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## Case Report

# A case report of pilocytic astrocytoma mimicking meningioma on imaging <sup>☆</sup>

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## ABSTRACT

Pilocytic astrocytoma (PA) is categorized as grade I gliomas with a good prognosis. Although PA mostly occurs in the cerebellum, it also can occur in the orbital and mostly presents as a cystic tumor with a mural nodule. PA often presents in the second decade of life, with 75% occurring under the age of 20 years. This case report describes a 10-year-old boy presented a left eye tumor for over 3 years. MRI examination showed unrestricted intracanal lesions in the optic nerve, visible from the optic canal to the anterior with well-defined borders. The excised tumor specimen depicted a nodular tumor tissue, measuring 35 × 28 × 20 mm, weighing 11 grams, solid with gray and white. The microscopic examination showed a classical biphasic pattern including combinations of loose glial tissue and compact pyloid tissue. Histopathology result revealed a pilocytic astrocytoma.

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## Introduction

The orbital tumors histologically and etiologically diverse space-occupying lesions of the orbit. Orbital tumors may invade the orbit from surrounding structures. Orbital tumors often lead to the displacement of the eyeball, motility disturbance, diplopia, and visual field defects [1]. Radiologic imaging plays an important role in making a diagnosis and differentiating between the benign and likely ma-

lignant process. The most common malignant neoplasms in orbital pediatrics are rhabdomyosarcoma, followed by lymphoproliferative disorders, optic glioma, meningioma, and astrocytoma.

Pilocytic astrocytoma (PA) most often presents in the second decade of life, with 75% occurring before the age of 20 years. The World Health Organization (WHO) categorized PA as grade I glioma with a good prognosis [2]. PA arises in the optic nerve and tends to be dura-bound fusiform swellings with distinctive imaging and histological characteristics. They of-

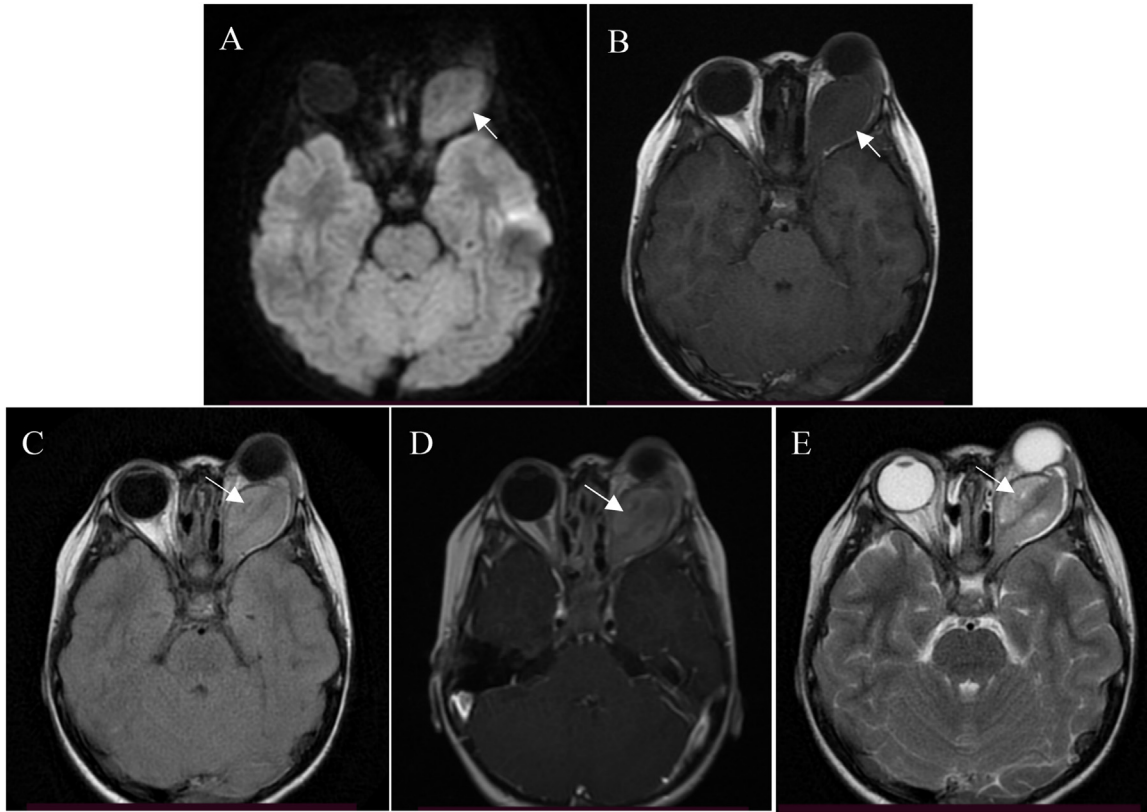
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**Fig. 1 – (A) DWI: Showed unrestricted intraconal lesions on the optic nerve, visible from the optic canal to the anterior with well-defined borders; (B) T1WI without Contrast: Isointense intensity with muscle, slight heterogenous and well demarcated. The lesion appeared to push the globe anteriorly, there was no infiltration with the orbital muscles; (C) Axial T2 Flair: An isointense intraconal lesion with orbital muscles, there was a slight hypointense line mimicking “tram track” calcification so that the initial diagnosis of optic nerve sheath meningioma; (D) Axial T2W: A slight hyperintense with hypointense intraconal lesion and a hypointense line suggesting tram track line sign; (E) T1W Contrast: A contrast-enhanced slight heterogenous intraconal lesion, well demarcated with a few hypointense areas in it, suggesting a necrotic area disorder.**

ten extend into optic chiasm and may be associated with neurofibromatosis type 1-3 [2].

There are wide spectrum of imaging features for PA. An avidly enhancing mural nodule has a significance to a cystic component found in 66%, while the cyst wall is also enhanced in 46% [3]. In this case report, we present a patient with PA in the retrobulbar region showing a solid and well-defined border on MRI, mimicking meningioma.

### Case report

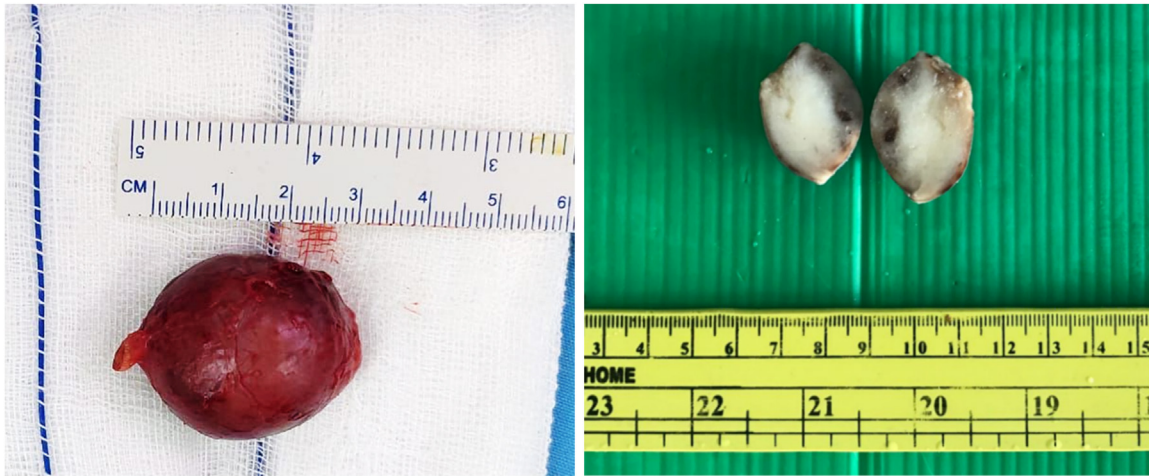
A 10-year-old boy presented with a tumor of the left eye for 3 years period. For the last 1 year, the tumor was getting prominent, an intermittent red and watery left eye accompanied by eyes glancing inward and reduced vision. The patient had been diagnosed with mild infection and only consumed pain-relieving drugs also some eye drops 4 times a day.

An objective condition showed VOS 4/60 (nasal side), Ishihara OS 0/38, eyeball movement (–2) inhibition in the superior quadrant, superotemporal, lateral, inferotemporal, (–1) in the

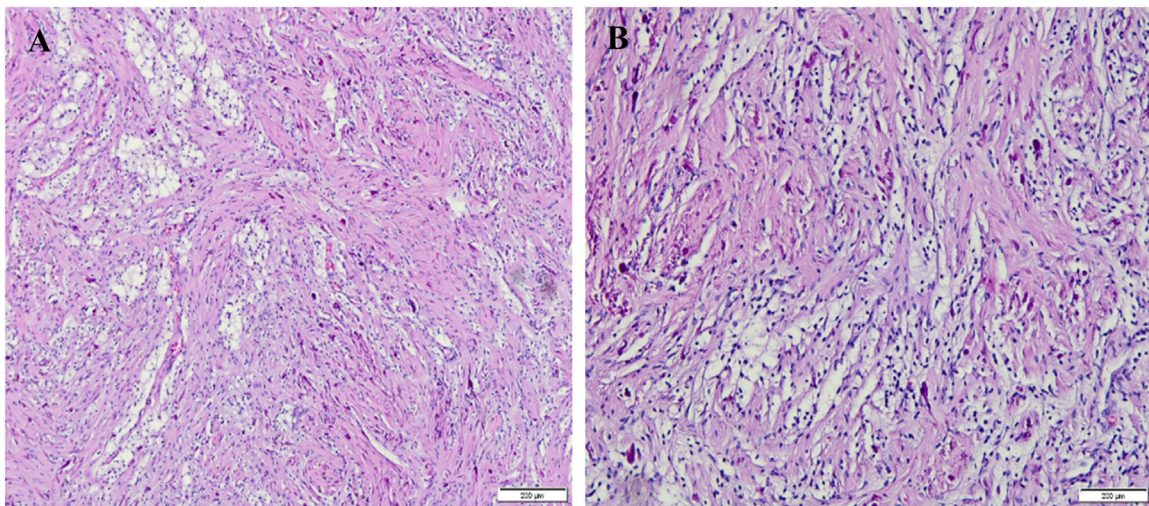
superomedial and inferior quadrant. OS globe displacement to inferonasal (+) also OS proptosis (+). Laboratory examination results were within normal limits.

The MR imaging showed a left intraconal slight heterogeneous solid and lobulated mass, with the size  $2.4 \times 2.8 \times 2.9$  cm, displaced the bulbus oculi anteriorly causing proptosis, and the optic canal looks widening. The differential diagnosis from CT imaging was optic glioma, lymphoma and meningioma. The MR imaging (Fig. 1) showed a solid mass measuring  $3.64 \times 2.26 \times 2.71$  cm in the left retrobulbar intraconal to the proximal side of the left optic nerve.

The lateral orbitotomy approach is appropriate for most intraconal masses. Our patient underwent tumor resection starting with a left superolateral orbital skin incision that was made deep to the periosteum. The macroscopic examination of excised tumor (Fig. 2) depicted a nodular tumor tissue, measuring  $35 \times 28 \times 20$  mm, and weighing 11 grams. The cut section showed a solid tumor with grayish-white. The microscopic examination (Fig. 3) revealed tumor tissue forming dense and loose areas, giving the appearance of a biphasic pattern. The dense area of the tumor was composed of bipolar cells with rounded nuclei, fine chromatin, and long processes;



**Fig. 2 – Macroscopically, the tumor was solid and well-circumscribed, with white and gray in color.**



**Fig. 3 – (A) Tumor showed dense and loose areas giving the appearance of biphasic pattern (H&E, 100x); (B) Rosenthal fibers are noticed within the dense areas of tumor (H&E, 200x).**

while the loose area was composed of multipolar cells forming microcysts. Rosenthal fibers are found within dense areas. Based on microscopic findings, a diagnosis of pilocytic astrocytoma was established.

## Discussion

Pilocytic Astrocytoma (PA) is a distinctive subtype of astrocytoma that occurs predominantly in children and adolescents younger than 20 years, and the incidence decreases with age [4]. The course of pilocytic astrocytoma is always over a period of several months. There are 4 main image patterns in the region of PA that have been described: (1) Tumors with augmentation on cyst wall and above wall nodules with strong enhancement (46% of cases); (2) Tumors with non-enhanced cyst and highly enhanced mural nodules (21% of cases); (3) Mainly solid mass minimal or no cystic component (17%); and (4)

Necrotic mass with the central non-concentrated zone (16%) [5]. Therefore, PA tends to show nodular enhancement with significant cyst components. Generally, these tumors have no peritumoral edema and obstructive hydrocephalus (usually from mild to moderate) occurs in the second half of the disease course [6–8].

The radiologic finding of orbital wall expansion provided additional evidence of a long-standing mass. The tumoral growth to a critical volume interfered with the ocular prosthetic fitting, which led to the diagnosis. PA on MRI is typically hypo- or isointense on T1 sequences and hyperintense on T2-weighted or FLAIR images and enhanced typically strongly and diffusely. In the posterior fossa, pilocytic astrocytoma may involve primarily the brainstem rather than the cerebellum, also the spinal cord can be affected [9,10]. On PA MR T1-weighted imaging, the tumor appears hypointense compared with orbital fat, but isointense is concerning for extraocular muscles. On T2-weighted imaging, the lesions are hyperintense to orbital fat and extraocular muscles. Areas of

chronic hemorrhage may show focal areas of increased signal on T1- and T2-weighted images. They typically demonstrate moderate contrast enhancement with gadolinium injection, particularly with fat suppression. Overall, several tissues of PA demonstrate high T1 signals: fat, subacute blood, and often melanin. Tissues with high fluid content (such as edema) demonstrate high T2 intensity. Fat-suppressed sequences are typically performed in the orbit to allow for differentiation of hyperintense structures from the underlying normal fat, for instance, in contrast-enhanced sequences. PA on DW Imaging and the apparent diffusion coefficient (ADC) help to evaluate ischemic tissues and cellular tumors like lymphomas, which can be bright on DWI and dark on ADC.

On histopathologic examination, the PA is characterized by intermingling, eosinophilic slender cell processes. Rosenthal fibers, which are thick elongated eosinophilic structures likely representing clumped intermediate filament proteins, are typically present. The term pilocytic refers to the hair-like appearance of bipolar tumor cells [11].

Our patient had a T1WI without contrast that showed isointense intensity with muscle, homogeneous and well-demarcated. The lesion appeared to push the globe anteriorly, there was no infiltration into the orbital muscles. The T1WI contrast showed a contrast-enhanced slight heterogeneous intraconal lesion, well demarcated with a few hypointense areas in it, suggesting a necrotic area disorder. The DW imaging of our patient showed an unrestricted intraconal lesion on the optic nerve, visible from the optic canal to the anterior with well-defined borders. This situation was in accordance with the theory states that PA tends to show accelerated diffusion, bright on ADC imaging and diffusion-weighted image (DWI) low signal helps to separate them from higher-grade lesions with restricted diffusion (dark on ADC and high signal on DWI). The same features are found in optic nerve sheath meningiomas on T1W and T2W imaging that shows homogeneous with the slight heterogenous “tram track” calcification distension posterior to the globe. Based on the imaging examination, we suspected that the tumor was a meningioma, so we carried out a further examination on anatomical pathology.

After establishing the tumor classification and lateral orbitotomy was performed, it was reported that *quo ad vitam* and *quo ad sanationam* were *dubia ad bonam*. In this case, we reported that the profile of orbital pilocytic astrocytoma was found different. It filled the distal part of the left optic nerves until the proximal part, pushing the oculi to the anterior. It consisted of a solid components, without any cystic part, and after contrast administration showed heterogeneous contrast enhancement. Also, we found a lesion that looks like a “tram track” sign in the middle of tumors, mimicking meningioma.

Histologically, the PA of our case has elongated, thin, hugely spindle-shaped tumor cells. PA shows a classical remarkable biphasic pattern including combinations of loose glial tissue and compact pyloid tissue. The latter include elongated bipolar high-density sheets cells that exhibit a fine fibrous (hair-like) process and typical abundance of rosen-

thal fibers. In contrast, loose glial tissue is built of multipolar cells, microcapsules and eosinophils granules. Based on those findings, a diagnosis of a pilocytic astrocytoma can be established.

## Conclusion

PA is the most common childhood brain tumor that may occur in the orbital. PA has a wide spectrum of imaging features, from a cystic tumor with mural nodules to solid and well-circumscribed tumor. PA should be considered in the differential diagnosis in the case of pediatric retrobulbar tumor.

## Patient consent statement

Informed consent obtained for publication of a case report.

Written informed consent was obtained from the patient for the publication of this case report.

## REFERENCES

- [1] Deveer M, Cullu N, Beydilli H, Sozen H, Yeniceri O, Parlak S. Spontaneous retrobulbar haematoma. *Hindawi Case Rep Radiol* 2020;1:3.
- [2] Mudhar H, Nuruddin M, Roy S. Primary orbital cystic pilocytic astrocytoma. *Ocul Oncol Pathol* 2016;2:280–4.
- [3] Tabash M. 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.
- [4] Komotor R J, Mocco J, Zacharia B E, Wilson D A, Kim P Y, Canoll P D. Astrocytoma with pilomyxoid features presenting in an adult. *Neuropathology* 2016;26(1):89–93.
- [5] Bornhorst M, Frappaz D, Packer R. Pilocytic astrocytoma. *Handbook Clin Neurol* 2016;134(3):329–44.
- [6] Wallner K, Gonzales M, Edwards M. Treatment results of juvenile pilocytic astrocytoma. *J Neurosurg* 1988;69(2):171–6.
- [7] PennMedicine. Orbital tumors. 2021. Retrieved from PennMedicine: <https://www.pennmedicine.org/for-patients-and-visitors/patient-information/conditions-treated-a-to-z/orbital-tumors>
- [8] AlRayahi J, Zapotocky M, Ramaswamy V, Hanagandi P, Branson H, Mubarak W, et al. Pediatric brain tumor genetics: what radiologist need to know. *Radiographics* 2018;38(7):2102–22.
- [9] Malik A, Deb P, Sharma M, Sarkar C. Neuropathological spectrum of pilocytic astrocytoma - an Indian series of 120 cases. *Pathol Oncol Res* 2006;12:164–71.
- [10] Murray R D, Penar P L, Filippi C G, Tarasiewicz I. Radiographically distinct variant of pilocytic astrocytoma: a case series. *J Comput Assist Tomogr* 2011;35:495–7.
- [11] Ironside W J, Moss T H, Luis D N, Lowe J S, Weller R O. Diagnostic pathology of nervous system tumours. *Churchill Livingstone*; 2002. p. 53–120.