugs can help reduce intraocular pressure. From the pediatric section, the patient also received mannitol to help reduce intracranial pressure, as well as to help reduce intraocular pressure. The patient was then planned to have a VP shunt re-installed by the neurosurgery department.[13,14,15]

The prognosis of papilledema is determined by the underlying cause. The late management and precise will lead to papillae atrophy. If papilledema appears quickly, this will be a sign of a poor prognosis. Papilledema with an elevation of more than 5 diopters, accompanied by bleeding and profuse exudate will worsen the visual prognosis.

After placing a VP shunt, signs and symptoms of increased intracranial

pressure. slowly reduced. At the fourth postoperative follow-up day, the headache had disappeared, and the intraocular pressure had returned to normal, so acetazolamide and mannitol were discontinued. However, the patient still complained of blurred vision and double vision and still had obstruction of lateral eye movement. In the posterior segment, papilledema was also found without obscuration.

Long-lasting papilledema can also cause progressive and permanent loss of nerve fibers. Both small and large fibers within the optic disc are affected, but more peripheral nerve fibers are affected earlier and in greater numbers. So that in chronic papilledema, it can cause limited visual field, especially the inferior area, this can gradually occur, which can further worsen into central vision loss and total blindness and at that time the chance of recovery is very small. In this patient also found a mass in the right hemicerebellum which is just below the right occipital. The presence of a mass may also cause compression of the right occipital area, which may be one of the reasons for the worse visual impairment in the patient's left eye than in the right eye. From the pediatric neurology department, the patient was still given paracetamol and ciproheptadine to reduce the patient's headaches, and from the neurosurgery department, the patient was scheduled for a re-perform head MRI for evaluation post VP Shunt installation. And after getting the results of the MRI, the plan will be consulted to the radiotherapy section for further management of pilocytic astrocytoma in this patient. [15,16]

CONCLUSION

Papilledema can be caused by a variety of intracranial conditions and reflects the associated increased ICP. It can also cause sudden or chronic visual loss through a multitude of pathophysiological mechanisms. Increased ICP can also causes increased ophthalmic venous pressure, which would be transmitted directly to the ocular fluid, thus increasing the IOP. Treatment options include medical and surgical modalities. This case needs a multidisciplinary treatment. As an ophthalmologists we should be able to diagnose as early as possible and along with neurosurgical expertise may contribute to the management of this patients when CSF diversion or decompressive procedures are indicated because of the risk of severe or progressive visual loss.

REFERENCE

1. Rigi M, Almarzouqi SJ, Morgan ML, Lee AG. Papilledema: epidemiology, etiology, and clinical management. Eye Brain. 2015 Aug;17;7:47-57.
2. McCafferty B, McClelland CM, Lee MS. The diagnostic challenge of evaluating papilledema in the pediatric patient. Taiwan J Ophthalmol. 2017 Jan-Mar;7(1):15-21.
3. American Academy of Ophthalmology, Basic and Clinical Science Course. Section 5 Neuro-Ophthalmology. Edisi 2020-2021. San Fransisco; 2021.
4. Stavern, Gregory. Optic Disc Edema. Seminars in neurology. 2007: 27. 233-43.
5. Killer HE, Jaggi GP, Flammer J. Cerebrospinal fluid dynamics between the intracranial and the subarachnoid space of the optic nerve. Is it always bidirectional? Brain. 2007. 514–20.
6. Miller NR, Subramanian P, Patel V. Walsh & Hoyt's clinical neuro- ophthalmology. Philadelphia: Wolters Kluwer Health; 2015.
7. Ray LA, Heys JJ. Fluid Flow and Mass Transport in Brain Tissue. Fluids. 2019.
8. Hayreh SS. Pathogenesis of optic disc edema in raised intracranial pressure. Prog Retin Eye Res. 2016 Jan;50:108-44.
9. Sakka L, Coll G, Chazal J. Anatomy and physiology of cerebrospinal fluid. Eur Ann Otorhinolaryngol Head Neck Dis. 2011 Dec;128(6):309-16.
10. Passi N, Degnan AJ, Levy LM. Imaging of papilledema and visual pathways: effects of increased intracranial pressure and

pathophysiologic mechanisms. AJNR Am J Neuroradiol. 2013. 919–24.
11. Li Z, Yang Y, Lu Y, Liu D, Xu E, Jia J, Yang D, Zhang X, Yang H, Ma D, Wang N. Intraocular pressure vs intracranial pressure in disease conditions: a prospective cohort study (Beijing iCOP study). BMC Neurol. 2012 Aug 3;12:66.
12. Wilson MH. The dynamic vascular and venous pathophysiological components of intracranial pressure. J Cereb Blood Flow Metab. 2016. 1338–50.
13. Lee AG, Wall M. Papiledema: Are we any nearer to a consensus on pathogenesis and treatment? Curr Neurol Neurosci Rep. 2012 :334–9.
14. Capo H. Don't miss this! red flags in the pediatric eye exam: blurred disc margins. Journal of Binocular Vision and Ocular Motility. 2019 Jul. 110–5.
15. Chen JJ, Bhatti MT. Papilledema. Int Ophthalmol Clin. 2019 Summer;59(3):3-22.
16. Schirmer CM, Hedges TR 3rd. Mechanisms of visual loss in papilledema. Neurosurg Focus. 2007;23(5):E5.