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Case Report

Occipital Intraparenchymal Myxopapillary Ependymoma: Case Report and Literature Review

Abstract

Myxopapillary ependymoma (MPE) is a histological variant of ependymoma found in the conus medullaris or filum terminale region. Intracranial occurrence of the tumor is a rarity. The most characteristic histological feature of myxopapillary tumors is the abundance of intercellular and perivascular mucin and the arborizing vasculature, which tends to form papillae. We are reporting a 14-year-old patient presented with seizures caused by the right occipital region intraparenchymal lesion. Histopathology confirmed it to be MPE. Lesion was excised completely. Literature reviews on the topic are discussed regarding the histological findings, natural history, and outcome of surgically treated MPE. This is the fifth reported case of cerebral intraparenchymal primary MPE.

Keywords: *Intraparenchymal, myxopapillary ependymoma, pseudorosettes*

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Introduction

Myxopapillary ependymoma (MPE) is a histological variant of ependymoma found in the conus medullaris or filum terminale region. Intracranial occurrence of the tumor is a rarity. According to the WHO classification, spinal cord MPEs are grade 1 tumors with favorable outcome after complete resection even though metastasis present. In literature, four cases are reported for cerebral intraparenchymal MPEs with no ventricular extension. We are reporting fifth case of occipital region MPE.

Case Report

A 14-year-old male patient presented with two episodes of seizures (generalized tonic–clonic) within 6 months duration. His medical history was unremarkable. On neurological examination, there was no deficit. There was no cranial nerve involvement, sensory motor deficits, gait changes, vision deterioration, gait changes, or higher mental function derangements.

Magnetic resonance imaging (MRI) of the brain demonstrated a mass hypointense on T1-weighted imaging, hyperintense on fluid-attenuated inversion recovery imaging in the right occipital lobe which was heterogeneously contrast enhancing [Figure 1]. Low-grade glioma was considered as the first diagnostic entity and operative intervention was planned. The patient was operated in prone position, right occipital craniotomy, and tumor near total resection done. Frozen section report was low-grade glioma.

Histopathological consideration

Histological examination of tumor specimen stained with hematoxylin and eosin showed a cellular tumor consisting of medium-sized neoplastic cells arranged around stromal vessels and forming papillary structures. Variable amount of mucoid material seen around blood vessels with red blood cells in vascular lumen. There were areas of pseudorosettes formation also consistent with MPEs [Figure 2]. For publication of data, we took consent from patient and approval from institute.

Postoperative course

In view of histopathological findings, MRI of the total neuraxis was performed, which did not reveal any lesion, particularly of the conus medullaris or filum terminale [Figure 3]. The patient recovered well after surgery. Regular follow-up with MRI of total neuraxis is planned. No adjuvant chemotherapy or radiotherapy is planned for this patient.

Discussion

Ependymomas make up 6% of all gliomas and account for 1%–9% of

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intracranial tumors and 60% of spinal cord gliomas.[1] Intracranial location of ependymoma is more common in children, whereas spinal cord is more involved in adults.[2] Virchow first described an ependymoma, which consisted of ependymal cells forming ependymal rosettes and perivascular pseudorosettes.[3] The term MPE was first used by Kernohan showing an ependymoma with heavily branching papillary form, hyalinized blood vessels, and abundant intercellular and perivascular mucin.[4,5] Histopathologically, MPEs are low-grade tumors as per the WHO classification they are classified as grade 1 tumors.[6]

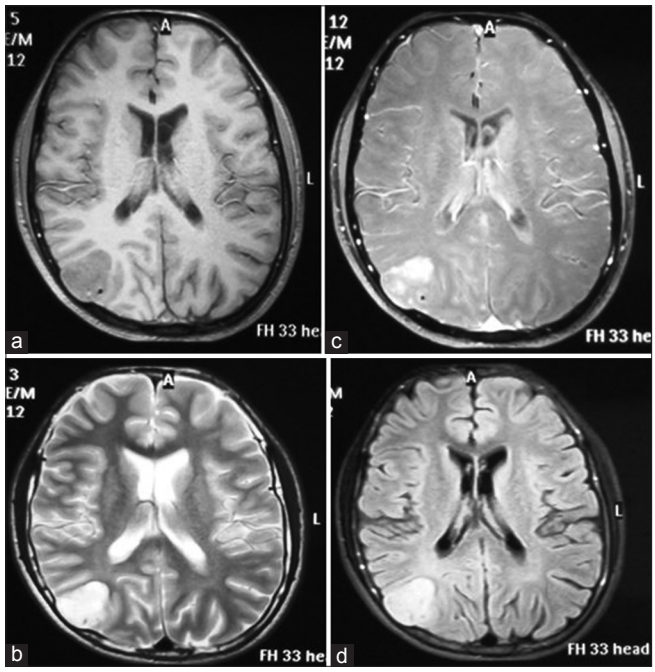

Figure 1: Magnetic resonance imaging brain showing axial image of hypointense mass on T1-weighted image (a), hyperintense mass on T2-weighted image (b), which is heterogeneously contrast enhancing (c), and hyperintense on fluid-attenuated inversion recovery sequence (d)

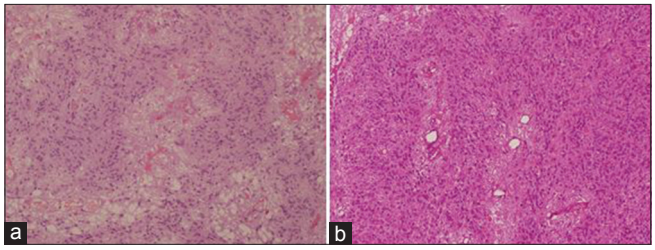

Figure 2: Histopathological examination show on hematoxylin and eosin preparation (a) medium-sized neoplastic cells arranged around stromal vessels and forming papillary structures with variable amount of mucoid material, (b) pseudorosettes formation

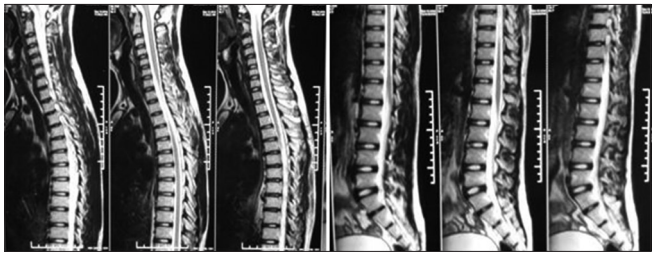

Figure 3: Magnetic resonance imaging spine showing T2-weighted images of magnetic resonance imaging spine showing no lesions

The usual age of presentation is from third to fifth decade.[7] Intradural MPEs almost exclusively occur at the conus medullaris or filum terminale. Extraspinal MPEs most commonly present as subcutaneous soft tissues in the sacrococcygeal region.[8-10] These are thought to arise as a result of neoplastic transformation of ependymal rest cells in this area.[10] Other rarer sources include the peritoneum-pelvic regions involving the ovary, broad ligament, meso-ovarium and omentum, and the lungs and mediastinum.[11] Reported cases show variable intracranial distribution including lateral ventricle,[5,12,13] forth ventricle,[7,14] cerebellopontine angle,[15] medulla oblongata,[16] falx cerebri,[13] and intraparenchymal.[4,17-19] Only five cases are reported for cerebral intraparenchymal location, including this case [Table 1].

MPEs are uncommon and make up approximately 15% of all ependymomas.[20] The microscopic appearance of MPE is characterized by pseudopapillary formation with symmetrical zones of mucoid matrix surrounding branching tumescent vessels as well as accumulation of mucin within and between tumor cells.[20] The mean proliferating index of this subtype is 2.4 versus 21% for anaplastic ependymomas.[21]

Although MPEs are grade I tumors with a tendency for slow growth and local recurrence, they are capable of spread within the central nervous system and extraneural metastasis.[22,23] There are occasional case reports of local bony invasion and peritoneal seeding.[23,24] MPE of the cauda equina region commonly recurs locally, often after an interval of many years, but the prognosis is good even after local recurrence or subarachnoid metastasis. Intracranial metastases of MPEs are rare, especially in the pediatric population.[25] Although the quoted recurrence rate of spinal MPEs ranges from 10% to 40% (with or without subset analysis of completeness of resection), the recurrence rates in children may be as high as 50%.[20,26-28]

There are no clear guidelines in the management of primary intracranial MPEs. These lesions are considered to be more benign than general ependymomas as being the WHO grade 1 tumor. No prospective study is available in the management of intracranial MPEs; however, the

Table 1: Reported cerebral intraparenchymal myxopapillary ependymomas

Author	Age/sex	Location
Maruyama *et al*.[17]	8/female	Right occipital lobe
Ralte *et al*.[18]	22/male	Left temporal lobe
Tzerakis *et al*.[4]	68/male	Left frontal lobe
Patangia *et al*.[19]	21/female	Right frontal lobe
Current case	14/male	Right occipital lobe

management strategies are guided by experience in treatment of spinal MPEs. Albeit from literature some conclusions can be drawn which are as follows:

- There are patients who are disease-free for many years after gross total resection without radiation therapy[3]
- Complete removal is the best therapeutic option[27-29]
- Recurrence and subarachnoid or systemic metastasis are possible even after gross total resection and radiation therapy. It is possible to achieve a good outcome with subtotal resection and irradiation[30,31]
- Histological features are not generally helpful in determining the prognosis of MPEs[20]
- There is no difference in outcome for irradiated versus nonirradiated patients, and this fact would suggest that radiation therapy is not effective although the consensus yet to be achieved. More recently, it has been shown that patients with spinal MPEs who received immediate postoperative radiation therapy (≥45 Gy) had 5- and 10-year progression-free survival rates that were significantly better than those of patients who did not receive any adjuvant therapy, including patients thought to have undergone a total resection.[29,31]

On the basis of these data, it is recommended that radiation be delayed until recurrence tumors undergoing gross total resection. Local radiation therapy may be indicated in subtotal resected tumors or recurrence (controversial). In the case of our patient reported here, no adjuvant radiation therapy is planned because serial neuraxis imaging does not demonstrate residual or recurrent pathology at this time.

Conclusion

This is the fifth reported case of histologically proven primary cerebral intraparenchymal MPE. It is hypothesized that intracranial MPEs arise from rests of fetal ependymal cells that migrated from periventricular regions. Although rare, MPEs outside of the filum terminale do exist.

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Conflicts of interest

There are no conflicts of interest.

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