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Juvenile Glaucoma Related to Pineal Tumor Post Ventriculoperitoneal Shunt Procedure: A Case Report

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ABSTRACT

Background: Glaucoma is a cause of irreversible blindness worldwide. This study aims to describe a case of juvenile open-angle glaucoma with a history of post-ventriculoperitoneal shunt perineal tumor. **Case presentation:** A 27 years old man came with complaints of blurred vision in both eyes about 2 years ago. The vision in both eyes is felt to be progressively decreasing, almost the same in both eyes. Blurred vision felt suddenly while driving 1 year ago. The patient had been diagnosed with a pineal tumor and underwent surgery to install a ventriculoperitoneal (VP) shunt for hydrocephalus indications 3 years ago. The patient also underwent radiotherapy 26 times for the treatment of the pineal tumor. Ophthalmological examination revealed 20/80 OD and OS vision; not advanced with pinhole, no eyelid edema, intraocular pressure OD 21 mmHg, OS 23 mmHg. Visual fields index on OD 85% and OS 81%. Central corneal thickness (CCT) examination showed CCT of the right eye at 541 μm and left eye at 547 μm . The results of a brain CT scan showed that there was a heterogeneous solid mass with calcification and cystic (necrotic) in the pineal region. This patient was diagnosed with juvenile glaucoma with bilateral papillary atrophy et causa pineal tumor post VP shunt. The therapy given was latanoprost 1 drop/day ODS and Cendo Lyteers 1 drop/6 hours ODS, and Thymol 0.5% 1 drop/12 hours ODS. **Conclusion:** Monitoring and evaluation of staging and clinical progression of glaucoma patients with increased intracranial pressure is a condition that needs attention. The correlation between the two is the involvement of the lamina cribrosa in the form of a translaminal pressure gradient.

1. Introduction

Glaucoma is an optic neuropathy characterized by progressive degeneration of the optic nerve. Glaucoma is currently the second leading cause of irreversible blindness worldwide. Juvenile open-angle glaucoma (JOAG) is a rare form of primary open-angle glaucoma, with younger onset (3 to 40 years), higher intraocular pressure, and more severe visual field loss than adult-onset open-angle glaucoma.¹⁻³

The most important and modifiable risk factor for glaucoma is intraocular pressure (IOP), so most of the available glaucoma therapies aim to reduce IOP so as to reduce optic nerve damage and visual field loss.^{2,3} A number of studies and cases have reported that

cerebrospinal fluid pressure (CSFP) is a risk factor for glaucomatous cupping. This study aims to describe a case of juvenile open-angle glaucoma with a history of post-ventriculoperitoneal shunt perineal tumor.

2. Case Presentation

A 27 years old man came with complaints of blurred vision in both eyes about 2 years ago. The vision in both eyes is felt to be progressively decreasing, almost the same in both eyes. A Blurred vision was felt suddenly while driving 1 year ago. There were no complaints of headache and nausea, or vomiting. The patient then went to an ophthalmologist

in Padang, and he found increased eye pressure and impaired visual field. The patient received latanoprost eye drops to reduce eyeball pressure.

Based on auto anamnesis, the patient had worn minus glasses for 15 years and last changed his eyeglass prescription 1 year ago. However, the glasses that are commonly used do not reduce the patient's complaints. In addition, the patient was diagnosed with a pineal tumor and underwent a ventriculoperitoneal (VP) shunt surgery for

hydrocephalus indications 3 years ago. The patient also underwent radiotherapy 26 times for the treatment of the pineal tumor.

Ophthalmological examination revealed 20/80 OD and OS vision; not advanced with pinhole, no eyelid edema, intraocular pressure OD 21 mmHg, OS 23 mmHg. On fundoscopic examination, the cup-to-disk ratio was enlarged in both eyes, there was excavation, and retina ODS was good (Figure 1).



Figure 1. Fundus photo. Impression: enlarged cup-to-disk ratio in both eyes, with papillary ODS c/d = 0.8-0.9. excavation (+), retina ODS is good.

Optical coherence tomography (OCT) examination showed retinal nerve fiber layer (RNFL) thinning, the cup-to-disk ratio at OD 0.7-0.8; OS 0.8-0.9. Perimetric examination showed visual field defects in both eyes, with an arcuate pattern. Visual fields index on OD 85% and OS 81%. Central corneal thickness (CCT) examination showed CCT of the right eye at 541 μm and left eye at 547 μm . The results of a brain CT scan showed that there was a heterogeneous solid mass with calcification and cystic (necrotic) in the pineal region. This patient was diagnosed with juvenile glaucoma with bilateral papillary atrophy et causa pineal tumor post VP shunt. The therapy given was latanoprost 1 drop/day ODS and Cendo Lyteers 1 drop/6 hours ODS, and Thymol 0.5% 1 drop/12 hours ODS. During the patient's follow-up in the first week of treatment, the complaints gradually decreased, and

both eyes became more comfortable.

3. Discussion

Glaucoma is a neurodegenerative disease characterized by specific changes in the optic nerve and retina, resulting in retinal ganglion cell (RGC) apoptosis. Loss of RGC causes progressive loss of visual field, which makes glaucoma the leading cause of incurable blindness.⁴⁻⁶ The case illustration presents a male patient with cupping glaucomatous and visual field defects with a history of an intracranial mass. The patient initially complained of loss of strength in both legs and falls. Then a brain CT scan was performed, and it was found that the patient had a previous history of pineal tumors. This condition is a sign and symptom that there has been an increase in intracranial pressure, and papilledema may occur

in the patient.

The patient had time to delay the treatment until later found hydrocephalus. The patient underwent surgical therapy for the installation of a VP shunt and radiotherapy. The patient only realized that his vision had decreased 1 year later. Hydrocephalus is a secondary condition and often results from tumors of the pineal region.⁷ Hydrocephalus can cause an increase in intracranial pressure, which can result in impaired consciousness, cerebral hernia, and even death due to compression of the cerebral aqueduct. From the history and examination, it is suspected that there is a correlation between the history of an intracranial mass that has undergone surgical therapy and the presence of eye complaints, although the possibility of glaucoma before the presence of an intracranial mass has occurred cannot be removed.

Eye complaints in new patients are felt after the condition the increase in ICP with papilledema. This condition occurs because of an imbalance between IOP and ICP and their relationship with the lamina cribrosa. There is a relative increase in IOP after ICP decompression with a VP shunt. The VP shunt will cause the initially high ICP to decrease, and this condition will affect the structure of the lamina cribrosa, which previously became thin due to the increase in ICP, resulting in an increase in TLPG.⁸⁻¹¹ This condition is thought to be the mechanism of nerve fiber layer damage due to an imbalance between IOP and ICP, where IOP becomes relatively high compared to ICP, which decreases after a VP shunt. Gallina et al. found a correlation between normal ICP after VP shunt placement and the occurrence of NTG.^{9,12}

The eye anatomy that plays a role in the association between increased ICP and the occurrence of glaucoma is the lamina cribrosa. The lamina cribrosa (LC) is a "nets" structure on the posterior part of the sclera that allows the optic nerve fibers to pass through the eye. The anterior lamina cribrosa is the inside of the eye, and the posterior part of the lamina cribrosa is the optic nerve which is surrounded by the meninges. The optic disc is located at the junction between the relatively high-pressure intraocular space

and the low-pressure subarachnoid space. Therefore, the pressure imbalance between these two areas may be the cause of damage to retinal ganglion cell axons that pass through the lamina cribrosa.¹³⁻¹⁵

Lamina cribrosa is under the influence of two separate pressure components, namely IOP and ICP, so it is acceptable that significant pathological changes in the lamina cribrosa could be caused by changes in these two pressure components. Indeed, recent studies have clearly demonstrated that ICP is responsible for optic papillary damage with elevated IOP.^{12,16,17}

Research on the role of ICT in glaucoma is still being carried out, but the alternative hypothesis, which is widely put forward, is the hypothesis that IOP has an opposite pressure on IOP which affects the optic nerve head (ONH) and the lamina cribrosa. Fleishman and Berdahl have even presented the theory of CSF glaucoma, and according to this theory, the balancing force results from the difference between IOP and ICP. In this condition, there is a barrier that plays a role in regulating the balance between ICP and IOP, namely the translaminar pressure gradient (TLPG), which is the difference between IOP and ICP divided by the distance between them (thickness of the lamina cribrosa), so that TLPG depends on ICP, IOP, and IOP. The thickness of the lamina cribrosa. High TLPG is expected to affect ONH and further contribute to the occurrence of glaucoma. The mean TLPG pressure in humans is estimated to be 20-33mm mmHg/mm, whereas ICP is used to calculate TLPG in most cases.^{8,18,19} Previous studies have shown that ICP is significantly lower in patients with POAG compared to non-glaucoma controls. A decrease in ICP was also found in NTG patients with a calculated ICP that was statistically lower than that found in POAG. In contrast, ICP was increased in patients with ocular hypertension compared with controls.^{8,9}

Efforts to reduce intraocular pressure are the only therapeutic outcome for glaucoma management. This effort is carried out by achieving the target IOP. The initial target range for IOP depends on the stage of glaucomatous damage. In mild POAG, the initial IOP

target range can be maintained at 15-17 mmHg, moderate 12-15 mmHg, and in the severe stage of damage, 10-12 mmHg, where this classification is based on glaucomatous cupping that occurs (cup disc ratio; mild 0.65, moderate 0.7-0.85 and weight >0.9). For normal-tension glaucoma, a 30% reduction in IOP from baseline IOP has been shown to significantly reduce glaucoma progression.

The determination of the IOP target is an important step in glaucoma management, but decreasing IOP does not provide a complete guarantee that disease progression can be prevented because many other factors also play a role in the development of glaucoma. On examination of the optic nerve, it is important to observe the inferior and superior quadrants to identify the presence of the neuroretinal thinning of the rim and associated retinal nerve fiber layer defects and to measure the amount of structural damage to the optic nerve. This condition can be confirmed by optical coherence tomography examination, with thinning of the RNFL in the anterior segment and occurs bilaterally. It is, therefore, important to lower the IOP and achieve the target IOP due to the depleted RNFL. The central corneal thickness is also a determining factor in determining the target IOP. The normal CCT is 545 μm , this condition is important, especially in patients with myopia, as the thicker the actual IOP becomes larger, and conversely, the thinner the CCT, the true IOP is also lower than the IOP results obtained. In this patient, the CCT was still relatively within the normal CCT range, so the IOP target was in accordance with the condition of the optic disc.^{3,15,20}

The previous history of radiotherapy can also affect the incidence of glaucoma in patients. Radiation-induced optic neuropathy (RION) is a rare complication following radiation therapy exposure. Radiation-induced ischemic Free radical damage to the vascular endothelium has been theorized as the pathogenesis of RION. The severity of visual impairment can lead to complete blindness. In most cases, visual function worsens soon after the initial development of RION. Although there is no definite

evidence of a link between this mechanism in patients, the existence of this theory can be a factor that influences visual impairment.^{10,11,21}

Patients are given anti-glaucoma therapy, which aims to lower IOP while maintaining control for IOP examination. There is no specific treatment for the part of the eye associated with intracranial pressure. Prognosis in glaucoma patients is highly dependent on success in achieving IOP targets. The presence of risk factors in the patient in the form of; young age, history of pineal tumor, and myopia, the target IOP in the patient must be achieved so that it is hoped that it will slow the progression of glaucomatous optic and visual field defects in the patient.

4. Conclusion

Monitoring and evaluation of staging and clinical progression of glaucoma patients with increased intracranial pressure is a condition that needs attention. The correlation between the two is the involvement of the lamina cribrosa in the form of a translaminal pressure gradient.

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