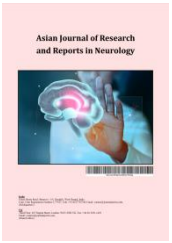

Asian Journal of Research and Reports in Neurology

Volume 7, Issue 1, Page 175-180, 2024; Article no.AJORRIN.126068

Severe Headache as the Sole Presenting Feature in a Rare Papillary Meningioma: A Case Report

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Authors' contributions

This work was carried out in collaboration among all authors. Author RC designed the study, wrote the protocol, and wrote the first draft of the manuscript. Author RKJ was the main neurosurgeon who treated and operated the patient. Author PG managed the literature searches. Author RM helped author PG in writing protocol and drafting of the manuscript. All authors read and approved the final manuscript.

Article Information

Open Peer Review History:
This journal follows the Advanced Open Peer Review policy. Identity of the Reviewers, Editor(s) and additional Reviewers, peer review comments, different versions of the manuscript, comments of the editors, etc are available here: https://www.sdiarticle5.com/review-history/126068

Case Report

Received: 03/09/2024
Accepted: 05/11/2024
Published: 09/11/2024

ABSTRACT

Meningiomas make up around 13-26% of all brain tumours. However, papillary meningioma accounts for only 1 – 2.5% of all meningiomas. Papillary meningiomas are rare and very aggressive in nature. The available literature regarding the ideal management of papillary meningioma is scarce. We present a case of a 30 years old man with complaints of no other symptoms except headache. On investigation, contrast MRI brain showed right parietal extra axial mass suggestive of

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Cite as: *Chhangte, Ramngaihzuala, Ranjeet Kumar Jha, Parul Gupta, and Rebecca Moncy. 2024. "Severe Headache As the Sole Presenting Feature in a Rare Papillary Meningioma: A Case Report". Asian Journal of Research and Reports in Neurology 7 (1):175-80. https://www.journalajorrin.com/index.php/AJORRIN/article/view/115.*

malignant meningioma. Routine blood test were otherwise within the normal range. GCS of the patient was 15/15 pre operatively. Excision of the lesion was done via right parietal craniectomy followed by lax duraplasty. Immediate post operative neurological status was same as the pre operative neurological status. Pathological investigation suggested that the tumour was papillary type of meningioma. Patient was advised to follow up in Radiation oncology department for radiotheraphy.

Keywords: Craniectomy; meningioma; papillary.

1. INTRODUCTION

Although rare, primary intracranial tumours account for 1.4% of newly diagnosed cancer cases and 2.4% of all cancer fatalities [1]. Pilocytic astrocytoma, embryonal tumours, and malignant gliomas are the most prevalent intracranial cancers in children, while meningiomas, pituitary tumours, and malignant gliomas are the most common adult brain tumour types [2].

Menigiomas account for around 13-26% of cerebral tumours. They are typically non-neuroepithelial progenitor arachnoid cap cells-derived benign tumors. They are also more in older age group and among females. Radiotherapy at a dose of 45-60Gy is being used currently in clinical practice although the role of radiotherapy is still unconvincing [3].

According to the Central Brain Tumour Registry Statistical Report 2009-2013, 81.1% of meningiomas were grade I (typical), 16.9% were grade II (atypical), and 1.7% were grade III (anaplastic) [4]. Papillary meningiomas are more aggressive than other forms of meningiomas, but according to WHO grading 2021 the phenotype is insufficient to classify it as a grade III tumour [5,6].

2. PRESENTATION OF CASE

A 30 year old male presented with a history of headache since one month. He had no other complaints other than headache. Headache was severe and intermittent in nature and there was no association with visual disturbance or vomiting. On examination, patient's GCS was 15/15, cranial nerves examination showed no abnormality and there was no sensory or motor weakness. Blood investigations including CBC, LFT and RFT were all within normal range. MRI showed a large ill-defined, T1 isointense, T2 hypointense lesion (Fig. 1) measuring 7x5x4cm in size with broad base towards the dura is noted along the convexity of the right parietal region. On post contrast (Fig. 2), the lesion shows moderate heterogenous enhancement with few non enhancing areas. It is causing buckling of the underlying parietal lobe. T2/FLAIR hyperintense areas are noted in the adjacent cortex suggestive of edema. These features are suggestive of neoplastic etiology likely malignant meningioma.

Patient was planned for excision of the tumor after anesthesia fitness was given. Under general anaesthesia, patient was positioned supine with head slightly tilted towards the opposite side. Right parietal craniectomy was done, and after dura was open, a grayish white mass of irregular surface with focal necrosis was seen. Meticulous resection under neurosurgery microscope was done and Simpson Grade II tumor resection was achieved. Immediate post operative neurological status was same as the pre operative status. The patient discharged after one week and was advised to take radiotherapy.

The histological examination showed a tumor tissue composed of sheets of cells arranged in a predominantly papillary pattern with perivascular pseudo-rosettes (Fig. 3). A pathological diagnosis of Papillary meningioma, was given.

3. DISCUSSION

The single most important risk factor for the development of meningiomas is ionising radiation to the skull. Other familial conditions, such as neurofibromatosis type 2 (NF 2), predispose to the development of meningioma. Other links have been proposed, including a history of head trauma, cigarette smoking, and cell phone use [7]. The clinical features of meningiomas depend on their location-intracranial or spinal dural surface, intraventricular. Clinical symptoms may include headaches due to increased intracranial pressure, focal neurological deficit, cranial nerve involvement, generalized and partial seizures caused by focal mass effect [8].

However papillary meningioma is a type of aggressive meningioma that accounts for 1-2.5% of all meningiomas. When compared to benign meningiomas, they typically exhibit aggressive clinical behaviour, as evidenced by a high rate of brain parenchymal invasion, local recurrence, and extracranial metastases [9] Papillary meningiomas are frequently misdiagnosed as metastatic carcinoma, chemodectoma, ependyoma, choroid plexus papilloma, astroblastoma, and amelontic melanoma. Papillary meningiomas are also more commonly reported in children [10].

Papillary meningiomas can be confirmed by histology. They have pseudo-papillary architecture, pseudo-rosettes, and necrosis on occasion. They are strongly EMA and Vimentin positive. Histopathological grading is a powerful predictive indicator that can be used to alter therapy regimens to the individual patient [11,12].

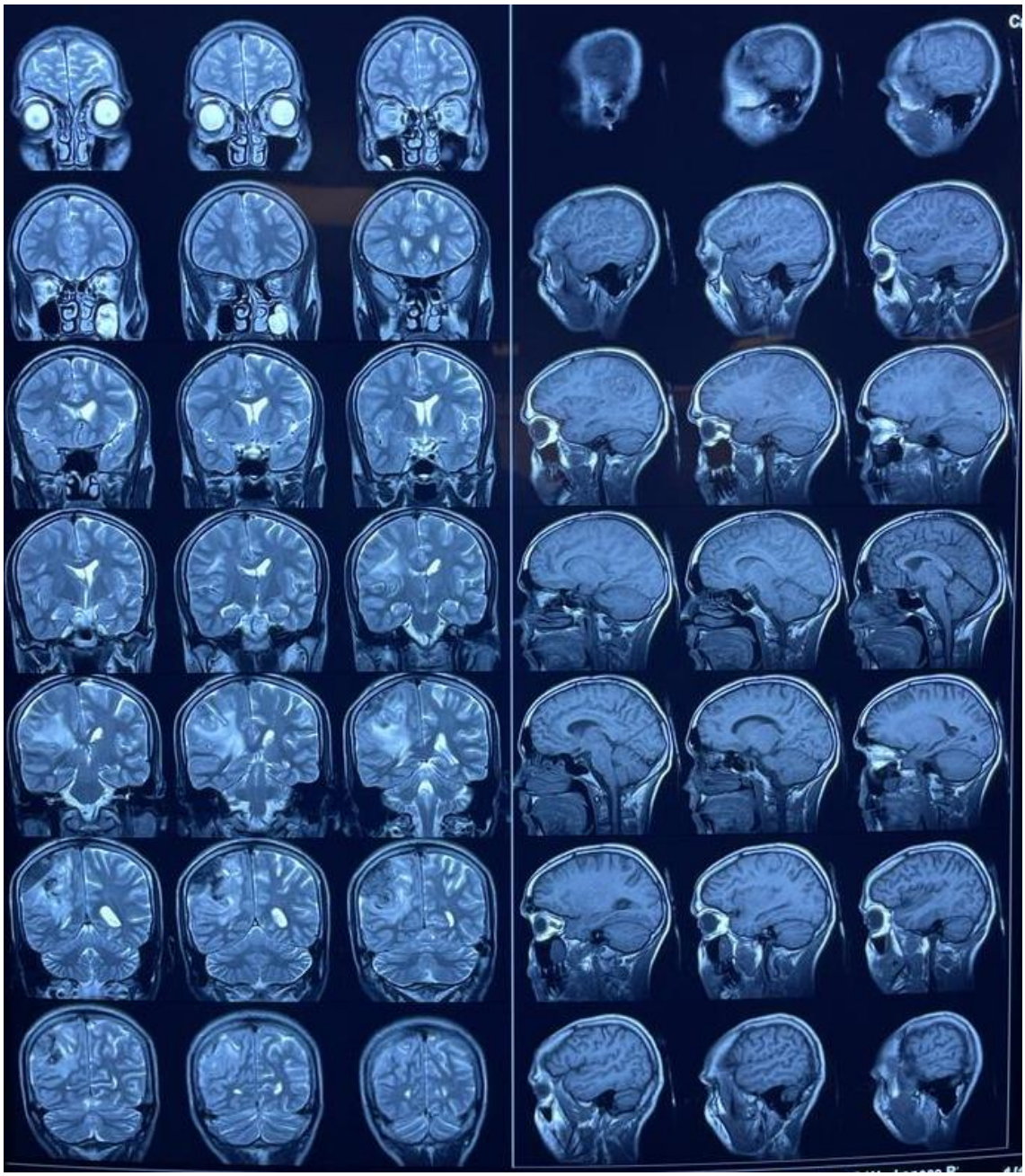

Fig. 1. Plain MRI showed approximately 7x5x4cm, ill defined, T1 isointense with T2 hypointense with broad base toward the dura along the convexity of the right parietal region

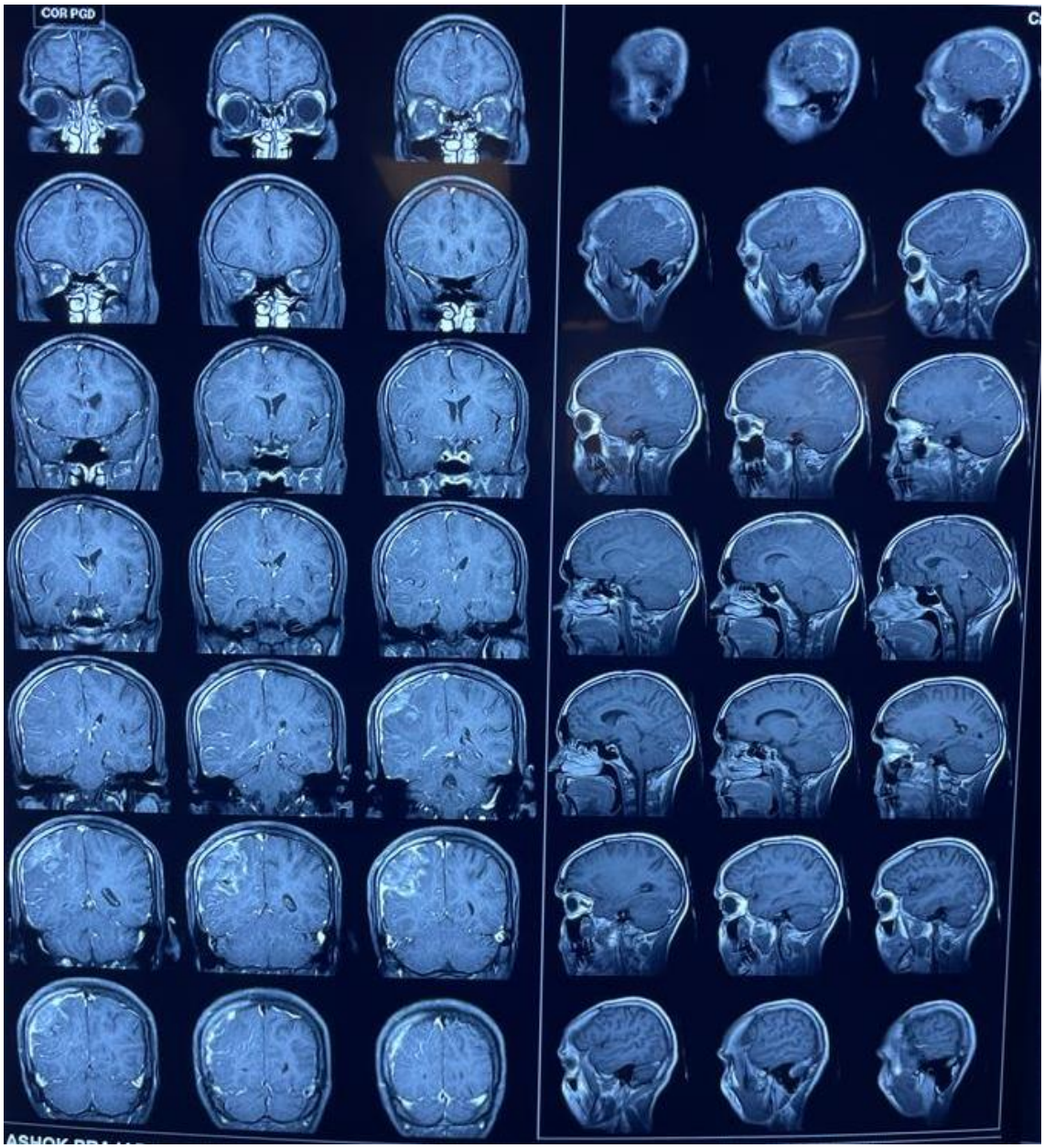

Fig. 2. On T1 post contrast, there was moderate enhancement with few non enhancing areas (heterogenous enhancement) which is causing buckling of the underlying parietal lobe

Meningiomas typically show hyperostosis, psammomatous calicifications, and enhanced vascular markings on plain radiographs. CT and MRI images show sessile or pedunculated isodense masses linked with the dura with distinctive 'mottling.' [13]. Papillary meningiomas have variable imaging findings but Lirng and collegues have suggested that MRI for papillary meningiomas shows a dura-based intracranial tumor with cystic component and enhancing solid tumor part [13].

Recurrences are not uncommon and lung is the most common site of extracranial metastases. To avoid recurrence or advancement of papillary meningiomas, aggressive excision is required. The role of adjuvant radiation is currently being debated. And the 5 year survival rate of papillary meningioma is just 40% as compared to the other types of meningiomas. Because aggressive tumour behaviour is associated with a poor clinical prognosis, early detection and treatments are critical [14].

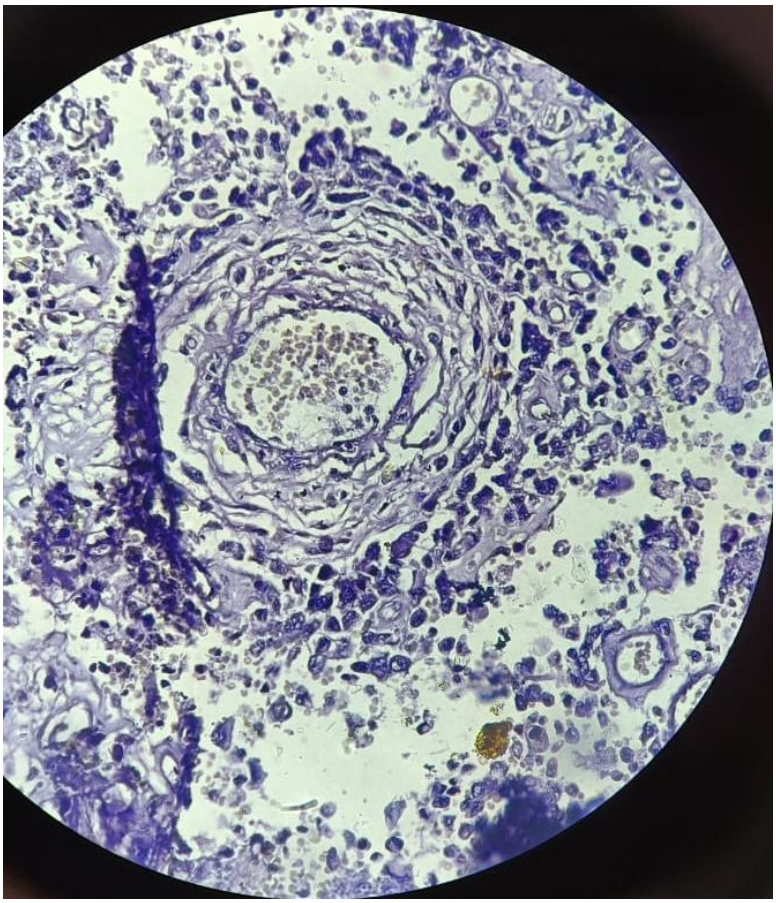

Fig. 3. On histological examination, the tumor showed a tissue composed of sheets of cells arranged in a predominantly papillary pattern with perivascular pseudo-rosettes

4. CONCLUSION

Papillary meningioma belongs to a very rare subtype of malignant meningioma. It has a very aggressive clinical course and is very prone to recurrence. Therefore, early diagnosis and timely appropriate management of the disease can significantly help improving the outcome of the disease positively.

CONSENT

All authors declare that 'written informed consent was obtained from the patient (or other approved parties) for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editorial office/Chief Editor/Editorial Board members of this journal.

ETHICAL APPROVAL

All authors hereby declare that all experiments have been examined and approved by the appropriate ethics committee and have therefore been performed in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki.

DISCLAIMER (ARTIFICIAL INTELLIGENCE)

Author(s) hereby declare that NO generative AI technologies such as Large Language Models (ChatGPT, COPILOT, etc) and text-to-image generators have been used during writing or editing of this manuscript.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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Peer-review history:
The peer review history for this paper can be accessed here:
https://www.sdiarticle5.com/review-history/126068