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Case Report

A rare case of falcotentorial meningioma type II of the pineal region

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Abstract

Background: Pineal region meningiomas are incredibly uncommon causes of pineal mass. Diagnosis is usually made when the tumours already reach huge diameters and cause mass effects, obstructing the ventricles and aquaducts.
Case Illustration: We present a case of a 32-year-old man complaining of worsening blurred visions and headaches. Brain Magnetic Resonance Imaging (MRI) disclosed a well-defined, lobulated pineal region mass that homogenously enhanced after contrast injection, with a dural adherence, close to the junction of the Vein of Galen and the straight sinus and inferior to the anterior edge of the tentorium, suggestive of a diagnosis of falcotentorial meningioma type II from the pineal region. The tumour compressed the mesencephalon and caused non-communicant hydrocephalus. The patient underwent tumour removal. Histological analysis of the tumour was consistent with meningioma WHO grade I.
Discussion : Pineal region meningioma, the falcotentorial type, can also be classified into four different types based on the tumour's adherence to the dura and the extension to its surrounding structures. Although rare, pineal region meningioma should be included as a differential diagnosis in a patient presenting with a pineal mass. MRI, both conventional and multimodal, still holds a crucial role in aiding clinicians and surgeons to make the correct diagnosis and deliver the best treatment option for the patients.
Conclusion : MRI can help differentiate pineal region meningiomas from intracranial and other pineal region tumours and aid the neurosurgeons in deciding the appropriate management approach for this rare intracranial neoplasm.

Keywords: *brain magnetic resonance imaging, meningioma, pineal region tumour, pineal region*

Abstrak

Latar Belakang: Meningioma daerah pineal adalah penyebab massa pineal yang sangat jarang terjadi. Diagnosis dapat ditegakkan ketika tumor sudah mencapai diameter yang besar dan menyebabkan efek massa, menghalangi ventrikel dan saluran air.
Ilustrasi Kasus: Seorang pria berusia 32 tahun yang mengeluhkan penglihatan kabur dan sakit kepala yang memburuk. Pada pemeriksaan Resonansi Magnetik Otak (MRI) didapatkan massa daerah pineal, lobulasi yang homogen setelah injeksi kontras, dengan perlekatan dura, dekat dengan pertemuan vena Galen dan sinus tentorial dan inferior ke tepi anterior tentorium, menunjukkan diagnosis meningioma falcotentorial tipe II dari daerah pineal. Tumor menekan mesencephalon dan menyebabkan hidrosefalus non-komunikan. Pasien menjalani pengangkatan tumor. Analisis histologis tumor konsisten dengan meningioma WHO grade I.
Diskusi: Meningioma daerah pineal, tipe falcotentorial, juga dapat diklasifikasikan menjadi empat tipe yang berbeda berdasarkan perlekatan tumor terhadap dura dan perluasan ke struktur sekitarnya. Meskipun jarang, meningioma daerah pineal harus dimasukkan sebagai diagnosis banding pada pasien yang datang dengan massa pineal. Pemeriksaan MRI baik secara konvensional maupun multimodal, masih menjadi baku emas dalam membantu dokter dan ahli bedah untuk membuat diagnosis yang benar dan memberikan pilihan pengobatan terbaik bagi pasien.
Kesimpulan: Pemeriksaan MRI dapat membantu membedakan meningioma daerah pineal dari tumor intrakranial dan tumor daerah pineal lainnya dan membantu ahli bedah saraf dalam memutuskan tatalaksana yang tepat untuk neoplasma intracranial.

Kata kunci: *daerah pineal, meningioma, pencitraan resonansi magnetik otak, tumor daerah pineal*

Background

Anatomically, the pineal region is located deeply, close to other vital structures in the brain's centre. It lies posteriorly along the midline, dorsal to the roof of the third ventricles and caudal to the tectum of the midbrain.[1,2] The incidence of the pineal region tumour is rare. It constitutes only 1% of all intracranial tumours in adult patients with various causes of lesions, such as tumours originating from pineal parenchymal tissue and various cells adjacent to the pineal gland, germinal cells, and metastatic tumours.[3-5] Although meningioma is the most commonly found central nervous system (CNS) tumour, pineal region meningioma is rare, accounting for only 2 – 8% of all pineal region tumours and 0,3 – 1,0% of all intracranial meningiomas.[2,6-8] Pineal region meningioma is characterized by meningioma that originates from falcotentorial junction or the posterior part of veluminterpositum and fills the quadrigeminal cistern with few or without adherence to the dura at all.[6,9] Pineal region meningiomas are predominantly benign with slow tumour progressivity. Thus, diagnosis is commonly made when the tumour is already large and elicits complications such as obstructive hydrocephalus.[10] Neuroimaging, particularly Magnetic Resonance Imaging (MRI), both standard and multimodal, still plays a pivotal role in diagnosing and differentiating benign and malignant mass in this deep, posterior part of the brain, including diagnosis of pineal region meningioma.[5]

Here, we report a rare case of a 32-year-old man with a falcotentorial meningioma type II of the pineal region.

Case Illustration

A 32-year-old-man had been complaining of blurred visions and headaches for three months. Blurry vision initially began with the left vision, followed by the right a month later. The Visual Analogue Scale (VAS) for his headache was 2-3. There were no vomiting and other neurology deficits, such as loss of consciousness, extremity weakness, or double vision. On examination, the patient was compos mentis with unremarkable vital signs. Both pupil size and pupillary light reflex were normal. However, his visual acuities were OD 6/30 and OS 1/300, and fundoscopy revealed bilateral papilledema. Other neurologic examinations found no contributory findings.

Magnetic Resonance Imaging (MRI) disclosed a sharply circumscribed mass lesion that was isointense and lobulated in the pineal region and enhanced after contrast injection (diameter, 3 x 3 x 3,5 cm), compressing the mesencephalon from the posterosuperior side. MRI also revealed noncommunicating hydrocephalus, deviated septum to the left, and an extra-axial cystic lesion in the left medial fossa with a differential diagnosis of an arachnoid cyst. Evans ratio was 0,4.

Serum biomarkers of pineal region tumours such as beta-HCG and alpha-fetoprotein (AFP) were normal, <1,2 mIU/mL and 2,3 ng/mL, respectively. Cerebrospinal fluid (CSF) analysis suggested clear colour, with cells count of 3 (3 MN, 0 PMN), CSF protein and glucose were 5 mg/dL and 79 mg/dL, respectively. Nonhematopoietic cells were not found.

The patient underwent a ventriculoperitoneal shunt surgery procedure and tumor removal. Tissue samples obtained from surgery confirmed our diagnosis of meningioma, WHO grade I. After surgery, the patient's vision and headache improved and he was later discharged and encouraged for routine follow-up visits.

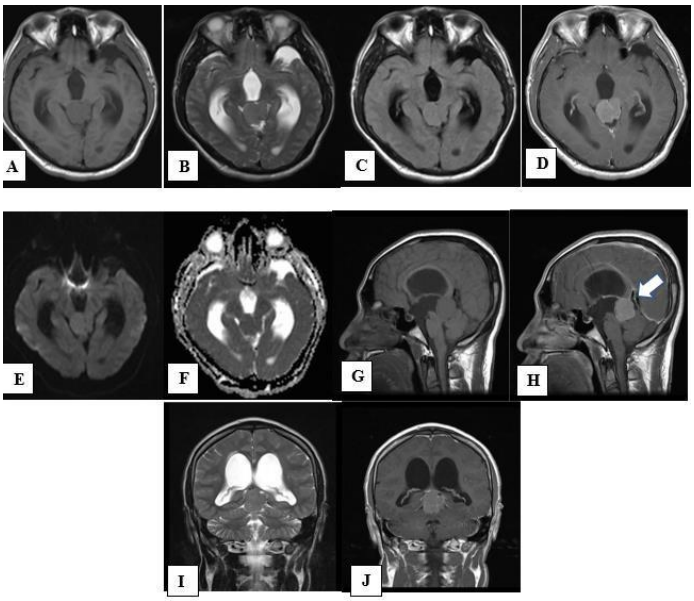

Figure 1. Brain MRI. **(A)** Axial T1-weighted, **(B)** Axial T2-weighted, **(C)** Axial FLAIR, **(D)** Axial T1weighted + gadolinium contrast, **(E)** Axial DWI, **(F)** Axial ADC image, (**G**) Sagittal T1-weighted, **(H)** Sagittal T1-weighted + gadolinium contrast, (**I**) Coronal T2-weighted, (**J**) Coronal T1-weighted + gadolinium contrast. Note that the tumor originated just beneath the anterior edge of the tentorium, near to the junction of galen vein and the straight sinus (white arrow), with a dural adherence apparent after contrast injection.

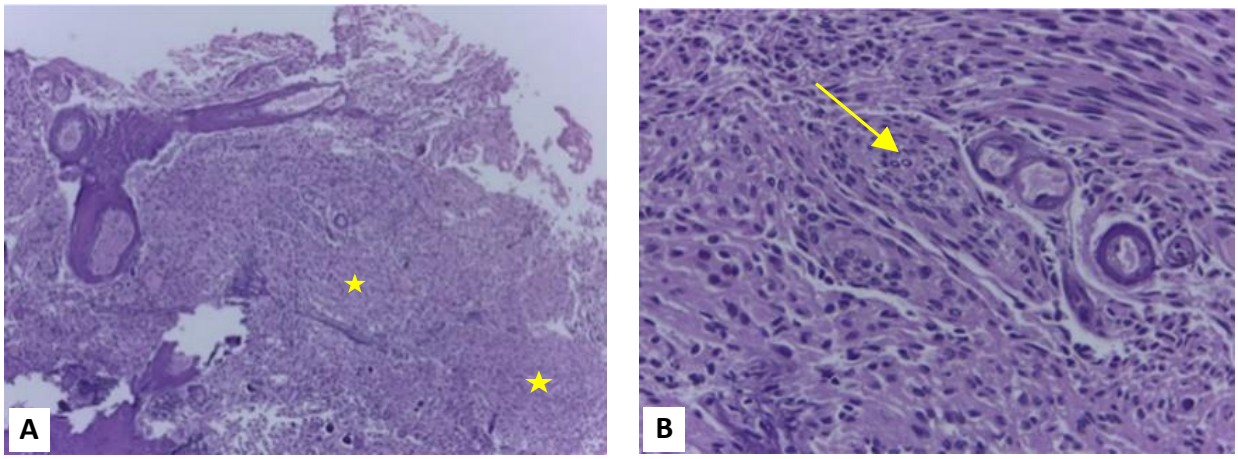

Figure 2. Histopathological finding of the tumor. (**A**) H&E 100X and (**B**) H&E 400x. The tumor shows lobular growth pattern (star). Tumor cells are largely uniform, with round to oval nuclei, fine chromatin and variable nuclear pseudo inclusions (arrow).

Discussion

As with other intracranial meningiomas, pineal region meningioma generally occurs in adulthood with a female predominance.[1,11] Some literature reports that the incidence of meningiomas increases with age, with a median age at diagnosis of 66 years.[12] Our patient presented a rarer case of pineal region meningioma in a 32year-old male.

In theory, pineal region meningiomas are broadly divided into two groups based on their origin: falcotentorial junction meningiomas and velum interpositum meningiomas. The clinical symptoms of the two do not show any difference. Thus, differentiation clinically is not plausible. The two lesions are distinguished by the relationship of the tumour to the dura at the falcotentorial junction. In falcotentorial junction meningiomas, meningiomas arise from the arachnoid membrane attached to the falcotentorial junction (dural folds of the tentorium and falx cerebri) and protrude anteriorly or posteriorly to the quadrigeminal cistern. Meanwhile, in the less common interpositum meningioma, the tumour originates from the arachnoid membrane that lines the velum interpositum.[2,6] Other literature also previously reported a case of pineal region meningioma arising from the arachnoid membrane of the vein of Galen.[6] Even with advanced imaging techniques, such as MRI and angiography, distinguishing these two types of tumours is often not possible, and the differentiation is usually confirmed after surgical operation.[2]

Pineal region meningioma, the falcotentorial type, can also be classified into four different types based on the tumour's adherence to the dura and the extension to its surrounding structures. In type I, the tumour derives from in the middle of both dural leaves of the cerebral falx, positioned superiorly to the Vein of Galen and the straight sinus junction. Type II originates from just beneath the anterior edge of the tentorium, closely positioned to the Vein of Galen and the straight sinus junction. Type III lateralized and located paramedian of one of the tentorial leaves. It originates from the dura and grows in a medial pattern towards the Vein of Galen. Type IV has a posterior direction. It is adherent to the dura in the falcotentorial junction down the straight sinus.[13] On MRI, our patient showed a meningioma closely adherent to the junction between the Vein of Galen and the straight sinus, inferior to the anterior edge of the tentorium, consistent with type II falcotentorial meningioma.

The course of the disease is generally slow, with an occult disease period of up to 25 years. Thus, pineal region meningiomas are often diagnosed when they have reached a large size and caused symptoms of mass effect and increased intracranial pressure. Obstructive hydrocephalus is caused by compression of the ventricles and aqueducts. Symptoms include headache, blurred vision, papilledema, gait disturbances, ataxia, cognitive impairment, hemifacial spasms, and Parinaud's syndrome (upward gaze paresis).[1,11,14] Consistent with the existing literature, our patient presented with symptoms of increased intracranial pressure in the form of gradually blurred vision and papilledema accompanied by headache. Furthermore, our patient did not have any extraocular movement disorders that are common in other tumours of the perineal region, confirming our suspicion of a pineal meningioma which, according to the available literature, rarely exhibits extraocular movement abnormalities.[7,11]

The radiological features of pineal meningiomas are nonspecific and similar to those of meningiomas at other CNS sites.[5,9] MRI generally shows a low to intermediate signal on T1weighted images, and the signal on T2-weighted images can vary, predominantly showing an intermediate to high signal on T2-weighted images. The lesions appear homogeneously enhanced after injection of gadolinium contrast. Because meningiomas are dura-based lesions, pineal region meningiomas exhibit a dura-tail appearance that differentiates them from other extra-axial tumours.[3,12] The presence of intratumoral haemorrhage, necrosis, or cysts with a more heterogeneous appearance indicates a more aggressive tumour.[12] On MRI examination, we found a solid, sharply-circumscribed, lobular, contrast-enhancing mass adherent to the dura that filled the pineal region, compressing the mesencephalon from a posterosuperior direction, suggestive of a mass of the pineal region origin. The tumour caused a mass effect and obstructed the normal flow of CSF, and caused noncommunicating hydrocephalus and deviated septum to the left. We also disclosed a cyst in the right and left temporal fossa, suggestive of an arachnoid cyst. The finding of an arachnoid cyst is incidental and is not related to the patient's clinical condition.

Serum and CSF biomarkers are performed to complete clinical, imaging, and pathologic findings. In our case, serum and CSF findings were unremarkable, lacking high CSF markers such as AFP and HCG. Thus, it furtherly excludes the possibility of

other pineal region tumours, such as germinoma, as the most prevalent type of all pineal region tumour that typically expressed high oncoproteins.[4]

Histopathological examination of the tumour confirmed the diagnosis of CNS WHO grade I meningioma. Based on histological criteria, the World Health Organization (WHO) in 2016 classified meningiomas into three grades: benign tumours (WHO grade I), and more aggressive meningiomas (WHO grade II), and malignant meningioma (WHO grade III).[8,12]

Definitive therapy is performed with surgery determined based on the relationship of the tumour to the deep venous system and surrounding structures, which are generally classified into two main approaches, namely the occipital transtentorial approach (OTA) and the supra-cerebellar infratentorial approach.[10,11] Due to its benign nature with slow disease progression, multistage and multidirectional surgery has also been reported and considered a safe approach for large pineal region meningiomas.[10] Most studies report improvement in patient's vision after tumour removal at the time of discharge from the hospital and follow-up.[14] In our case, the patient's headache and visual acuity had improved for both eyes after tumour removal.

Conclusion

Pineal region meningioma, falcotentorial meningioma type II in particular, is a rare case that ordinarily may not be considered a differential diagnosis in pineal region tumours. The diagnosis of pineal region meningioma is often a challenge for clinicians, surgeons, radiologists, and pathologists. MRI can help differentiate pineal region meningiomas from intracranial and other pineal region tumours and aid the neurosurgeons in deciding the appropriate management approach for this rare intracranial neoplasm.

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