

Parinaud Syndrome and Obstructive Hydrocephalus as Initial Manifestations of an AFP-Secreting Pineal Region Germ Cell Tumor: A Case Report

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ABSTRACT

A 14-year-old male presented with Parinaud syndrome, obstructive hydrocephalus, and markedly elevated serum AFP (272 ng/mL) and β -hCG (317 mIU/mL), suggesting a non-germinomatous germ cell tumor (NGGCT) of the pineal region. MRI confirmed a pineal mass with pituitary stalk involvement. Endoscopic third ventriculostomy (ETV) relieved hydrocephalus and enabled CSF tumor marker sampling. Four cycles of CE/IE chemotherapy achieved complete biochemical remission with significant neurological recovery. This case highlights the neuro-ophthalmic clues to early pineal GCT diagnosis and the effectiveness of ETV combined with chemotherapy.

Keywords: *alpha-Fetoproteins; Ocular Motility Disorders; Ventriculostomy*

INTRODUCTION

Intracranial germ cell tumors (GCTs) are uncommon, comprising less than 5% of primary brain tumors, occurring predominantly in the pineal and suprasellar regions of adolescent males. Parinaud syndrome combined with obstructive hydrocephalus as the inaugural presentation of a pineal GCT is rare and may mimic other intracranial pathologies, delaying diagnosis. Early recognition is critical given the high chemosensitivity and excellent outcomes when treated promptly. This case is distinctive

due to its markedly elevated AFP, indicating NGGCT, creating diagnostic and therapeutic challenges uncommon in typical pineal tumors. This report adheres to SCARE 2025 and TITAN guidelines. ^{1,2,3,4,5,6}

CASE REPORT

A 14-year-old male presented with one month of persistent headache, blurred vision, and vomiting. On examination, he had a low GCS (GCS 5/15) with a spastic right lower limb. Cranial nerve assessment demonstrated light-

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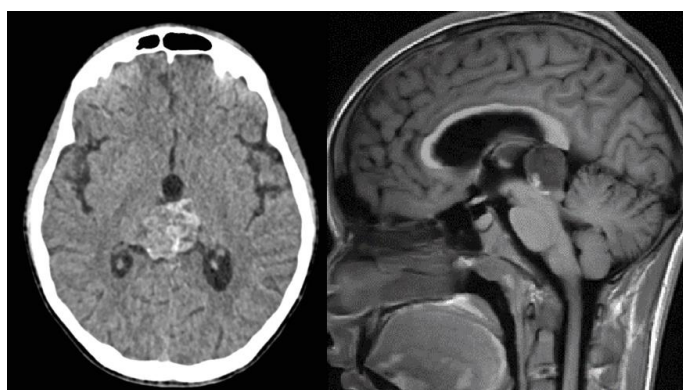


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near dissociation, limitation of upward gaze, and convergence-retraction nystagmus, consistent with Parinaud syndrome. Visual acuity was 6/18 (right) and 6/9 (left). Focal seizures were present on admission. Both testes were normal with no lymphadenopathy.

Serum β -hCG was markedly elevated at 317.40 mIU/mL and AFP at 272.35 ng/mL. Routine bloods were otherwise unremarkable.

Brain MRI revealed a 12×9×13 mm pineal mass: T1 hypointense, T2/FLAIR hyperintense, with peripheral rim enhancement, pituitary stalk thickening (3.2 mm), and obstructive hydrocephalus (Figures 1, 2). CT confirmed internal calcifications. Whole-spine MRI demonstrated no leptomeningeal dissemination. Systemic staging CT and scrotal ultrasound were negative for metastasis.

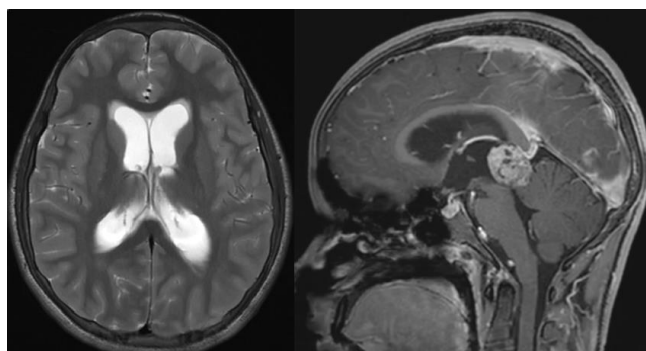


1a

1b

Figure 1:1a Axial Plain CT image of the brain shows a well-defined hyperdense solid mass lesion in the region of the pineal gland. The mass shows internal calcifications within.

1b: Sagittal T1WI shows a well-defined T1 intermediate to low signal intensity mass lesion within the pineal gland, showing T1 high signal intensity areas in the anterior aspect, suggesting calcifications. The mass is causing compression of the tectal plate and posterior midbrain.



2a

2b

Figure 2: 2a: Sagittal post-contrast T1WI shows heterogeneous enhancement of the mass with some non-enhancing areas. **2b:** Axial T2WI shows mild dilatation of the lateral ventricles with effacement of the subarachnoid spaces, along with peri-ependymal T2 high signal intensity, suggesting active hydrocephalus

Focal seizures were controlled with levetiracetam, phenytoin, and clobazam. ETV was performed via a right frontal burr hole; a translucent, thinned third ventricular floor overlying the interpeduncular tumor was identified (Figure 3) and fenestrated anterior to the mammillary bodies using a Fogarty balloon catheter. Intraoperative CSF confirmed elevated β -hCG.

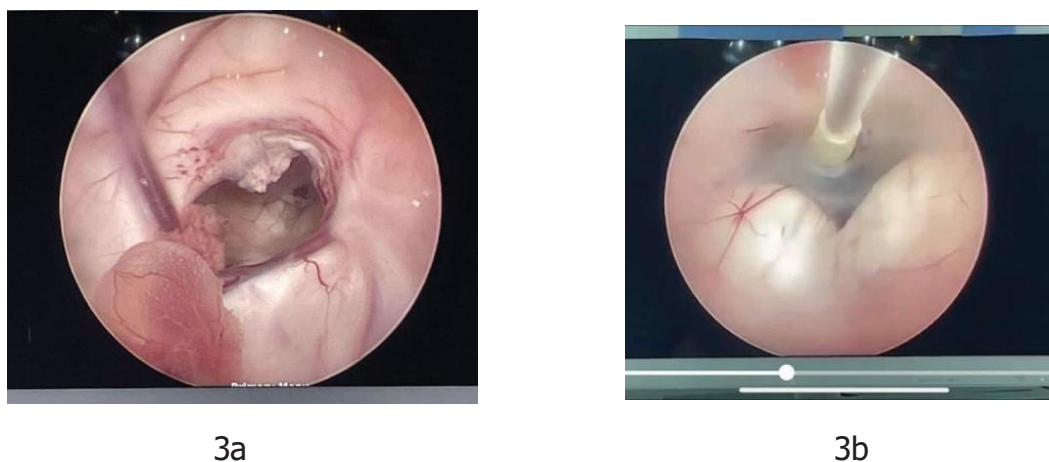


Figure 3: Intraoperative endoscopic views during endoscopic third ventriculostomy

3a: Visualization of the dilated third ventricle with a translucent and thinned-out floor overlying the tumor mass in the interpeduncular cistern.

3b: Fenestration of the third ventricular floor anterior to the mammillary bodies using a Fogarty balloon catheter, confirming cerebrospinal fluid (CSF) flow into the prepontine cistern

CE/IE chemotherapy was then initiated: Cycle 1 - carboplatin (AUC 5) and etoposide (100 mg/m², Days 1–3); Cycle 2 - ifosfamide (1800 mg/m²/day) and etoposide (100 mg/m²/day) for five days with mesna uroprotection. After four cycles, AFP fell below 10 ng/mL and β -hCG to <5 mIU/mL (complete biochemical remission). The patient achieved progressive neurological recovery with resolution of diplopia and normalization of eye movements by week 12.

DISCUSSION

Parinaud syndrome from dorsal midbrain compression - upward gaze palsy, light-near dissociation, and convergence-retraction nystagmus - is classic for pineal masses but frequently misattributed to stroke or demyelinating disease. Concurrent obstructive hydrocephalus from aqueductal compression amplifies intracranial hypertension and further obscures the diagnosis. Their coexistence in an adolescent male should prompt urgent evaluation for a pineal mass.^{8,9,10}

Elevated serum and CSF tumor markers often suffice to establish GCT diagnosis without biopsy. MRI typically shows a well-defined midline lesion with T1 hypointensity, T2/FLAIR hyperintensity, and contrast enhancement. Per WHO 2021 CNS classification, AFP >10 ng/mL excludes pure germinoma and indicates NGGCT or mixed GCT. ETV provides dual benefit: hydrocephalus relief and simultaneous CSF sampling. CE/IE was preferred over standard BEP given neurological instability and borderline

renal reserve, minimizing neurocognitive and endocrine sequelae while endorsed by COG and Japanese protocols. Five-year survival exceeds 90% for germinomas and reaches 70% for NGGCTs; long-term follow-up remains essential.⁶

Three key learning points emerge. First, Parinaud syndrome with hydrocephalus in an adolescent should strongly raise suspicion for GCT. Second, serum and CSF tumor markers can establish a diagnosis without biopsy. Third, early ETV expedites both symptomatic relief and diagnostic confirmation. Markedly elevated AFP heralding an NGGCT phenotype and excellent response to CE/IE make this a valuable teaching case, particularly in resource-limited settings

CONCLUSION

Parinaud syndrome with obstructive hydrocephalus in an adolescent male should prompt urgent evaluation for pineal GCT. Serum and CSF tumor markers can establish a diagnosis and guide treatment without biopsy. Early ETV provides both symptomatic relief and

diagnostic confirmation. Markedly elevated AFP indicating NGGCT and excellent response to CE/IE chemotherapy make this case a valuable clinical reference, especially in resource-limited settings..

CONFLICT OF INTEREST

None

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