# Concurrent Medulloblastoma and Ependymoma in a Single Patient: A Rare Illustrative Case

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## ABSTRACT

Simultaneous detection of two different primary brain tumours such as medulloblastoma and ependymoma, which develop from different types of cells of the central nervous system, in a single patient is a highly rare occurrence. In this article, we present a case in which these two primary brain tumours were found together and treated subsequently. A 22-year male patient was evaluated with symptoms of increased intracranial pressure. Investigations revealed two mass lesions, one in the cerebellum and other in the 3rd ventricle. Surgical excision of the tumours was performed along with a third ventriculostomy. Histopathological analysis confirmed the mass in the cerebellum as medulloblastoma and the mass in the third ventricle as ependymoma. Although the mass in the cerebellum recurred during follow-up, the patient did not show signs of hydrocephalus, probably due to the third ventriculostomy. The simultaneous occurrence of different primary brain tumours poses a significant challenge regarding diagnosis, treatment, and prognosis. Such complex cases require versatile and multidisciplinary approaches. This unique case highlights the importance of a comprehensive understanding of tumour development and consideration of atypical tumour presentations.

**Key Words:** *Ependymoma, Medulloblastoma, Concurrent tumour, Third ventriculostomy, Primary brain tumour.*

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## INTRODUCTION

Primary brain tumours (PBTs) consist of a wide variety of neoplasms that can arise from various cell types within the central nervous system. Medulloblastoma and ependymoma are subtypes of PBTs. Medulloblastoma, found mostly in the posterior fossa, is a malignant tumour of neuroectodermal origin that usually affects children, whereas ependymoma develops from ependymal cells lining the ventricles of the brain and the central canal of the spinal cord.[1]

The simultaneous occurrence of two different PBTs in one patient is extremely rare.[2] In addition, most of these tumours develop following radiation therapy or in association with inherited cancer conditions, and there have been a few reported cases where they have occurred simultaneously.[3] Based on a comprehensive review of current researches, there are no reported cases of individuals diagnosed with both medulloblastoma and ependymoma. The presence of these tumours in the same patient can complicate diagnosis, treatment planning, and prognosis, and requires a comprehensive evaluation and collaborative management strategy.

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This case report describes a patient with two intracranial tumours, medulloblastoma and ependymoma, and examines the unusualness of this case and its aetiology, pathogenesis, and management.

## CASE REPORT

A 22-year male patient was admitted to the emergency department with a persistent headache accompanying vomiting that lasted for 20 days and intensified in the last five days. In his medical history, he was particularly lacking in previous conditions. No abnormality was detected in the physical examination, but the presence of papilloedema was observed. Subsequent imaging investigations revealed a mass in the left cerebellar region and third ventricle, and a visible enlargement of the ventricles (Figure 1).

First, near-total excision of the mass in the posterior fossa was performed. Then, mass samples were taken from the right Kocher point with an endoscopic procedure and the tumour was excised endoscopically by aspirating the tumour from the third ventricle. After the procedure, the patient underwent a ventriculostomy. External ventricular drainage was placed into the ventricle as a precaution. The ventricular drainage was removed because the patient did not develop hydrocephalus in the postoperative period. Daily neurological examinations revealed no unusual findings and the patient was discharged in the first postoperative week. Histopathological examination of tumour sample from the posterior fossa showed tumour composed of small round cells with an increased nucleus/cyto-

plasm ratio (Figure 2A). Tumour cells were positive for synaptophysin. In the sample of the third ventricular tumour, a uniform tumour with prominent perivascular pseudorosette structures was observed. Tumour cells were stained in a dot-like pattern with Epithelial Membrane Antigen (EMA) (Figure 2B). The pathology of the tumour in the posterior fossa was medulloblastoma and that of the mass in the third ventricle was ependymoma.

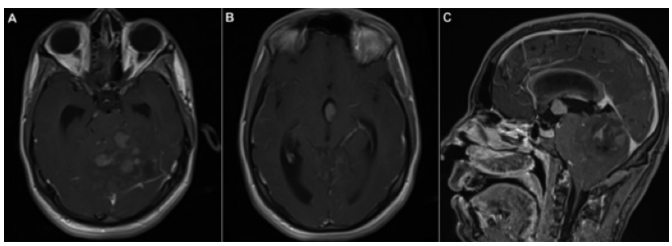


**Figure 1: Contrast-enhanced T1-weighted Magnetic Resonance Imaging (MRI) images preoperatively. (A) Axial, cerebellar tumour; (B) Axial, third ventricular tumor; (C) Sagittal, both tumours can be seen in the same image.**

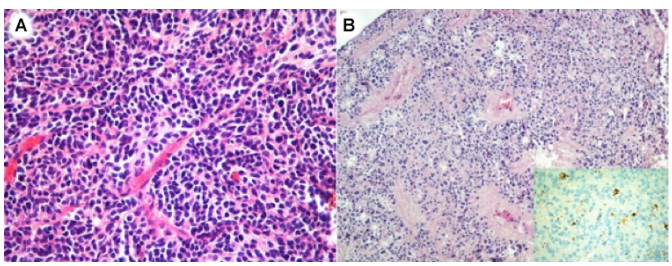


**Figure 2: (A) Medulloblastoma: Tumour consisting of cells with a small size and increased nucleus/cytoplasmic ratio (H&E, x400); (B) Ependymoma: True and pseudo-rosette structures in the tumour (H&E, x100) and dot-like staining pattern by EMA (DAP, x400) immunohistochemistry (inset).**

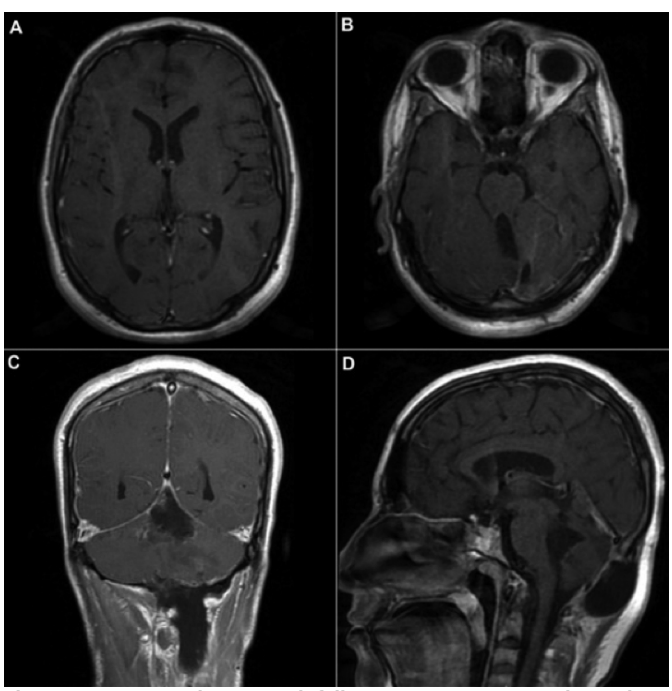


**Figure 3: Postoperative 9-month follow-up MRI, contrast-enhanced T1-weighted MRI images; (A) Axial, level of the third ventricle; (B) Axial, level of the cerebellum; (C) Coronal; (D) Sagittal.**

On follow-up examination at five months after the surgical intervention, it was noted that despite being referred to the departments of medical oncology and radiation oncology, the patient did not engage with either service owing to non-compliance. Thus, a recurrence of the tumour in the posterior fossa was observed, but hydrocephalus was not found. The patient underwent surgery again. After the surgical procedure, the patient was directed to the medical oncology and radiation oncology units for further treatment. No recurrence was observed in the 9-month follow-up and no shunt was required (Figure 3).

## DISCUSSION

PBTs are a heterogeneous group of tumours that can arise from different cells in the central nervous system. Multiple metastatic brain tumours or multiple primary brain tumours with the same histological origin are frequently observed in both adult and paediatric populations. Nevertheless, the simultaneous occurrence of multiple PBTs with distinct cellular origins is an uncommon event in clinical practice.[4]

Advancements in molecular pathology have refined our understanding of medulloblastomas, highlighting the roles of WNT and Sonic Hedgehog (SHH) pathways in their genesis. WNT activation leads to intracellular accumulation of beta-catenin, which subsequently triggers transcriptional activity. Conversely, SHH signalling promotes the proliferation of cerebellar precursor cells. Ordinarily, the Phenylthiocarbamide (PTC) receptor inhibits the Smoothened (SMO) receptor, but SHH binding negates this inhibition. This results in Gli1 accumulation, which induces cellular proliferation, angiogenesis, and the suppression of apoptosis.[5]

In the formation of ependymomas, many molecular pathological changes have been observed. These changes are characterised by the location of the ependymoma. Therefore, ependymomas are now classified into 3 main anatomical groups: supratentorial, posterior fossa, and spinal.[6]

The discovery of a second tumour may occur before, during, or even after surgery for the first intracranial tumour, and sometimes months or years may pass after successful treatment of the first tumour before the second tumour is detected. In some cases, the presence of the second tumour is only revealed during a post-mortem examination. There have even been cases where a patient has been found to have more than two PBTs.[7]

In the late 19$^{th}$ century, Julius Cohnheim proposed that tumours originate from embryonic remnants capable of uncontrolled proliferation. While this theory could offer some insight into a concurrent case of medulloblastoma and ependymoma, it falls short in explaining the sporadic nature of cancer development from such cells. Moreover, the different cellular origins and locations of the tumours in the subject case, along with the absence of predisposing factors, suggest that their simultaneous occurrence may be coincidental. However, this case highlights the complexity of tumour development and the different factors that can potentially contribute to this process.

In their study, Butti *et al*. discovered that 45% of multiple primary intracranial tumours are found in neighbouring areas, suggesting that this may imply a causal link. They hypothesised that one tumour could potentially act as a stimulus, promoting the growth of another nearby tumour. In the present case, due to the non-contiguous location of the tumours, their simultaneous occurrence was probably coincidental.[8]

Radiation exposure may be an important factor in the development of multiple PBTs. It has been observed that individuals who undergo cranial radiotherapy for a first brain tumour have an increased risk of subsequently developing a secondary neoplasm.[9] It has also been suggested that trauma is a potential contributing factor of tumour formation.[7]

However, this patient had no symptoms or family history of phacomatosis, trauma, or radiation exposure.

The widespread use of neuroimaging techniques may be perceived as artificially inflating the incidence of multiple brain tumours due to better detection of these conditions.

Another point worth mentioning is that, in the literature, studies showed that third ventriculostomy in addition to tumour resection reduces postoperative shunt dependency in the treatment of hydrocephalus due to posterior fossa tumours. Additionally, carrying out an endoscopic third ventriculostomy before tumour removal presents benefits for patients' post-surgical recovery. It reduces the complexity of their postoperative course by eliminating the risk of cerebrospinal fluid infection related to external ventricular drainage and decreasing the likelihood of over-drainage complications.[10] In our clinical practice, intraoperative third ventriculostomy for intraventricular neoplasms is not a standard procedure. However, given the high risk of recurrence for the cerebellar mass in this patient, a third ventriculostomy was performed in the same session after endoscopic excision of the neoplasm located within the third ventricle. Accordingly, even when the mass in the cerebellum of the patient recurred, the patient had no signs of hydrocephalus. Therefore, we believe that in appropriate cases, experienced neurosurgeons should consider performing a third ventriculostomy if the third ventricle has already been entered.

## CONCLUSION

Neurosurgeons need to be aware of rare cases of multiple PBTs with different histological features that could be present in the same patient and pose diagnostic and therapeutic challenges.

### PATIENT'S CONSENT:

An explicit consent of the patient was obtained to publish the case.

### COMPETING INTEREST:

All authors involved in this article have declared no conflict of interest. Additionally, it is important to note that this research project has been conducted without any external funding or financial support.

### AUTHORS' CONTRIBUTION:

CT: Carried out and prepared the literature review.
UTS: Prepared the figures.
SE: Provided pathological staining and diagnosis.
MC: Shaped the final version of the article.
All authors approved final version of the manuscript to be published.

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