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A Rare Case of WHO Grade 3 Posterior Fossa Ependymoma in an Adult: A Case Report

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Abstract: Ependymomas are rare tumors of the central nervous system (CNS), accounting for about 2-6% of all intracranial neoplasms. High-grade posterior fossa ependymomas, particularly WHO Grade 3 anaplastic ependymomas, present significant clinical and radiological challenges due to their aggressive behavior and recurrence potential. This report describes a case of a 60-year-old female with a posterior fossa mass causing obstructive hydrocephalus, ultimately diagnosed as WHO Grade 3 posterior fossa ependymoma. This case emphasizes key imaging characteristics, differential diagnoses, and the clinical implications of high-grade ependymomas.

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I. INTRODUCTION

Ependymomas originate from ependymal cells lining the ventricular system and spinal canal. While these tumors are more common in pediatric populations, posterior fossa ependymomas also occur in adults and frequently lead to hydrocephalus by obstructing cerebrospinal fluid (CSF) pathways [1]. The 2021 WHO classification defines anaplastic ependymomas as WHO Grade 3 tumors, characterized by increased mitotic activity, necrosis, and microvascular proliferation [2]. MRI remains the preferred imaging modality for evaluating these lesions, typically revealing a mix of cystic, hemorrhagic, and enhancing areas [3]. This report highlights the imaging features, differential considerations, and treatment strategies for high-grade posterior fossa ependymomas.

II. CASE PRESENTATION

A 60-year-old female presented with a progressive loss of balance while walking for six months, followed by symptoms of headache, giddiness, nausea, vomiting, and a single episode of loss of consciousness over two days. She had a history of diabetes but was not hypertensive. Clinical examination showed decreased bilateral plantar reflexes, while routine blood investigations were normal.

III. IMAGING FINDINGS

MRI of the brain with contrast revealed a well-defined, lobulated mass in the inferior fourth ventricle near the obex, measuring 2.9 × 3.8 × 3.8 cm. The lesion displayed a T1 isointense to hyperintense and T2 heterogeneous signal pattern with multiple T2 hyperintensities that suppressed on FLAIR, indicating cystic changes.

- Anteriorly, the lesion exhibited an indistinct interface with the floor of the fourth ventricle (pons and medulla), causing mass effect and splaying of the medulla.
- Anterolaterally, extension into the bilateral foramina of Luschka (left > right) was noted.
- Posteriorly, the lesion caused mass effect on the cerebellar vermis.
- Inferiorly, extension into the cisterna magna via the foramen of Magendie was seen.
- Superiorly, the lesion resulted in ventricular dilatation (fourth, third, and lateral ventricles) and periventricular CSF seepage, suggesting obstructive hydrocephalus.
- The lesion showed SWI blooming, indicating hemorrhage or calcification, and demonstrated heterogeneous contrast enhancement on post-contrast T1-weighted images. There was no evidence of peritumoral edema or diffusion restriction on DWI.

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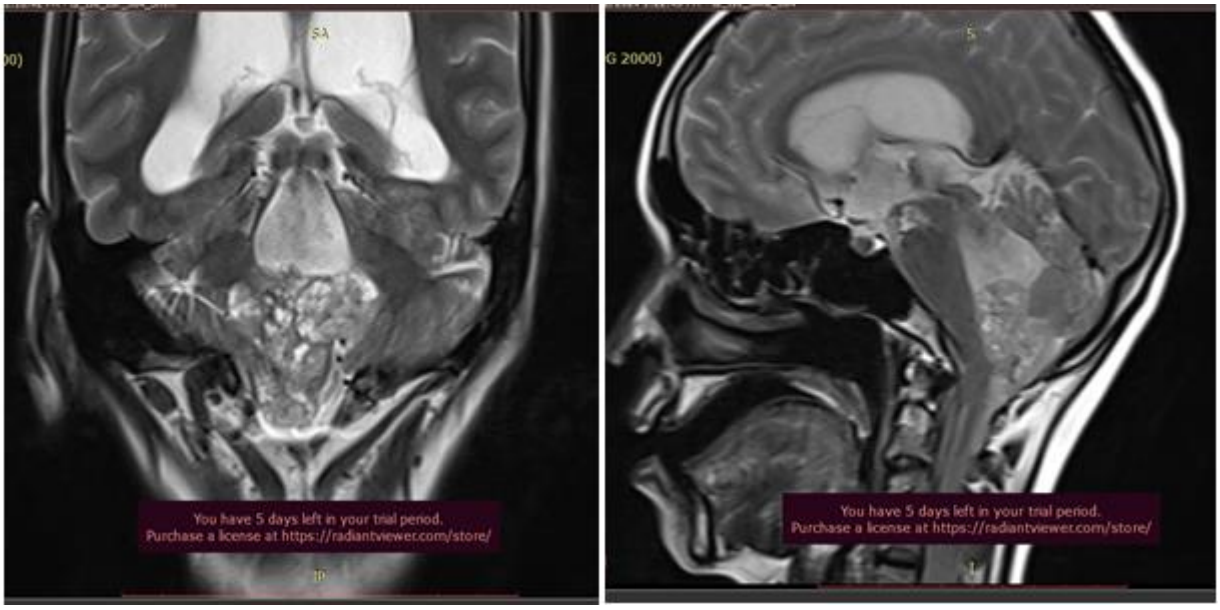
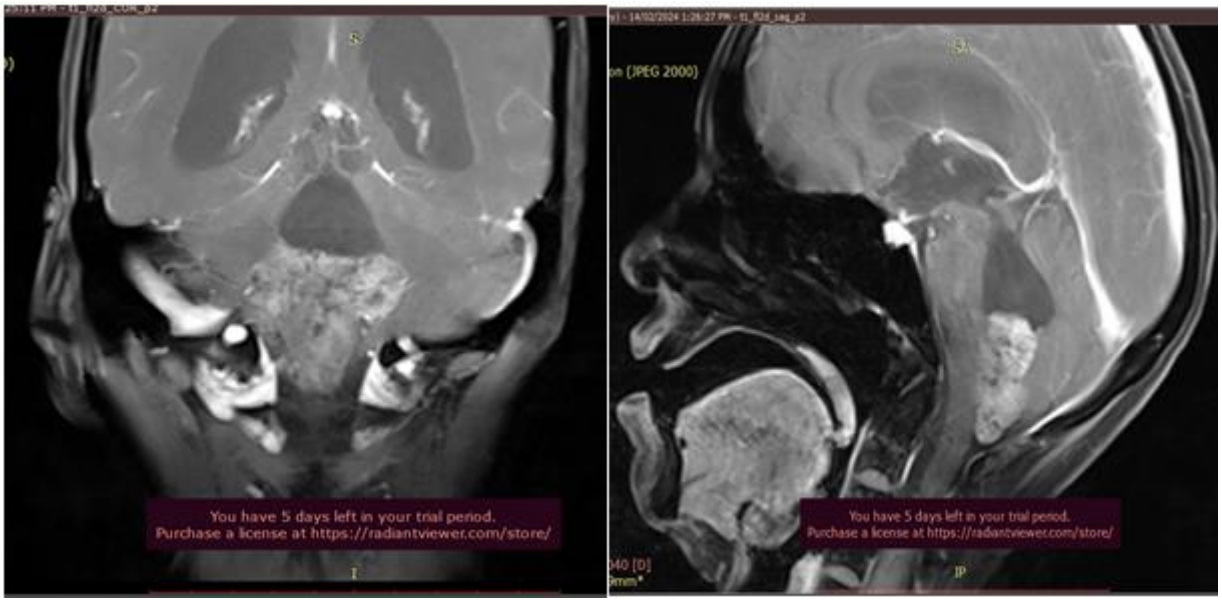
IV. T2 WEIGHTED IMAGES: CORONAL AND SAGI



T2 WEIGHTED IMAGES: CORONAL AND SAGITTAL



T1 POST CONTRAST IMAGES: CORONAL AND SAGITTAL

V. DIFFERENTIAL DIAGNOSIS

Based on the imaging findings, the following differentials were considered:

- Posterior fossa ependymoma (Type B) – Typically characterized by a heterogeneous T2 signal, cystic degeneration, and extension through the foramina of Luschka and Magendie [4].
- Subependymoma – Usually well-circumscribed and non-enhancing, with a lower grade and minimal associated mass effect or hemorrhage [5].
- Choroid plexus papilloma – Frequently lobulated, with strong contrast enhancement and less infiltration into adjacent structures [6].

VI. HISTOPATHOLOGY AND FOLLOW-UP

The patient was referred to a tertiary neuro-oncology center (NIMHANS), where a biopsy confirmed WHO Grade 3 posterior fossa ependymoma. The treatment plan included surgical resection followed by adjuvant radiation therapy.

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VII. DISCUSSION

Posterior fossa ependymomas are a notable subset of adult posterior fossa tumors. WHO Grade 3 anaplastic ependymomas tend to display aggressive biological behavior and have a high recurrence risk even after surgical excision [7].

MRI findings play a crucial role in diagnosis and surgical planning. These tumors often exhibit a heterogeneous signal intensity on T2-weighted imaging, with cystic, necrotic, and hemorrhagic areas. The “cap sign” on T2-weighted sequences, indicative of hemosiderin deposition, is a distinguishing feature of ependymomas [8]. Additionally, due to the risk of CSF dissemination, spinal MRI is recommended for staging high-grade cases [9]. The preferred treatment approach involves gross total resection (GTR), when feasible, to reduce recurrence risk. Adjuvant radiation therapy has been shown to improve progression-free survival significantly [10]. Chemotherapy is generally less effective but may be considered in recurrent or metastatic cases [11]. Despite aggressive treatment, the long-term prognosis remains cautious, with 5-year survival rates ranging between 50-70%, depending on resection extent and response to radiation [12].

VIII. CONCLUSION

High-grade posterior fossa ependymomas pose significant diagnostic and management challenges. MRI is crucial for recognizing tumor characteristics, differentiating from similar lesions, and aiding surgical planning. Early detection and comprehensive management, including surgical excision and adjuvant therapy, are key to improving patient outcomes.

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