JNI Jurnal Neuroanestesi Indonesia

Case Report

Optic Nerve Pilocytic Astrocytoma in a Pediatric Patient: A Case Report

I Made Stepanus Biondi Pramantara[*)], Agung Bagus Sista Satyarsa[*)], Nyoman Golden[*)], I Wayan Niryana[*)], Tjokorda Gde Bagus Mahadewa[*)], Sri Maliawan[*)], I Putu Pramana Suarjaya[**)]

[*)]Neurosurgery Division, Department of Surgery, Faculty of Medicine Universitas Udayana – Prof. Dr. I.G.N.G. Ngoerah General Hospital Bali, [**)]Department of Anesthesiology and Intensive Therapy, Faculty of Medicine Universitas Udayana–Prof. Dr. IGNG Ngoerah Hospital, Bali

Received: April 11, 2024; Accepted: October 03, 2024; Publish: October 24, 2024

Correspondences: niryana@gmail.com

Abstract

Pilocytic astrocytoma is a type of low-grade glioma that can develop in any part of the central nervous system. It primarily affects individuals in the pediatric and young adult age groups. Optic nerve pilocytic astrocytoma is an uncommon, gradually developing type of brain tumor known as a glioma. It is categorized as a grade I tumor by the World Health Organization (WHO). We aim to report rare optic nerve pilocytic astrocytomas and discuss their clinical findings and interconnection with the current literature. A 7-year-old male patient exhibited left-sided eye proptosis and complete loss of vision. A magnetic resonance imaging (MRI) performed before surgery showed the presence of a tumor in the left eye socket. The mass has a spherical shape and smooth boundaries, resulting in the compression of the left optic tract from behind. The left optic tract appears darker on T1-weighted images and brighter on T2-weighted images. On contrast administration, there is heterogenous contrast enhancement. A total resection of the mass was performed. Histopathology results show pilocytic astrocytoma (WHO Grade I). Optic nerve pilocytic astrocytoma is a tumor that can affect the optic nerve pathway. Early diagnosis and multidisciplinary team management are required. Treatment must be individualized, and the options include chemotherapy, radiotherapy, and surgical intervention. Surgery is only recommended in cases of painful or disfiguring proptosis and exposure keratopathy in eyes with severe vision impairment.

Keywords: Optic nerve pilocytic astrocytoma, surgical intervention, visual loss

J. neuroanestesi Indones 2024;13(2): 136–40

I. Introduction

Pilocytic astrocytoma is a type of brain tumor that mainly affects children and young people. It is classified as a low-grade glioma. Pilocytic astrocytoma of the optic nerve, primarily found in children, constitutes 3–5% of all brain tumors in this age group. Both men and women are affected with equal frequency. The average age of diagnosis was 7.0 years, and 90% of patients were diagnosed before to reaching the age of 19.[1,2] Optic nerve pilocytic astrocytomas generally target the visual pathway. They might start in orbit with the optic nerve and then spread to the optic chiasm and the frontal brain lobe. These tumours typically manifest as fusiform swelling, bound by the dura, and they have unique imaging and histologic traits. Optic nerve pilocytic astrocytomas are frequently connected with Neurofibromatosis Type I (NFI).[1,2] Optic nerve pilocytic astrocytomas are uncommon gliomas that grow slowly and are classed as grade-I tumors by the World Health Organization (WHO). The signs and symptoms of this condition typically last for several months and are closely linked to the size, location, and occurrence of hydrocephalus associated with the tumor. Visual loss is the most prevalent symptom by a significant margin. Optic nerve pilocytic astrocytomas usually present as a well defined cystic mass with a distinct nodule on the surface.[3]

doi: https://doi.org/10.24244/jni.v13i3.594
ISSN (Print): 2088-9674 ISSN (Online): 2460-2302

JNI is accredited as Sinta 2 Journal: https://sinta.kemdikbud.go.id/journals/profile/796

How to cite: Pramantara IMSB, et al, "Optic Nerve Pilocytic Astrocytoma in a Pediatric Patient: A Case Report".

The difficulties in treating optic nerve pilocytic astrocytomas stem from the intricate nature of their symptoms and their proximity to vital tissues. Optic nerve pilocytic astrocytomas, because to their proximity to important brain structures and involvement with visual pathways, sometimes result in substantial neurological damage when total removal or optimal radiation dosage is attempted. Consequently, the treatment of optic nerve pilocytic astrocytomas must be tailored to each individual.

Unless there is evidence of tumor growth or the presence of visual complaints, most patients are typically monitored through a series of imaging tests during a period of observation. Possible treatments encompass chemotherapy, radiation, and surgical intervention. Surgery is solely advised for instances of painful or disfiguring proptosis and exposure keratopathy in eyes with significant visual impairment.[2,4] In this article, we aim to report rare optic nerve pilocytic astrocytomas and discuss their clinical findings and interconnection with the current literature.

II. Case

History

A 7-year-old boy who had lost all eyesight and suffered left eye proptosis for two years was brought to the hospital. There was no trauma or significant systemic illness history. The patient, however, had an uneventful prenatal, natal, and postnatal account.

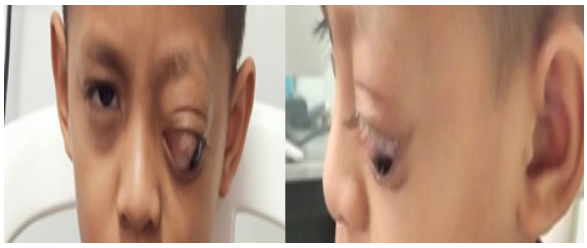

Figure 1. Preoperative image as we can observe exposure keratopathy, significant pro-ptosis, and lateral and downward globe displacement

Informed consent was obtained from the patient's family for the publication of this case report and any accompanying images, ensuring that confidentiality is maintained and no identifying information is disclosed.

Physical Examination

Upon examination, the left eye had no perception of light. There was exposure keratopathy, significant proptosis, and lateral and downward globe displacement. In all gazes, there was a restriction on eye movement. (Figure 1)

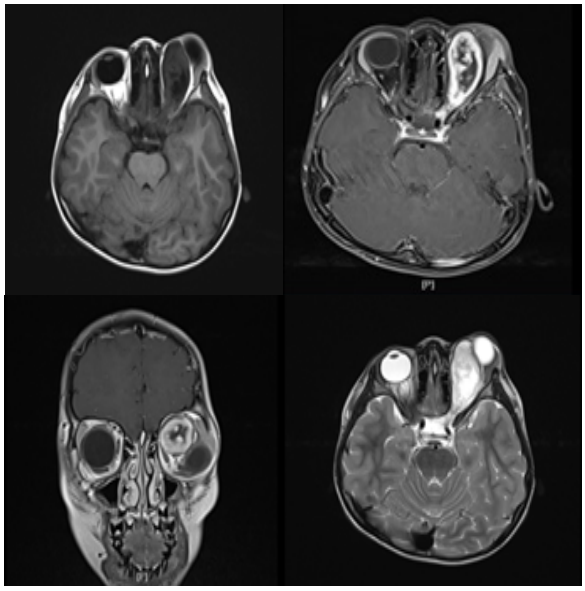

Figure 2. Preoperative MRI shows a mass in the left pars intraorbital to pars intracranial, measured +/- 2.2x5,2x2.4 cm. The mass appears round with regular edges, causing posterior compression of the left optic tract, which appears hypointense on T1WI (a), and hyperintense on T2WI, and contrast administration, there is heterogenous contrast enhancement (b,c,d).

Supporting Examination

Preoperative MRI revealed a mass in the left pars intraorbital to pars intracranial, measured +/- 2.2x5, 2x2.4 cm. The mass appears to be round with regular edges causing posterior compression of the left optic tract, which appears hypointense on T1WI, and hyperintense on T2WI, and contrast administration, there is heterogenous contrast enhancement (Figure 2).

Anesthesia Management

Tumour removal was performed through a hemicoronal skin incision (Figure 3). A supraorbital orbitotomy was performed, followed

by an incision in the orbital fascia, and then periorbital fat was removed while the patient was under general anesthesia. General anesthesia management included sedation with propofol continuous infusion at 50 – 50 mcg/kg/hour and sevoflurane at 1 – 2%. For analgesia, fentanyl 25 mcg was administered every 30 – 45 minutes, and for muscle relaxation, rocuronium 0.15 mcg/kg was given every 30 – 45 minutes. The patient was monitored throughout the surgery with blood pressure charts, pulse rate, respiratory rate, and partial pressure of carbon dioxide (PCO_2).

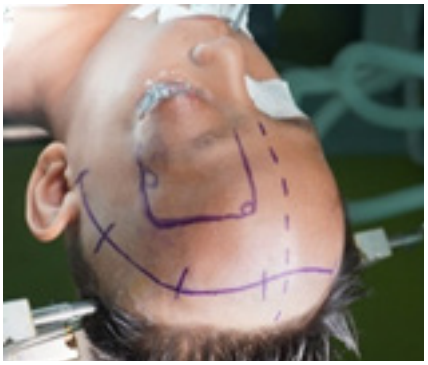

Figure 3. Semicoronal skin incision

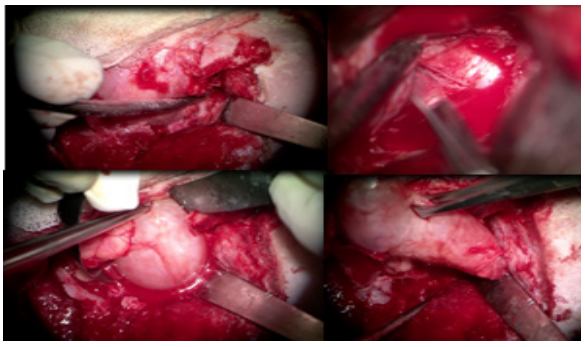

Figure 4. Supraorbital orbitotomy (a), incision in the orbital fascia (b), tumour identification (c) and complete tumour resection was performed (d)

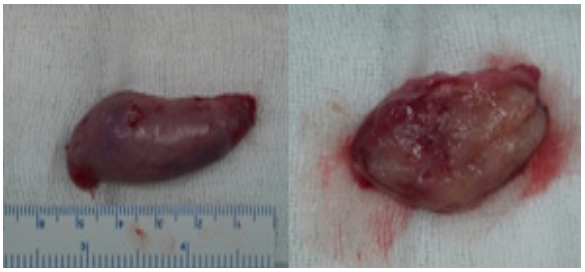

Figure 5. Complete Tumour Removal and Size of the Tumour

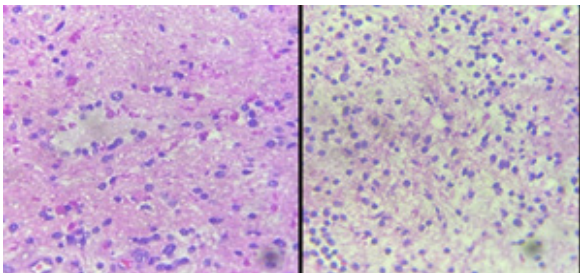

Figure 6. Histopathology examination result showed bipolar cells with hair-like processes and Rosenthal fibers

Ventilation mode was carefully adjusted. Then, we made tumour identification, and a complete tumour resection was performed using blunt and sharp dissection (Figures 4 and 5). Upon microscopic analysis (Figure 6), it was observed that the tumor tissue displayed a biphasic pattern, with dense and loose patches. The tumor was densely populated with bipolar cells that had rounded nuclei, fine chromatin, and lengthy processes. On the other hand, the loose region consisted of multipolar cells that formed microcystins. Rosenthal fibres are located within compacted areas. The diagnosis of pilocytic astrocytoma was confirmed based on microscopic observations.

Post-surgical Management

Intravenous methylprednisolone 250mg q.i.d for 3 days was administered followed by oral prednisone 50mg for 11 days. On the fifth day of the hospital stay, the patient was released with no changes to their ophthalmologic condition. The third day after being discharged, the patient went back to the outpatient clinic.

III. Discussion

Optic nerve pilocytic astrocytoma is a low-grade glioma tumour that usually occurs in children with a median diagnosis of seven years, and most patients are diagnosed before 19 years old. The incidence of this tumour in children is about 3 – 5% in pediatric brain tumours.[1,2] Optic nerve pilocytic astrocytoma, usually associated with a high incidence of NF1, has long been recognized. An inactive mutation causes this autosomal genetic disease in the tumour suppressor gene encoding neurofibromin, which

results in the stimulation of RAS signalling and the subsequent risk of developing RAS-induced tumours.[2] Based on a systematic review of study data and meta-analyses, 48.7% of patients had known cases of NF-1.5 According to the average age of patients diagnosed with Optic nerve pilocytic astrocytoma, our patient was seven years old at the time of diagnosis. Optic nerve pilocytic astrocytoma tumours are incredibly uncommon and rarely observed in ordinary clinical practice with 83.7% of cases diagnosed in pediatric patients, most commonly between 1 – 4 years of age.[6] This tumour can grow anywhere in the optic route, including the optic-chiasmatic and hypothalamic areas, from the optic nerves to the occipital brain. They typically occur in the chiasmatic-hypothalamic area. However, regardless of the site, visual loss is the most prevalent symptom in patients. If the tumour appears from the anterior optic tract, it clinically manifests as uniocular loss of vision, strabismus and proptosis. Proptosis is often discrete, but it can sometimes be severe and linked to other issues, such as corneal ulcers and partial occlusion of the eyelid. Patients with NF1 have a higher prevalence of proptosis than people without NF1.[1–4] Upon examination, our patient showed signs of a left eye with no perception of light. In addition to lateral and downward globe displacement, there was exposure keratopathy and considerable proptosis. There were limitations on eye movement in each gaze. Optic nerve pilocytic astrocytoma on MRI are typically hyperintense on T2 imaging and hypo- to iso-intense on T1 scans. After gadolinium injection, more than 50% of tumours exhibit bright lesion enhancement.

Optic nerve pilocytic astrocytoma may only occur in some portions of the visual system, such as the optic nerve or the chiasm, or it may manifest more widely. Imaging reveals a well-defined nerve enlargement, sometimes with a twisted or kinked appearance, when a tumour is localized to the optic nerves.[2] Most of the mass detected by preoperative MRI was located in the pars intraorbital of the left orbit. The mass seems circular with regular margins and causes posterior compression of the left optic tract, which appears hypointense on T1WI and hyperintense on T2WI, and on contrast administration, there is heterogeneous contrast enhancement. Despite being low-grade tumours, Optic nerve pilocytic astrocytoma can behave aggressively, making treatment challenging and patient-specific. Observation, surgery, chemotherapy, and radiation therapy are the treatment modalities available for patients with optic nerve pilocytic astrocytoma. One of the disease's most difficult and controversial elements is deciding how to treat an optic nerve pilocytic astrocytoma patient. The current agreement is to treat the patient who exhibits signs of visual or neurological decline. As NF1-associated optic nerve pilocytic astrocytoma is known to be more indolent, children with NF1 should be evaluated with greater caution.[2,4,7,8]

No consensus exists about the role of surgery in the treatment of optic nerve pilocytic astrocytoma. Surgery is only recommended in cases of painful or disfiguring proptosis and exposure keratopathy in eyes with severe vision impairment. Complete resection can improve cosmetic appearance in optic nerve pilocytic astrocytoma, where the tumour is restricted to the optic nerve and a prominent orbital component, resulting in proptosis, total blindness, discomfort from exposure keratoconjunctivitis, and deformity.[2,4,7,8] In our patient, most of the mass was located in the pars intraorbital of the left orbit. The left eye was blind; there was exposure keratopathy, proptosis, and lateral and downward globe displacement. In all gazes, there was a restriction on eye movement. Total resection was performed during surgery, as well as complaints and cosmetic improvements. At a microscopic level, PA is characterized by its biphasic structure, which includes areas that are loosely packed and areas that are tightly packed. However, there is significant variation in the microscopic appearance of PA, and only a few tumors conform to this typical description. Tumors consist of bipolar spindle cells and usually contain Rosenthal fibers.[8-10] Still up for question is the origin of these Rosenthal fibres. Nevertheless, they most likely reflect degraded glial fibres.

The microscopic sign of an indolent tumour, known as "eosinophilic granular masses," is also

present in certain tumours. Hyalinized arteries, mitotic figures, and calcifications are frequently seen in PAs. In the past, vascular proliferation along the cystic component has been mistakenly interpreted as a marker of aggressive behaviour since it is often structured in a linear pattern.[2,11] The pathological analysis reveals the presence of cancer tissue exhibiting both dense and loose patches, resulting in a biphasic pattern. The compact region of the tumor consisted of bipolar cells with spherical nuclei, delicate chromatin, and elongated processes, whereas the less dense region consisted of multipolar cells that formed microcystins. Rosenthal fibres are located in regions of high density.[12]

IV. Conclusion

Optic nerve pilocytic astrocytoma affects the optic nerve pathway, necessitating early diagnosis and multidisciplinary management. Treatment approaches, including chemotherapy, radiotherapy, and surgical intervention, should be tailored to individual cases. Surgical intervention is recommended primarily for cases involving painful or disfiguring proptosis and exposure keratopathy in eyes with significant vision impairment.

Conflict of Interest

The authors declare that they have no conflicts of interest.

Funding
None.

Author Contribution
All authors participated in the writing of this article.

References

1. Menon SG, Raju VNJ, Bhandary SV, Addoor KR. Recurrent optic nerve pilocytic astrocytoma: A rare case. J Clin Diagn Res. 2017;11(5):ND03-ND04. doi:10.7860/JCDR/2017/26991.9824

2. Fried I, Tabori U, Tihan T, Reginald A, Bouffet E. Optic pathway gliomas: a review. CNS Oncol. 2013;2(2):143-159. doi:10.2217/cns.12.47

3. Apanisile I, Karosi T. Surgical management of pilocytic astrocytoma of the optic nerve: a case report and review of the literature. Case Rep Oncol Med. 2017:1-7. doi:10.1155/2017/4283570

4. Hill CS, Khan M, Phipps K, Green K, Hargrave D, Aquilina K. Neurosurgical experience of managing optic pathway gliomas. Child's Nerv Syst. 2021;37(6):1917-929. doi:10.1007/s00381-021-05060-8

5. Yousefi O, Azami P, Sabahi M, Dabecco R, Adada B, Borghei-Razavi H. Management of optic pathway glioma: a systematic review and meta-analysis. Cancers (Basel). 2022;14(19):4781. doi:10.3390/cancers14194781

6. Farazdaghi MK, Katowitz WR, Avery RA. Current treatment of optic nerve gliomas. Curr Opin Ophthalmol. 2019;30(5):356-63. doi:10.1097/ICU.0000000000000587

7. Tabash MA. Characteristics, survival and incidence rates and trends of pilocytic astrocytoma in children in the United States; SEER-based analysis. J Neurol Sci. 2019;400:148-52. doi:10.1016/j.jns.2019.03.028

8. Bornhorst M, Frappaz D, Packer RJ. Pilocytic astrocytomas. Handb Clin Neurol. 2016;134:329-44. Doi: 10.1016/B978-0-12-802997-8.00020-7

9. Salles D, Laviola G, de Moraes Malinverni AC, Stávale JN. Pilocytic astrocytoma: a review of general, clinical, and molecular characteristics. J Child Neurol. 2020;35(12):852-58. Doi: 10.1177/0883073820937225

10. Reis GF, Bloomer MM, Perry A, Phillips JJ, Grenert JP, Karnezis AN, Tihan T. Pilocytic astrocytomas of the optic nerve and their relation to pilocytic astrocytomas elsewhere

in the central nervous system. Mod Pathol. 2013;26(10):1279-87. Doi: 10.1038/modpathol.2013.79

11. Mahakul DJ, Premsagar IC. Primary pilocytic astrocytoma of cerebello-pontine angle in the pediatric age group: literature review and case report. Egypt J Neurosurg. 2023;38(1):25.

12. Avuthu OP, Salunkhe S, Patil MG, Buch AC, Mane SV, Chugh A. Pilomyxoid astrocytoma presenting with developmental regression: a case report. Cureus. 2024;16(8). Doi: 10.7759/cureus.67167