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Case report

# Supra-sellar clear cell ependymoma in a 2-year-old female: A case report

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ABSTRACT

*Introduction and importance:* Clear cell ependymomas (CCEs) are rare glial tumors, with suprasellar localization being extremely uncommon in pediatric populations. Their nonspecific presentation often overlaps with more common tumors such as optic pathway gliomas and craniopharyngiomas, making early and accurate diagnosis particularly challenging.

*Case presentation:* A 2-year-old female presented with progressive visual disturbances and developmental delay. MRI revealed a T2-hyperintense, contrast-enhancing lobulated sellar and suprasellar mass. The initial radiologic impression favored optic pathway glioma. A transcranial approach enabled maximal safe resection. Histopathological analysis confirmed clear cell ependymoma, showing clear cytoplasm, perivascular pseudorosettes, and GFAP positivity. Critical structures including the optic nerves, pituitary stalk, and hypothalamus were preserved.

*Clinical discussion:* Suprasellar CCEs are exceptionally rare in children and can mimic more prevalent pediatric brain tumors radiologically. Histopathologic examination remains essential for definitive diagnosis. The lesion's location necessitates careful surgical planning to minimize morbidity. Due to the potential for recurrence, adjuvant therapy and vigilant long-term follow-up are recommended.

*Conclusion:* This case highlights the importance of including rare tumors like CCE in the differential diagnosis of pediatric suprasellar masses. A multidisciplinary approach involving imaging, histopathology, surgery, and adjuvant therapy is essential for accurate diagnosis and optimal patient outcomes.

## 1. Introduction

Ependymomas are glial tumors originating from ependymal cells lining the ventricles of the brain and the central canal of the spinal cord. While ependymomas are relatively common in pediatric neuro-oncology, clear cell ependymomas (CCEs) represent a rare histological variant. These tumors are characterized by clear cytoplasm within tumor cells, often exhibiting distinct pseudovascular patterns. CCEs are typically found in the spinal cord or posterior fossa, with suprasellar occurrences being exceedingly rare in pediatric patients [1]. Only a limited number of cases have been documented in the literature. As per recent literature, CCEs in the suprasellar region are exceedingly rare, with only a handful of cases reported, typically misdiagnosed as optic pathway gliomas or craniopharyngiomas. This underscores the importance of including this diagnosis in differential diagnoses for pediatric patients with visual disturbances and developmental delay. Reports suggest that the difficulty in diagnosing these tumors lies in their nonspecific clinical presentations and radiological overlap with more common suprasellar tumors, such as gliomas and craniopharyngiomas [2]. This case report presents the clinical course, diagnostic challenges, and treatment approach for a 2-year-old female diagnosed with suprasellar clear cell ependymoma, underscoring the importance of early recognition and intervention in rare pediatric brain tumors [3].

## 2. Case presentation

A 2-year-old female was brought to the neurosurgery department with a history of progressive visual disturbances and developmental delay. Her parents noted reduced engagement with visual stimuli and occasional irritability over the past few months. The child also exhibited signs of poor visual tracking and difficulty with eye contact, suggestive of early optic pathway involvement. There was no family history of

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neurological disorders or congenital anomalies.

On physical examination, the patient had a normal head circumference (95th percentile for age) and demonstrated mild developmental delay. No focal neurological deficits were observed. Her other cranial nerves examination were within normal limit, and motor and sensory examinations were unremarkable. Detailed ophthalmologic examination revealed no papilledema, but abnormal visual evoked potentials were noted, suggesting optic pathway involvement.

MRI revealed a hyperintense lobulated suprasellar and sellar mass on T2W image (Fig. 1) A1, A2 and A3 that was initially thought to be an optic pathway glioma. Head and neck CT- angiography revealed a suprasellar and right parasellar hypodense mass with no significant contrast enhancement on arterial post-contrast phase (Fig. 2.A). Given the clinical and radiologic findings, surgical biopsy was advised because the possibility of other pathology could not be ruled out. The MRI also showed no signs of intracranial CSF dissemination otherwise the spine MRI was not done to exclude extracranial CSF dissemination.

A transcranial approach was used to access the tumor, allowing for maximum safe resection while preserving critical structures such as the optic nerves, pituitary stalk, and hypothalamus.

Histopathological analysis of the tumor revealed a well-circumscribed mass composed predominantly of clear cells arranged in sheets and nested patterns. The tumor cells displayed round nuclei with well-defined, vacuolated cytoplasm. Notably, pseudopapillary and pseudovascular architectures were observed, and the tumor cells demonstrated strong, diffuse cytoplasmic staining for GFAP (Glial Fibrillary Acidic Protein) on immunohistochemical analysis, consistent with the histopathological features of clear cell ependymomas (Fig. 3).

Postoperative imaging showed marked reduction in the volume of the lesion as compared to previous MRI with post-surgical defects and cavitation (Fig. 2.B–E).Given the possibility of residual tumor, adjuvant radiation therapy was planned. The patient was referred to the oncology department but was subsequently lost to follow-up.

## 3. Discussion

Clear cell ependymomas are typically managed with a combination of surgical resection and adjuvant therapy. In this case, a transcranial approach was used, and safe resection was achieved; however, due to the suprasellar location, histological grade, and potential for recurrence, adjuvant radiation therapy was recommended. Chemotherapy is generally reserved for pediatric ependymomas with significant residual disease or when radiation therapy is contraindicated due to patient age [4].

Surgical resection of suprasellar tumors presents unique challenges due to the close proximity to critical structures, including the optic chiasm, hypothalamus, and pituitary gland. Meticulous surgical planning and intraoperative neurophysiological monitoring were crucial in minimizing damage to these vital areas. The prognosis of pediatric ependymomas depends on multiple factors, including tumor location, extent of resection, and presence of residual disease. The 5-year survival rate for pediatric ependymomas is generally favorable if complete resection is achieved. However, recurrence remains a concern, particularly for supratentorial tumors [5]. The case highlights the importance of precise tumor resection and the necessity of radiation therapy to minimize the risk of recurrence.

The role of chemotherapy in ependymomas remains an area of ongoing research. It is typically reserved for cases with residual or recurrent disease following surgery and radiation. Given the rarity of suprasellar clear cell ependymomas, long-term follow-up is essential to monitor for recurrence and late treatment-related complications [6]. Recent studies also emphasize the potential benefits of molecular profiling and genetic testing to guide treatment, although this remains an area under active investigation [7].

## 4. Conclusion

This case highlights the rarity and clinical challenges associated with suprasellar clear cell ependymomas in pediatric patients. This report emphasizes the importance of early diagnosis, careful differential diagnosis (including optic pathway glioma and craniopharyngioma), and timely intervention to optimize patient outcomes. Diagnosis relies on a combination of neuroimaging and histopathology, with immunohistochemical studies confirming the tumor's glial origin. Early diagnosis, precise surgical management, and consideration of adjuvant therapies are critical for optimizing patient outcomes. Due to the risk of recurrence, long-term follow-up with periodic neuroimaging including spine imaging is essential. Increasing awareness of such rare tumors can aid in timely diagnosis and improved management strategies in pediatric neuro-oncology.

## SCARE guidelines

This work has been reported in accordance with the SCARE criteria (Surgical Case Report Guidelines) [8].

## Patient consent

Written informed consent was obtained from the patient's guardians for publication and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal upon request.

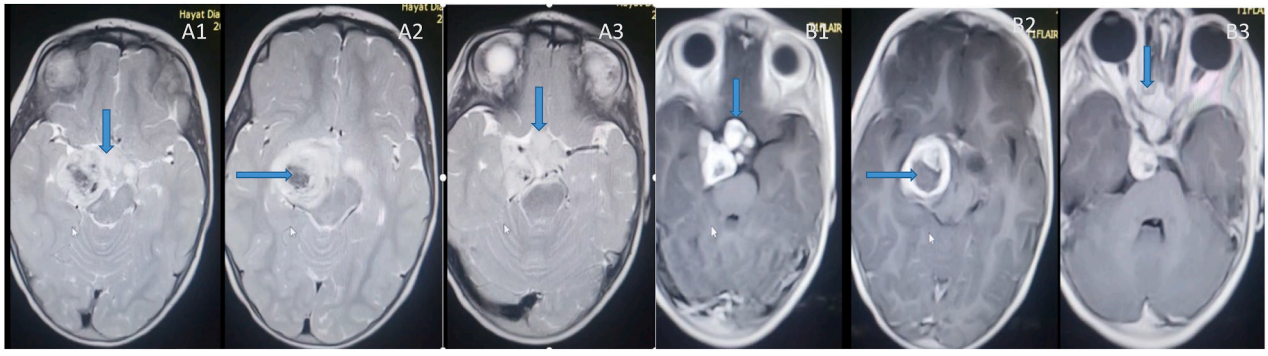



**Fig. 1.** Brain MRI.
A. Axial T2-weighted images show a multilobulated hyperintense suprasellar and right parasellar mass (vertical arrows, A1 & A3) with a focal hypointense area suggestive of necrosis (horizontal arrow, A2).
B. Post-contrast axial T1-FLAIR images reveal intense enhancement of the mass (vertical arrows, B1 & B3) and a non-enhancing focus (horizontal arrow, B2) corresponding to the necrotic region. The mass is separate from the pituitary on sagittal T1+C (not shown).

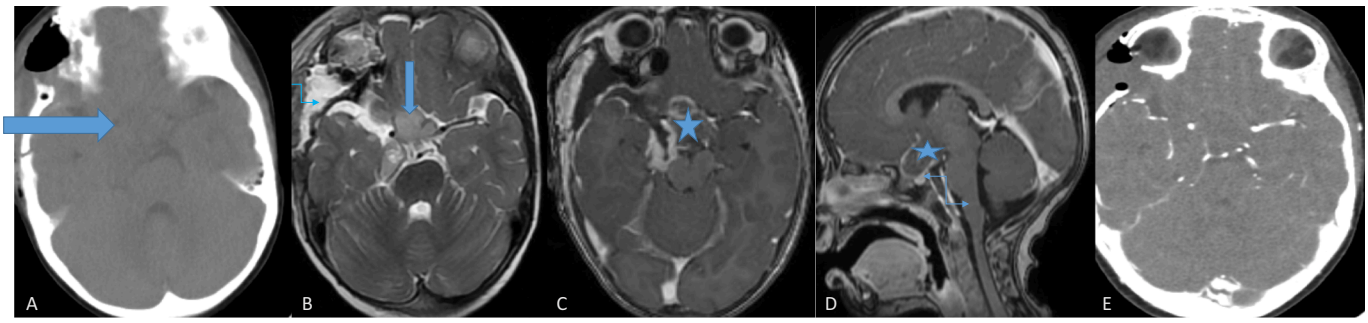



**Fig. 2.** Immediate post-operative non-contrast axial head CT with soft window showed isodense to hypodense mass on the suprasellar location with no calcification (horizontal arrow) (A). Axial T2-weighted image showed residual lobulated hyperintense lesion (vertical arrow) with marked interval reduction in size and right frontal and temporal skull acute post-surgical fracture and scalp fluid collection (elbow arrow) (B) and suprasellar and right parasellar multilobed mass which has post-surgical cavitation and showed peripheral contrast enhancement indicated by star on Ax and Sag T1+C images (C &D) and contrast enhanced pituitary gland separately visualized and indicated by connector double elbow arrow on Sag T1+C (D) and lesion showed no significant enhancement on early arterial phase of CTA (E).

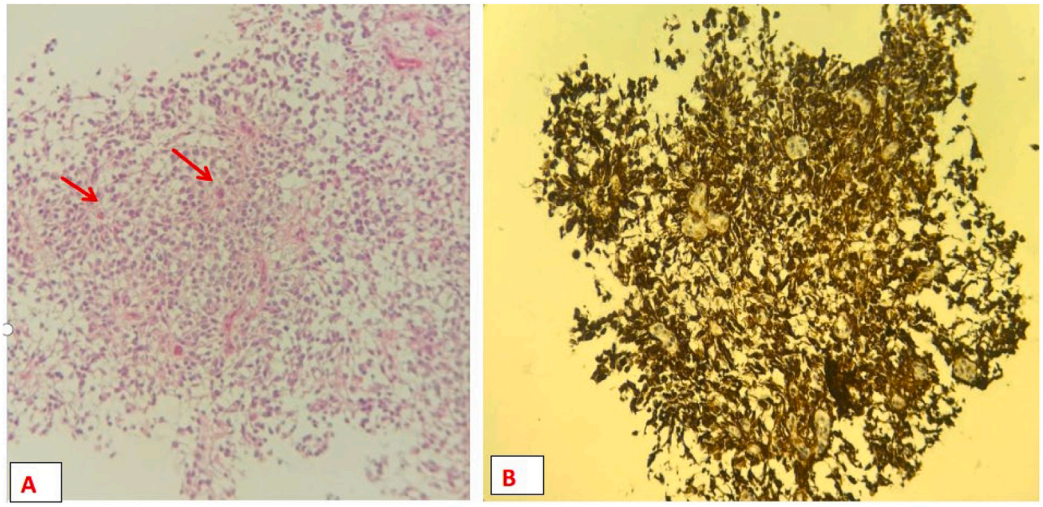



**Fig. 3.** Histopathological features of clear cell ependymoma.
The tumor is composed of cells with pale, well-defined cytoplasm and round to oval nuclei, arranged around blood vessels forming numerous perivascular pseudorosettes (arrow) (a) H&E-stained section, 20× magnification. (b) Immunohistochemistry demonstrates strong and diffuse cytoplasmic positivity for GFAP, supporting glial differentiation; GFAP immunostaining.

## Ethical approval

This study was approved by the Research Ethical Review Committee of Ethio Istanbul Hospital with a reference number (EIH-026/24) and approval date of 07/01/24.

## Guarantor

Wubshet Assefa Tesema

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No external funding was received for this study.

## Author contribution

WT, MM, and AW conceived the study, and wrote the manuscript for publication. All the authors critically read and approved the final manuscript.

## Conflict of interest statement

The authors declare no conflicts of interest.

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