

# A rare presentation of medulloblastoma in adults as primary leptomeningeal involvement

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## Keywords

Medulloblastoma, Children, Headache, Leptomeningeal Metastasis

## Abstract

Medulloblastoma is one of the most common primary tumors of the central nervous system of children but it is uncommon in adult age. We report a case of young male presented with a history of generalized headache that was found to have medulloblastoma with diffuse leptomeningeal involvement without prominent mass lesion. To our knowledge, our patient is the second reported case of medulloblastoma in adults identified with primary leptomeningeal involvement without a prominent mass lesion. The clinical, radiological and pathologic features of this entity are described.

## Introduction

Medulloblastoma is one of the most common primary tumors of the central nervous system of children but it is uncommon in adult age. Although subarachnoid seeding is found in up to 33% of all patients with medulloblastoma, primary leptomeningeal involvement is extremely rare.<sup>1</sup>

We report a case of young male presented with a

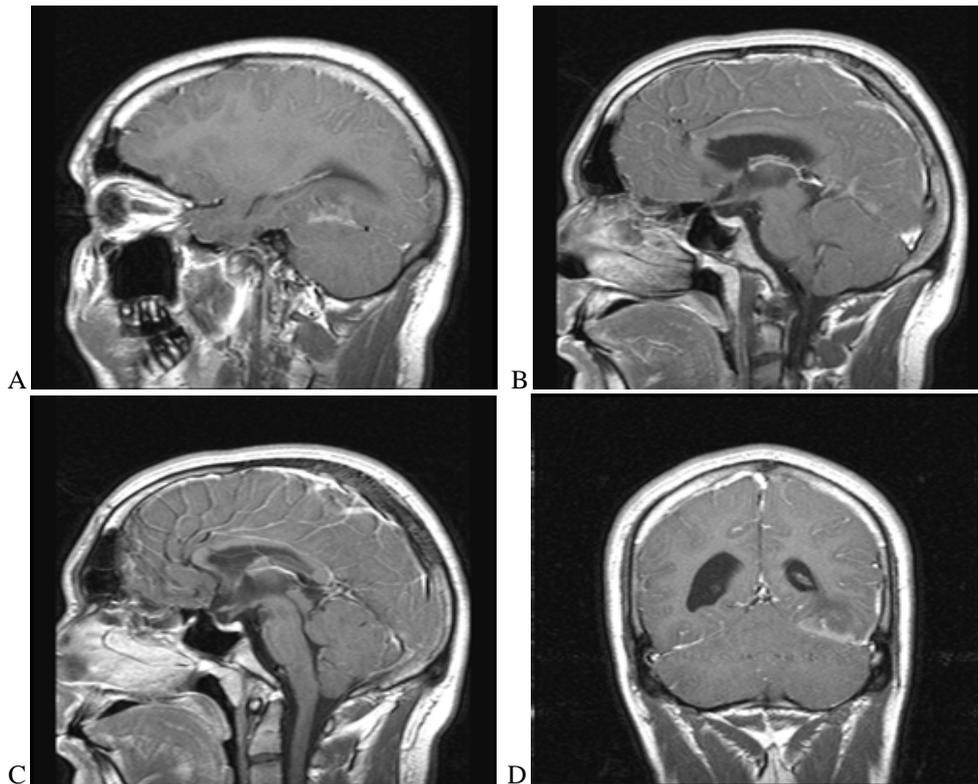
history of generalized headache that was found to have medulloblastoma with diffuse leptomeningeal involvement without prominent mass lesion. To our knowledge, our patient is the second reported case of medulloblastoma in adult identified with primary leptomeningeal involvement without a prominent mass lesion. The clinical, radiological and pathologic features of this entity are described.

## Case report

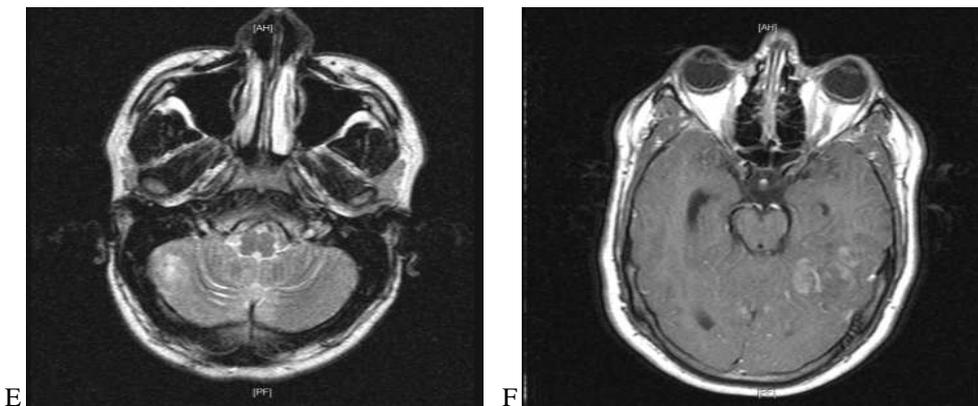
A 22-year-old male presented to our department with a 3-months history of generalized headache, hearing loss, tinnitus and blurred vision. Physical examination of the patient revealed decreased visual acuity (right eye 20/80 and left eye one meter finger count), bilateral papilloedema, bilateral hearing loss and bilateral upward plantar responses. Other neurological examinations including muscle strength, sensory and gait were normal.

Brain magnetic resonance imaging (MRI) with and without contrast showed intense dural enhancement of cerebellar folia and cerebral convexity, moderate hydrocephalus and intraparenchymal signal changes in right cerebellar hemisphere (Figures 1 and 2).

Lumbar punctures were done three times that



**Figure 1.** Sagittal (A, B, C) and coronal (D) post-gadolinium brain magnetic resonance imaging showing intense dural enhancement of cerebral convexities and tentorium



**Figure 2.** (E) Axial T2-weighted brain magnetic resonance imaging that shows abnormal signal changes within right cerebellar hemisphere. (F) Post-contrast axial brain MRI shows abnormal parenchymal enhancement

had negative results for gram stain, culture, Wright, angiotensin-converting enzyme, Indian ink, cerebrospinal fluid (CSF) adenosine deaminase, polymerase chain reaction for tuberculosis, CSF cytology and human immunodeficiency virus (HIV) antibody. CSF pressure was extremely high (about 70 cmH<sub>2</sub>O). CSF analysis showed normal cellular counts but elevated protein level and decrease glucose level was significant. Complete blood count, biochemistry profiles were normal. Erythrocyte sedimentation rate was 11 and C-reactive protein result was negative. Serum HIV antibody, HBs antigen, HCV antibody were all negative. Tuberculin purified protein derivative

(PPD) was performed that had negative result. Serum Wright, ANA, ds DNA, Antineutrophil cytoplasmic antibody (cANCA and pANCA) and antiphospholipid antibody were within normal limits, too. Paranasal sinus, chest, pelvic and abdominal CT-Scan with contrast all had normal results. Audiometric evaluation showed bilateral asymmetrical sensory neural hearing loss.

A suboccipital craniotomy was performed and the posterior fossa dura, pia arachnoid and cerebellar hemispheres were subject to biopsy. Histological examination of biopsy material showed highly cellular neoplasm that is composed of cells with

small to medium-sized, hyperchromatic nuclei and little apparent cytoplasm. Immunohistochemical stains performed on the biopsy material revealed positive reaction of tumor cells with neuron-specific enolase and glial fibrillary acidic protein and negative result with CD99. Synaptophysin shows weak positive reaction (Figure 3, A-H).

### Discussion

Medulloblastoma is a common brain tumor in children, accounting for 15-30 percent of all pediatric neoplasm of the central nervous system. In adults, however, the tumor is quite rare, accounting for only one to three percent of all primary brain tumors. Medulloblastoma has a classic radiological appearance on MRI as heterogeneous enhancing, well-defined mass lesions, often with surrounding cystic degeneration or necrosis. Although subarachnoid seeding is found in up to 33% of patients with medulloblastoma at time of diagnosis, primary leptomeningeal medulloblastoma is exceedingly rare.<sup>1</sup>

Ferrara et al. described a case of 5-year-old boy presented with primary leptomeningeal medulloblastoma of the posterior fossa without true mass lesion. The diagnosis was suspected first on the basis of the CT-scan findings, and was confirmed by cytological examination of the CSF and pathology.<sup>2</sup> Suman et al. described a 10-year-old girl with diffuse leptomeningeal involvement of posterior fossa with no mass lesion. The patient underwent biopsy.

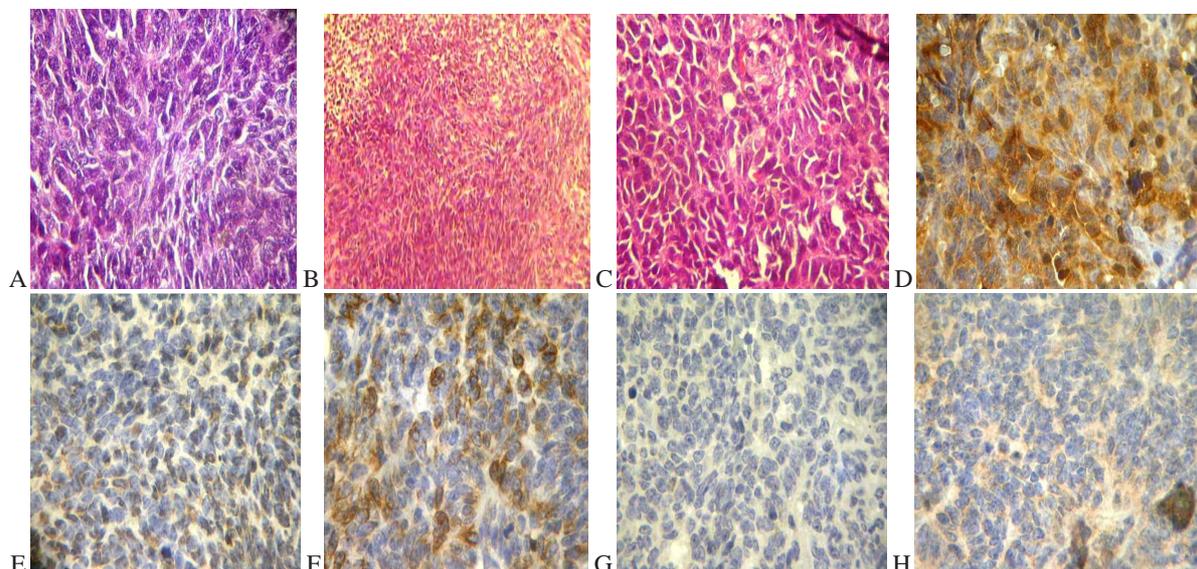
Histopathology was consistent with medulloblastoma.<sup>3</sup>

In another report, the authors reported a case of 8-year-old boy with primary leptomeningeal involvement of medulloblastoma and no mass lesion. The diagnosis was first documented on CSF cytological examination and then biopsy of the cerebellum and was later confirmed at necropsy.<sup>4</sup> Rushing et al. presented the first case of an occult leptomeningeal neoplasm in a 30-year-old man with imaging features of Chiari type I malformation that proved to have large cell medulloblastoma within the subarachnoid space.<sup>5</sup>

Although a peripheral primitive neuroectodermal tumor could not be completely be ruled out, it is unlikely because no mass lesion was discovered on whole body imaging studies. Previously reported cases showed rapid neurologic decline resulting in death within few weeks of initial presentation. Our patient had the same grave course.

### Conclusion

According to our literature review, this is the second reported case of medulloblastoma identified with primary leptomeningeal involvement without a prominent cerebellar mass in adults. In the case of leptomeningeal involvement, medulloblastoma should be considered as one of differential diagnosis. It could contribute to the development of more rapid diagnosis of patients with the same clinical presentation.



**Figure 3.** Histological examination of biopsy material

- A: Highly cellular neoplasm that are composed of cells with small- to medium-sized, hyperchromatic nuclei and little apparent cytoplasm (X40)
- B: Low power view
- C: Vague Homer-Wright rosettes
- D: Positive immunohistochemistry reaction for neuron specific enolase (NSE)
- E: Weak positive reaction for Synaptophysin
- F: Positive Glial fibrillary acidic protein (GFAP) reaction
- G: Negative epithelial membrane antigen (EMA) differentiate it from meningioma which is EMA positive)
- H: Negative CD99

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