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CASE REPORT

Temporal Ependymoma: A Case Study Emphasising Diagnostic Dilemmas and Therapeutic Challenges

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ABSTRAK

Ependimoma temporal merupakan neoplasma intrakranial yang sangat jarang berlaku, yang bermula dari sel ependimal dalam lobus temporal. Disebabkan tumor ini jarang dijumpai maka diagnosis dan rawatannya amat sukar dalam bidang neuro-onkologi. Tumor ini sering mengalami diagnosis yang salah sebagai meningioma atau lesi tambahan paksi yang lain disebabkan oleh lokasi atipikal dan ketiadaan ciri tersendiri. Oleh itu, pemeriksaan histopatologi adalah penting untuk penilaian dan diagnosis yang tepat. Kajian ini membentangkan kes unik pesakit lelaki yang berusia 25 tahun yang mengalami ependimoma supratentorial di lobus temporal. Laporan ini menerangkan penyampaian klinikal pesakit termasuk sakit kepala yang bersifat progresif, tingkah laku yang tidak normal dan muntah, yang membawa kepada penyiasatan diagnostik yang tepat pada masanya. Selepas pembedahan, diagnosis ependimoma disahkan oleh analisis histopatologi. Pesakit gagal membuat rawatan susulan selepas pembedahan dan seterusnya menunjukkan gejala yang semakin teruk selepas tempoh enam bulan. Pengimejan radiologi membuktikan pertumbuhan ketara tumor. Pembedahan kedua mengakibatkan defisit neurologi yang lebih teruk. Malangnya, radioterapi adjuvan tidak lagi memberi manfaat kepada beliau ketika ini. Pesakit menerima rawatan paliatif. Kajian kes ini menekankan kesukaran dalam mendiagnosis dan merawat penyakit ependimoma temporal, menitikberatkan kepentingan kerjasama pelbagai disiplin dan pendekatan berasaskan bukti untuk hasil pesakit yang optimum.

Kata kunci: Ependimoma; lobus temporal; neoplasma otak; paksi tambahan; supratentorial

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ABSTRACT

Temporal ependymomas are exceedingly rare intracranial neoplasms originating from ependymal cells within the temporal lobe. The rare nature of these tumours makes diagnosis and treatment extremely difficult in the field of neuro-oncology. These tumours frequently encounter misdiagnosis as meningiomas or other extra-axial lesions due to their atypical location and the absence of distinctive features. Consequently, histopathological examination becomes essential for accurate evaluation and a precise diagnosis. This article presented a unique case of a 25-year-old male patient's supratentorial ependymoma at the temporal region. The report described the patient's clinical presentation, including progressive headaches, abnormal behaviour and vomiting, leading to timely diagnostic investigations. After a surgical resection, the diagnosis of ependymoma was confirmed by histopathological analysis. Following surgery, the patient defaulted follow-up and presented six months later with worsening symptoms. Radiological imaging proved the tumour's significant growth. The second resection worsened his neurological deficits. Unfortunately, adjuvant radiotherapy would not be beneficial to him at this point. He received palliative care. The case study emphasises the difficulty in diagnosing and treating temporal ependymomas, underscoring the importance of multidisciplinary collaboration and evidence-based approaches for optimal patient outcomes.

Keywords: Brain neoplasms, ependymoma; extra-axial; supratentorial; temporal lobe

INTRODUCTION

Ependymomas are rare primary tumours arising from the ependymal lining of the central nervous system that present unique challenges in neuro-oncology due to their varied clinical behaviour and histological heterogeneity. Globally, ependymomas account for 2 to 9% of all primary brain and central nervous system tumours. It is well known that the sites of ependymoma tumour typically vary in children and adults, with the majority of cases occurring in the spinal cord in adults and the posterior fossa in children (Elsamadicy et al. 2020). From a radiological standpoint, supratentorial extra-axial, more precisely temporal ependymomas, are often misdiagnosed as meningiomas or other extra-axial lesions due to their atypical location and the absence of distinctive features. Consequently, histopathological examination becomes essential for accurate evaluation and a precise diagnosis (Lombardi et al. 2021). The scarcity of these cases underscores the importance of documenting and studying individual experiences to enhance our understanding of this rare disease.

This case study reported a rare supratentorial extra-axial temporal ependymoma, shedding light on the intricate clinical course, diagnostic evaluation and therapeutic management. The report centered on a specific patient's experience, detailing the initial clinical presentation, radiological findings, and pathological characterisation of the temporal ependymoma. The case offered

a valuable opportunity to analyse the complexities involved in diagnosing this rare tumour, as well as the subsequent decision-making process in developing an evidence-based treatment plan tailored to the patient's unique condition.

Through a meticulous examination of this ependymoma case report, our research aspired to bridge the gap between clinical experience and scientific inquiry, ultimately guiding future research directions and optimising patient care for those diagnosed with this challenging tumour. By pooling knowledge and expertise, we strive to further unravel the complexities of ependymomas, fostering advancements in diagnosis, treatment, and ultimately, improved patient outcomes.

CASE REPORT

A 25-year-old male complained of generalised headache, abnormal behaviour and episodes of vomiting. Neurological examination revealed no significant findings. A plain computed tomography (CT) scan of the brain revealed a left temporal tumour with clear mass effect (Figure 1). On 15th August 2022, the patient underwent a left craniectomy coupled with tumour debulking, all within 24 hours of the first presentation. The ensuing histopathological analysis (Figure 2), indicated characteristics that were consistent with a high-grade tumour with neuroglial differentiation and a polymorphous immunophenotype, but the final report was not available. Post-

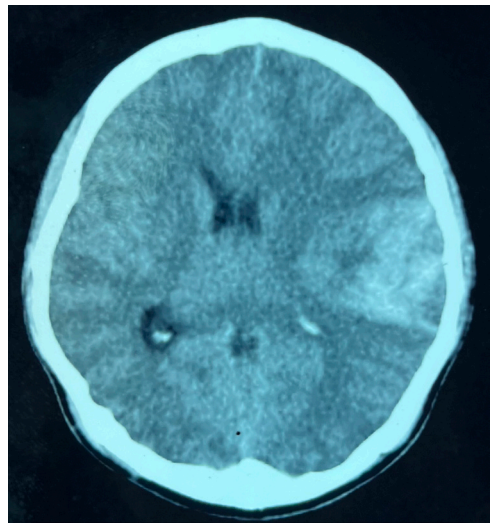

FIGURE 1: Plain computed tomography (CT) brain

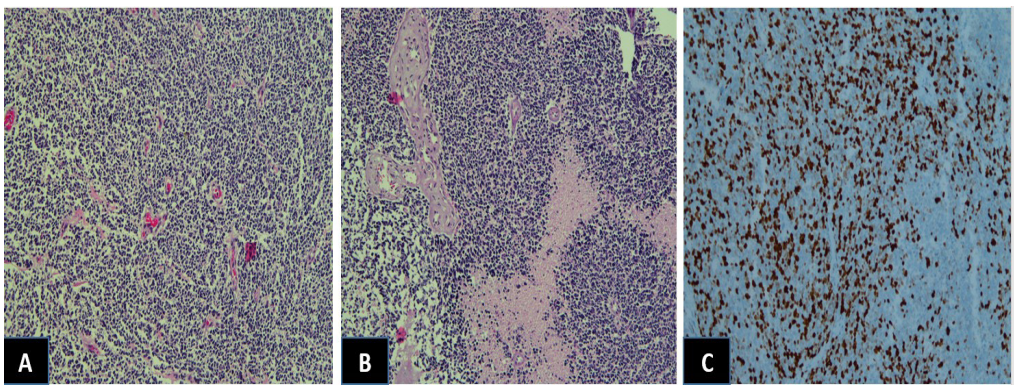

FIGURE 2: (A) Hypercellular tumour (x20); (B) Tumour necrosis and microvascular proliferation (x20); (C) Immunohistochemistry Ki-67

operatively, the patient experienced no neurological deficits and was discharged after two weeks. Following the initial surgical operation, the patient failed to return for follow-up. When he returned in June 2023, he had progressive right-sided body weakness and expressive dysphasia. During this visit, a neurological examination revealed that he had 3/5 and 4/5 power over his right upper and lower limbs, respectively. A follow-up contrast-enhanced CT scan (Figure 3) and magnetic resonance imaging (MRI) (Figure 4) revealed that the tumour had recurred. A secondary intervention was initiated, which included an emergency left re-craniotomy and tumour debulking. The histopathological report denoted a poorly differentiated high-grade brain tumour. The histopathological samples were sent to a neuropathologist for further analysis. Post-operative MRI brain showed residual tumour with encephalomalacic changes, no evidence of drop metastases (Figure 5). After convening a multidisciplinary meeting including neurosurgeons, radiologists, pathologists and oncologists, consensus was reached to classify and treat the tumour as a supratentorial ependymoma. Finally, a grade III supratentorial ependymoma was diagnosed, in accordance with the World Health Organisation classification.

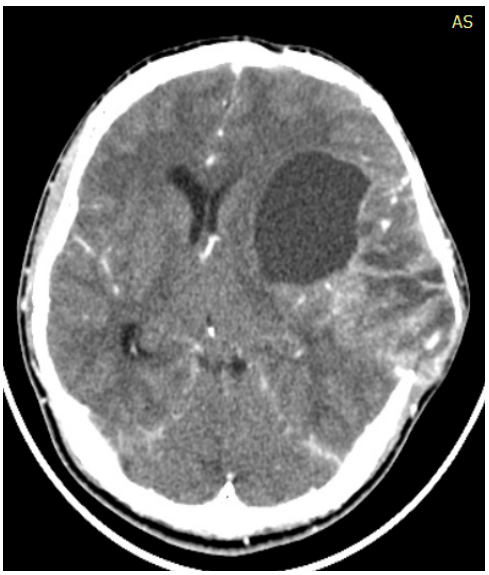

FIGURE 3: Contrast-enhanced computed tomography (CECT) brain

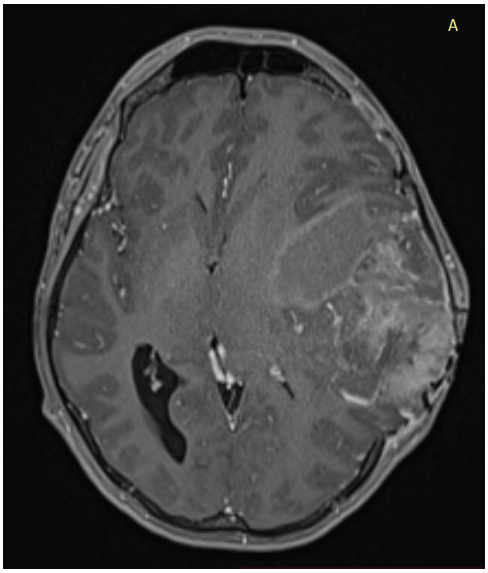

FIGURE 4: Magnetic resonance imaging (MRI) brain

DISCUSSION

Ependymomas are rare primary tumours arising from the ependymal lining of the central nervous system with an incidence rate of 0.42 patients per 100,000 population in the United States. They make up 1.6% of all primary brain tumours (Ostrom et al. 2020). The diagnostic journey for ependymomas is often complex, owing to their rarity and variety of clinical manifestations. In this particular case, the

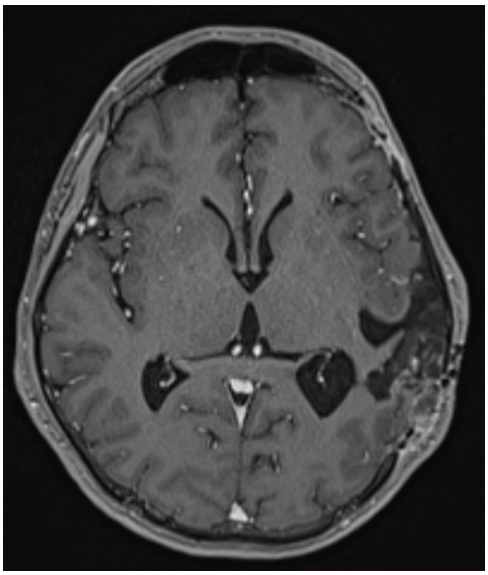

FIGURE 5: Post-operative magnetic resonance imaging (MRI) brain after the patient's second surgery

initial clinical symptoms of progressive headaches, abnormal behaviour and vomiting prompted thorough diagnostic investigations. A plain CT scan of the brain showed an initial diagnosis of temporal meningioma based on its location and radiological features. Similar issues of misdiagnosing meningiomas based on radiological features were encountered in many other instances that have been previously reported of (Bohara et al. 2023; Nagayasu et al. 2022; Osman et al. 2023; Uttam et al. 2022). There are radiological similarities between extra-axial ependymomas and meningiomas, such as the dural tail, subarachnoid plane and diffuse contrast enhancement. This leads to a high rate of misdiagnoses. Despite this, maximum safe resection is advised for both extra-axial ependymomas and meningiomas. The patient underwent craniectomy and tumour debulking surgery within 24 hours upon presentation. Following tumour debulking, a histopathological analysis that revealed characteristics consistent with a high-grade tumour with neuroglial differentiation and a polymorphous immunophenotype.

A post-operative MRI of the brain reported a residual extra-axial lesion measuring 0.9 x 0.7 cm. According to guidelines for evidence-based treatment, adjuvant radiotherapy would have been the best course of action to minimise the risk of recurrence. The European Association of Neuro-Oncology advocated for radiotherapy in adult cases with WHO grade III ependymomas, and in adult cases of incomplete resection of WHO grade II ependymomas (Rudà et al. 2018). However, this patient defaulted follow-up following the first resection and presented after six months with worsening right-sided body weakness and expressive dysphasia. A second tumour resection was then performed. A grade III supratentorial ependymoma was ultimately identified based on the histopathological report, which initially indicated a poorly differentiated high-grade brain tumour. At this stage, the patient was rendered unfit to proceed with adjuvant radiotherapy. Therefore, he was given palliative care.

The rarity of temporal ependymomas makes these cases particularly valuable, as they provide crucial contributions to the sparse literature on this tumour subtype. This case report's findings align with previous studies on the clinical presentation, radiological features and histopathological characteristics of ependymomas. The rarity of temporal ependymomas emphasises the importance of individual case reports, as they contribute crucial contributions to the

limited literature on this tumour subtype.

Limitations and Future Perspectives

This case report, like any individual case study, is subjected to certain limitations. The rarity of temporal ependymomas restricts the generalisability of the findings. Additionally, the absence of genetic profiling and molecular analysis leaves scopes for future research to explore potential diagnostic and prognostic markers. Collaborative efforts through multi-center studies and international databases are essential to overcome these limitations and gather more robust data to advance ependymoma research.

CONCLUSION

In conclusion, this case report provides a valuable addition to the existing knowledge base on ependymomas, particularly those of the supratentorial extra-axial subtypes. By contextualising the case findings within the current literature, this discussion contributes to the ongoing efforts in improving diagnosis, treatment and overall care for patients diagnosed with supratentorial ependymomas.

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