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RESEARCH ARTICLE

ANAPLASTIC EPENDYMOMA (WHO GRADE III) OF THE FOURTH VENTRICLE IN A CHILD: CASE REPORT AND LITERATURE REVIEW

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Abstract

Anaplastic ependymoma (WHO grade III) is an uncommon and aggress ive glial tumor in children, most often located in the posterior fossa, typically arising from the fourth ventricle. We report the case of a 6-year-old boy who presented with persistent headaches and recurrent vomiting.Brain MRI revealed a heterogeneously enhancing fourth-ventricle mass causing triventricular hydrocephalus.The patient underw ent gross total resection followed by adjuvant conformal radiotherapy with a total dose of 54 Gy. The postoperative evolution was favorable, with no neurological deficits or radiological signs of recurrence at follow up. This observation emphasizes the importance of early diagno sis, multidisciplinary management, and complete surgical removal combined with adjuvant radiotherapy in improving outcomes for pediatric anaplastic ependymomas.

Introduction:-

Ependymomas are rare glial tumors derived from ependymal cells of the ventricular system and spinal canal. In children, they mainly arise in the posterior fossa, most often within the fourth ventricle (1). The anaplastic form (WHO grade III)shows higher mitotic activity, vascular proliferation, and necrosis, leading to a greater risk of recurrence and poorer survival (2-3). Management requires **a** multidisciplinary approach, including cerebrospinal fluid diversion when needed, maximal safe resection, and postoperative radiotherapy. Recent molecular studies identifying PF-A and PF-B subgroups have improved prognostic assessment (3-4).We report the case of a six-year-old boy diagnosed with a fourth ventricle ependymoma, who underwent complete surgical resection followed by adjuvant radiotherapy at a total dose of 54 Gy, with a favorable clinical and radiological outcome.

Case Presentation:

The patient was a 6-year-old child with no significant personal or family medical history. Symptoms began approximately 2 months prior to admission, marked by the gradual onset of vomiting and progressive headaches that did not respond to symptomatic treatment. As the condition worsened, the child was referred to a neurosurgery department for further evaluation.On initial physical examination, the child was classified as WHO performance status 1, with stable hemodynamic and respiratory parameters. Neurological assessment revealed a conscious patient with a Glasgow Coma Scale (GCS) score of 15. The child was able to stand and walk independently.Cranial nerve

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examination was normal. Motor function was preserved, with muscle strength rated 5/5 in all four limbs. No signs of cerebellar syndrome were observed, and deep tendon reflexes were present and symmetrical.
Brain magnetic resonance imaging (MRI) showed a fourth ventricle mass exerting a mass effect and causing mild upstream hydrocephalus with slight transependymal resorption, findings highly suggestive of a fourth ventricle ependymoma.

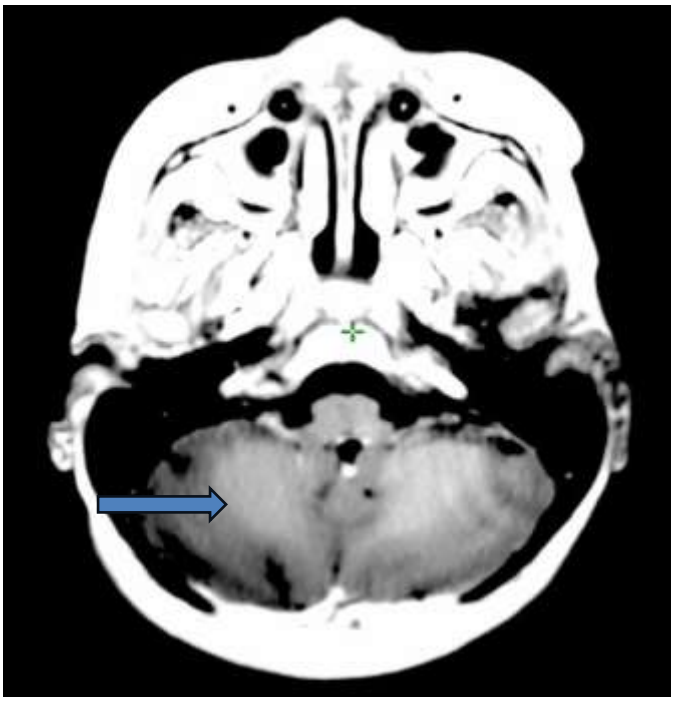


Figure 1. **MRI findings at diagnosis, with the blue arrow highlighting the fourth ventricle ependymoma.**

The patient initially underwent a ventriculo-cisternostomy (VCS), which was performed without any intraoperative complications.This was followed by what was considered a complete surgical resection of the lesion.
Histopathological and immunohistochemical examination confirmed the diagnosis of a Grade III anaplastic ependy moma.
Postoperative craniospinal MRI revealed a mild triventricular dilatation, more pronounced on the right side. Postsurgical changes were observed in the posterior fossa, with no definite residual tumor identified. Spinal MRI findings were normal, showing no evidence of metastatic spread or other abnormalities**.**

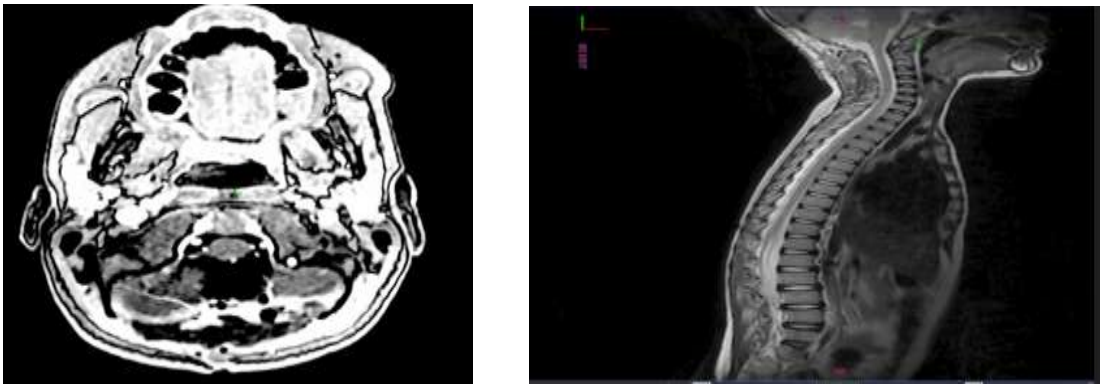
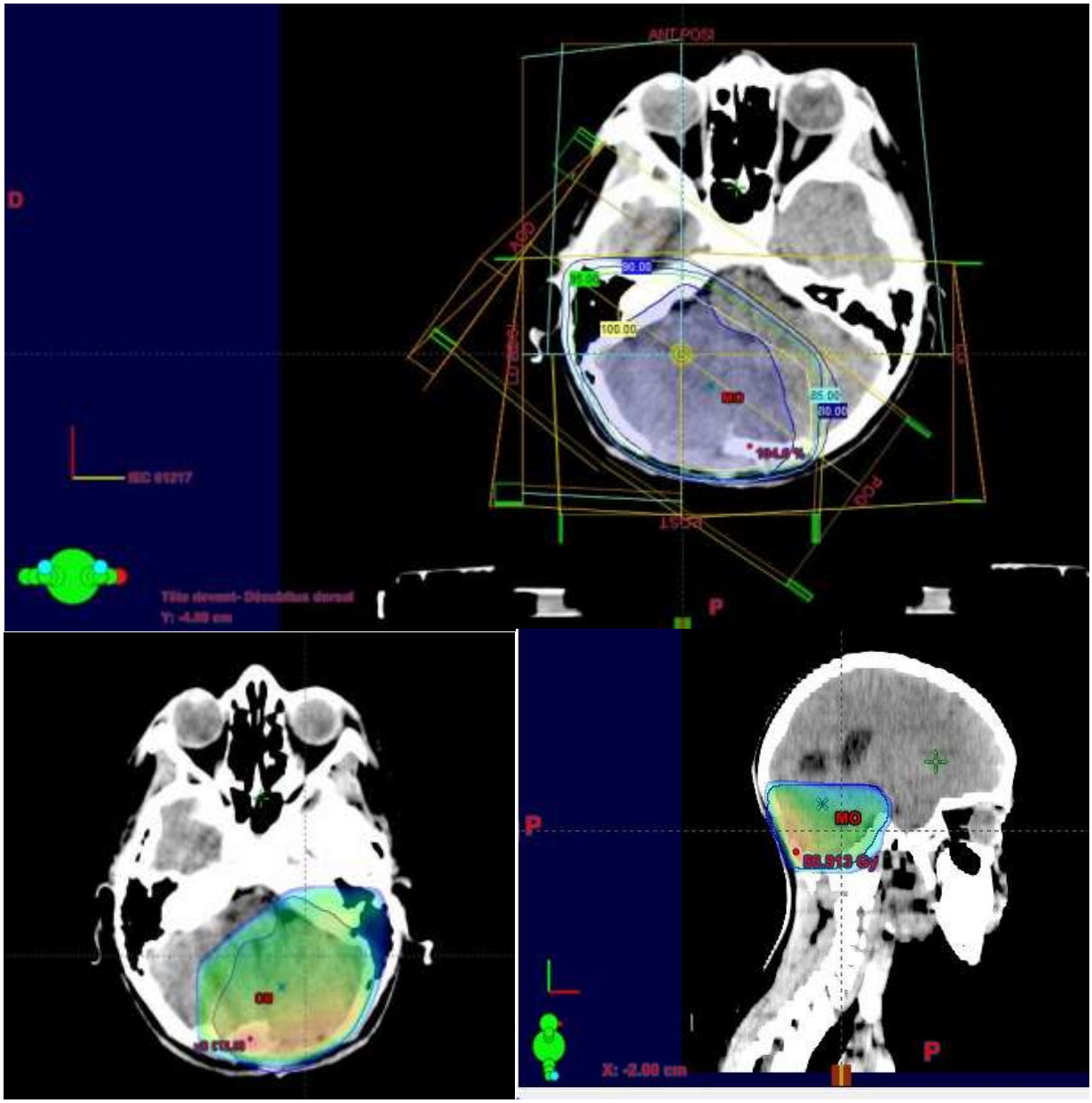


Figure 2.3Postoperative craniospinalMRI: no **definite residual tumoridentified. Spinal MRI findings were normal, showing no evidence of metastatic spread or other abnormalities**.

The patient's case was discussed during a multidisciplinary neuro-oncology tumor board, where the decision was made to proceed with adjuvant postoperative radiotherapy.

A high-resolution cranial planning CT scan was performed to enable precise treatment planning. Target volumes and organs at risk were delineated following image fusion and co-registration with pre- and postoperative magnetic resonance imaging (MRI) scans. Due to the unavailability of advanced techniques such as volumetric modulated arc therapy (VMAT) and tomotherapy in our department, three-dimensional conformal radiotherapy (3D-CRT) was administered. Treatment was delivered to the operative bed with a total dose of 54 Gy in 30 daily fractions of 1.8 Gy, in accordance with current recommendations for this tumor type.



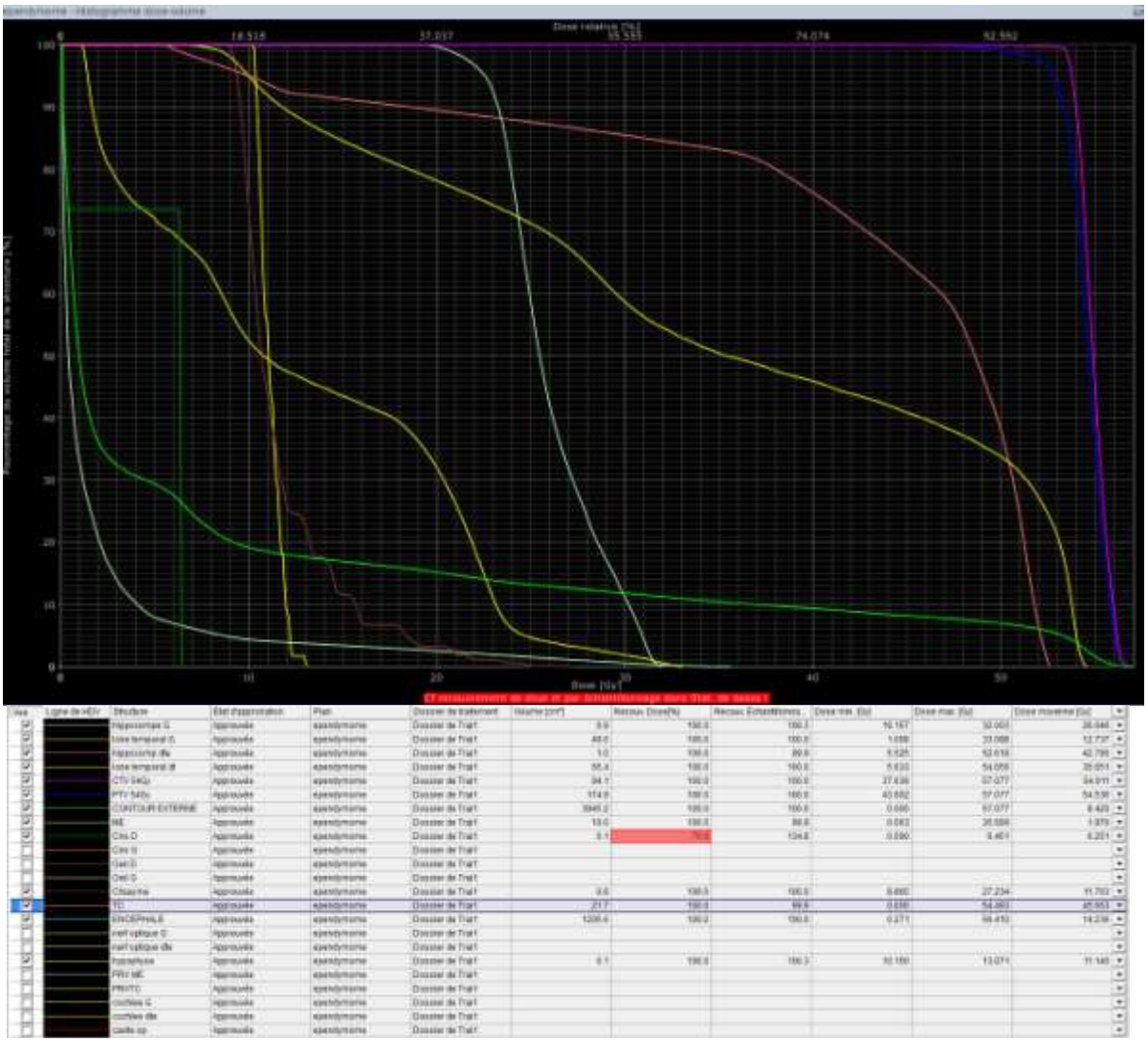


Figure 4-8Dosimetric images of cranial irradiation with a total dose of 54 Gy delivered using three-dimensional conformal radiotherapy (3D-CRT).

The total treatment duration was 46 days, with no interruption exceeding two days.
Adverse effects included two areas of alopecia and grade I radiodermatitis according to CTCAE criteria. No toxicity above grade II was observed.

Post-treatment MRI**,** performed one month after the completion of radiotherapy, demonstrated a complete disappearance of the fourth ventricle lesion previously described on imaging.
At six months, brain MRI demonstrated a slight regression of postoperative and post-radiotherapy changes in the posterior fossa, with no visible residual tumor. There was also a regression of the previously noted mild triventricular dilatation. Bilateral thickening of the paranasal sinus walls was observed, suggestive of chronic inflammatory changes.

After three years of follow-up, clinical examination showed that the patient was fully conscious, with no neurological deficits and normal psychomotor development.Themost recent MRI revealed a further reduction of post-therapeutic changes around the fourth ventricle and occipital paramedian bones, with no evidence of tumor recurrence**.**

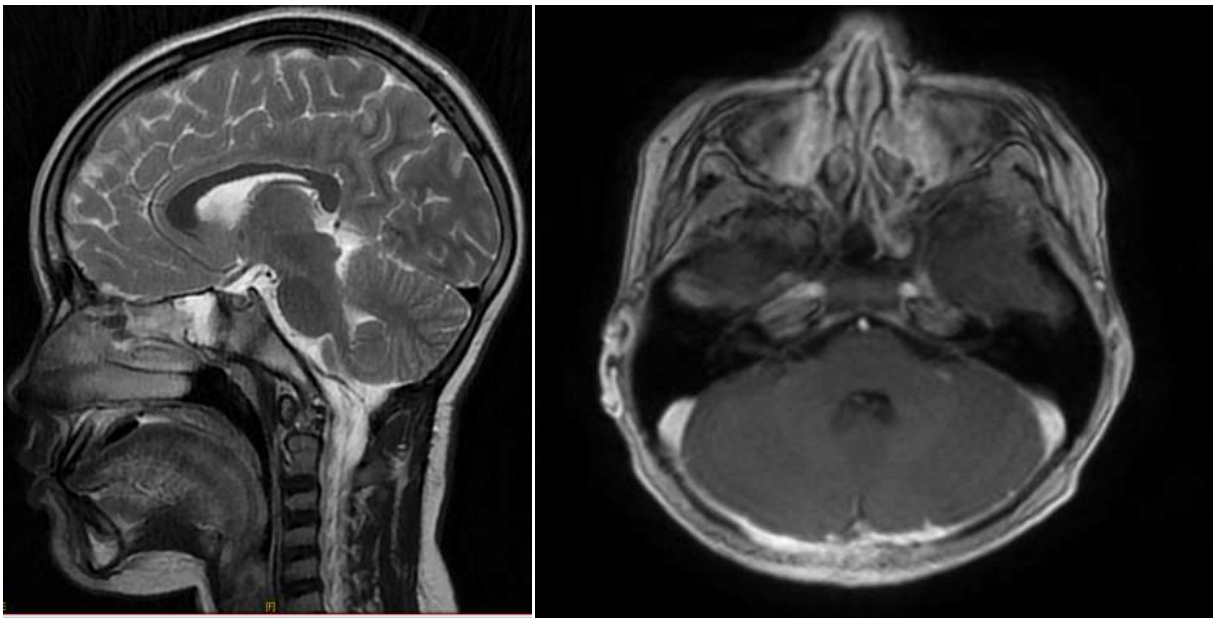


Figure 9 :**MRIrevealed a further reduction of post-therapeutic changes around the fourth ventricle and occipital paramedian bones, with no evidence of tumor recurrence**.

Post-therapeutic follow-up for this patient is based on clinical and radiological evaluations every six months during the first five years, followed by annual assessments thereafter. The primary goal is to maximize long-term remission while allowing prompt intervention in the event of recurrence. This follow-up also facilitates the early detection of late treatment-related complications, monitoring of the patient's overall clinical course, and assessment of functional and neurological prognosis, thereby enabling tailored adjustments to patient care as needed.

Discussion:-

Pediatric posterior fossa ependymomas, particularly those located within or adjacent to the fourth ventricle, present significant clinical challenges. Their anatomical location complicates surgical management, and the frequent occurrence of obstructive hydrocephalus often leads to headaches, vomiting, and cerebellar signs such as gait disturbances and ataxia. Early diagnosis is critical to prevent neurological deterioration [5,6].

Epidemiology and Clinical Presentation:

Pediatric ependymomas are uncommon glial tumors, most frequently arising in the posterior fossa, particularly within the fourth ventricle (1-2). The anaplastic subtype (WHO grade III) is more aggressive than grade II tumors and carries a higher risk of local recurrence and reduced overall survival (5; 17). Symptoms usually result from obstructive hydrocephalus or mass effect on the cerebellum and brainstem, including headache, nausea, vomiting, gait instability, and balance difficulties (18). Prompt recognition of these clinical features is crucial to prevent neurological deterioration and enable timely intervention (12).

Imaging and Diagnosis:

Magnetic resonance imaging (MRI) is essential for initial diagnosis, surgical planning, and postoperative monitoring. Typical MRI features include iso- or hypointensity on T1-weighted sequences, hyperintensity on T2-weighted sequences, and heterogeneous enhancement after gadolinium administration. MRI also provides crucial information on hydrocephalus, brainstem or cerebellar involvement, and potential spinal dissemination ("drop metastases") [7].

Surgical Resection:

Surgery remains the cornerstone of treatment. Gross total resection (GTR) is strongly associated with improved progression-free survival (PFS) and overall survival (OS). Subtotal resection consistently correlates with higher recurrence rates and poorer outcomes [5,7]. Achieving GTR is often difficult when tumors involve the floor of the fourth ventricle, lateral walls, or adjacent cisternal spaces, highlighting the importance of specialized pediatric neurosurgical expertise [7,8].

Adjuvant Radiotherapy:
Postoperative radiotherapy is recommended for WHO grade II and III ependymomas, even after GTR, to minimize local recurrence. Combining GTR with modern conformal radiotherapy (3D-CRT or IMRT) improves local control and survival while sparing normal tissue, thereby reducing acute and late toxicity [9]. For example, Merchant et al. reported a 10-year local control rate of 100% for patients treated with GTR plus radiotherapy, compared to 50% for GTR alone [9].

Histology, Molecular Classification, and Prognostic Factors:
While histological grade provides some prognostic value, extent of resection and molecular subgroup are more predictive of outcomes [10]. Posterior fossa (PF) ependymomas are classified into PF-A and PF-B subgroups. PF-A tumors are more aggressive, occur in younger children, and have worse prognosis, whereas PF-B tumors generally have better outcomes [11,12]. Other prognostic factors include age at diagnosis, tumor volume, severity of hydrocephalus, and presence of spinal or CSF dissemination [11,13].

Clinical Implications:-
Our case illustrates these principles. A six-year-old child with a fourth ventricle ependymoma underwent ventriculo-cisternostomy followed by GTR and adjuvant radiotherapy (54 Gy), achieving intact neurological function and no recurrence at three years. This underscores the favorable impact of combined GTR and radiotherapy and highlights the importance of multidisciplinary management involving neurosurgery, radiation oncology, imaging, and pathology.

Future Directions:
Routine molecular profiling should guide risk-adapted therapy. PF-A tumors may benefit from early inclusion of novel therapies, while PF-B tumors could be candidates for dose-modified radiotherapy or de-escalation strategies. Prospective multicenter registries and trials are needed to refine protocols, especially for infants and very young children who cannot tolerate immediate radiotherapy [5,6]

Conclusion:-
Fourth ventricle ependymoma remains a rare pediatric tumor that requires prompt and multidisciplinary management [14 ;15]. Complete surgical resection combined with adjuvant radiotherapy represents the standard of care and is a key determinant of prognosis [15,16]. Our case highlights the importance of early diagnosis and regular follow-up to optimize survival outcomes and maintain quality of life in affected children [14,15].

Conflict of Interest:
None declared.

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