

Primary Intracerebral Myxopapillary Ependymoma: A Rare Case Report

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ABSTRACT

Myxopapillary ependymoma (MPE) is a variant of ependymoma occurring almost exclusively in the conus medullaris or filum terminale. Myxopapillary ependymoma primarily in the brain is extremely rare. Here we report the case of a 21 year female with a right frontal myxopapillary ependymoma which did not demonstrate any connection to the lateral ventricles. Patient is presented with complaint of headache and vomiting with no history of convulsion, loss of consciousness. On radiological examination, MRI of brain revealed an ill-defined, cystic mass lesion in frontal lobe. On histopathological examination it was reported as myxopapillary ependymoma. This is the sixteenth reported case of histologically proven primary myxopapillary intracranial ependymoma and fourth cases of supratentorial intraparenchymal lesion.

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Introduction

Myxopapillary ependymoma was first described by Kernohan in 1932^[1], as a variant of ependymoma occurring in the conus medullaris or filum terminale exclusively^[2]. Although it can affect patients of all ages, they are most frequent in young adults. Bailey and Cushing recognised ependymomas as an independent entity in their first brain tumour classification (1926). Myxopapillary ependymoma occurring primarily in the brain parenchyma is extremely rare and only Fifteen cases have been reported in the literature till date.^[3-16]

The characteristic histological feature of myxopapillary ependymomas are the juxtaposition of pseudo-papillary structures consisting of a centrally located vessel surrounded by a ring of mucin lined by cuboidal ependymal cells. The most characteristic histological feature is the abundance of intercellular and perivascular mucin and the arborizing vasculature, which tends to form papillae. Myxopapillary ependymoma should be a possible differential diagnosis whenever an intracranial cystic tumor is found.

Case History

A 21 year female patient presented with complaint of headache and vomiting with no history of convulsion, loss of consciousness. On radiological examination, MRI of brain revealed an ill-defined, cystic mass lesion measuring 6.2 x 5.9 x 4.8cm involving cortical and sub cortical region of right frontal lobe; these areas appeared hypointense to gray matter on T1, hyperintense on T2W and FLAIR images; on post contrast images cystic lesion show rim enhancement with central non-enhancing areas. Small mural nodule also seen. Faint calcification seen within the lesion. There is mild perilesional white matter edema not

showing enhancement thus favouring vasogenic edema. Ventricles were normal but there was a mid-line shift to left side. (Figure 1 & 2) Based on MRI findings differential diagnosis given were primary brain tumor, ganglioglioma/ cystic oligodendroglioma/ low grade astrocytoma. Patients complete blood counts and serum biochemistry were within normal limits. Patient was subjected to excision of mass with duroplasty.

On gross examination multiple soft tissue pieces were received of which largest tissue measured 4 x 3 x 2.5 cm with presence of normal brain tissue. On microscopic examination sections revealed characteristic finding of myxopapillary ependymoma (Figure 3 & 4) i.e. presence of papillary architecture with abundant perivascular mucin and areas of calcifications (psammoma bodies) (Figure 5 & 6). Alcian blue staining demonstrated perivascular mucin. X-Ray spine excluded a primary tumor in filum terminale region which is the most common site thus ruling out the possibility of intracerebral metastasis.

Intervention: The lesion was totally resected. After surgery, the patient was neurologically intact and had an uneventful recovery. On post-operative follow up CT scan of brain revealed acute extradural hemorrhage in right fronto-parietal region causing mass effect over adjacent cerebral parenchyma. Mild shift to left is present. Other regions of cerebral parenchyma, cerebellum and brainstem appears normal.

Discussion

Ependymomas are tumors having origin from the ependymal cells lining cerebral ventricles, spinal central canal, and cortical rests.^[17,18] The most common location for ependymomas is the 4th ventricle, followed by the spinal

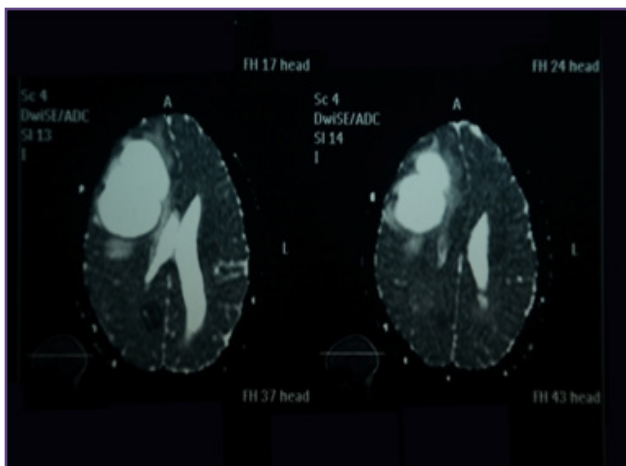


Fig. 1: MRI scan showing cystic lesion in right frontal lobe which is well separated from lateral ventricle.

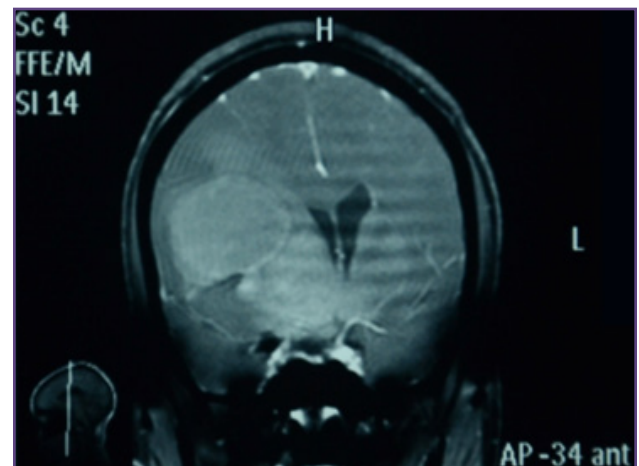


Fig. 2: MRI Scan reveals a cystic lesion with sparing of ventricles and midline shift.

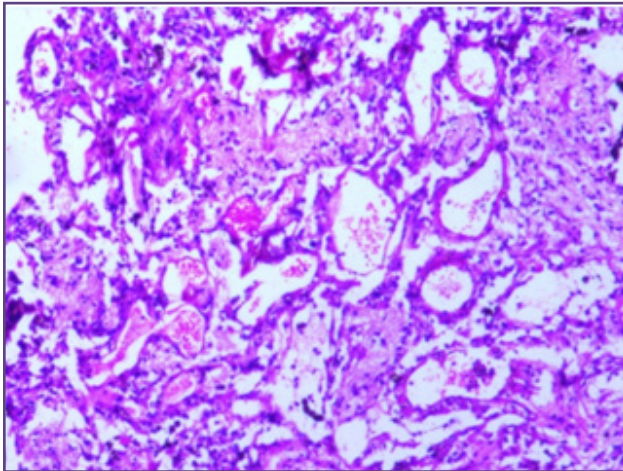


Fig 3. H&E section (10x) showing pseudopapillary structures consisting of a centrally located vessel surrounded by a ring of mucin lined by cuboidal ependymal cells.

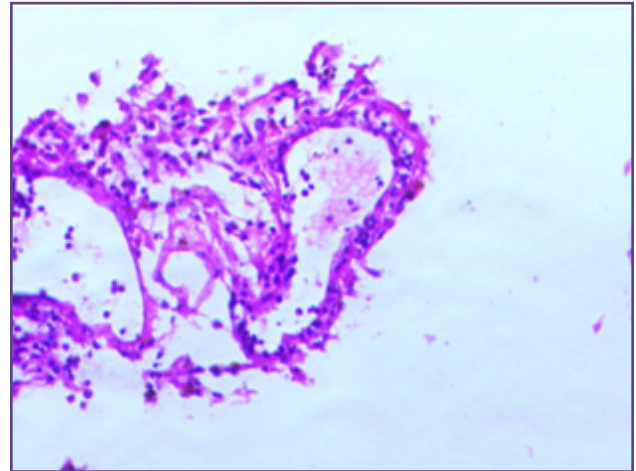


Fig. 4: H&E section (20x) showing pseudopapillary structures consisting of a centrally located vessel surrounded by a ring of mucin lined by cuboidal ependymal cells.



Fig. 5: Alcian blue (20x) staining showing perivascular mucin.

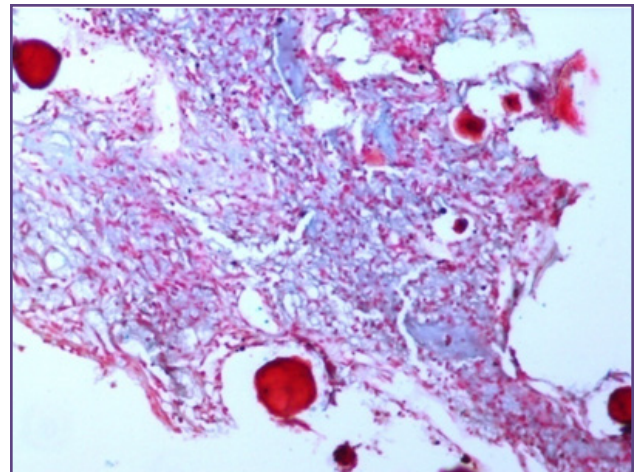


Fig 6. Alcian blue staining (10x) showing abundant mucin and psammoma bodies are also present.

cord, filum terminale/cauda equina, and supratentorium of the brain ^[19]. Occasionally it may be seen in the subcutaneous tissue overlying the sacrum. ^[20]

Myxopapillary ependymoma is a variant of ependymoma, characterized microscopically by a papillary arrangement of neoplastic cells surrounding a fibrovascular core containing both hyalinized blood vessels and myxoid degeneration. Histologically, it is a benign tumor ^[12] (WHO grade 1) with a peak incidence in the 4th decade of life which is generally confined to the lumbosacral region of the spinal cord. It constitutes 27% of ependymomas occurring in the spinal cord. ^[1,2] It may rarely disseminate via the CSF pathways into the brain and be mistaken as a primary intracranial myxopapillary ependymoma. ^[22-24] It is therefore important to rule out the possibility of metastasis from a spinal

myxopapillary ependymoma before making a diagnosis of primary intracranial myxopapillary ependymoma.

Including the presented case, 16 primary intracranial myxopapillary ependymomas have been reported ^[3-16]. Present case is the fourth reported case of intracranial intraparenchymal myxopapillary ependymomas. ^[4,7,9] Nine tumors were located supratentorially while seven were infratentorial. Of the supratentorial group, four were intraventricular (3 from lateral ventricle and 1 from third ventricle), four were intra- parenchymal and one was transependymal. Among infratentorial group 3 were in fourth ventricle, 2 in cerebellopontine angle, one in each medulla and cerebellum. (Table 1)

All of them were treated by total surgical resection. Radiological features of these tumors were all well-

Table 1: Reported cases of Intracranial Myxopapillary Ependymoma.

Author (Year)	Age	Sex	Site	Location
Sato et al (1983) ³	29	M	Right Lateral Ventricle	ST
Maruyama R et al (1992) ⁴	8	F	Right occipital lobe	ST
Warnick RE et al (1993) ⁵	37	F	Lateral Ventricle	ST
Matyja E et al (2003) ⁶	37	M	Right Lateral Ventricle	ST
Ralte et al (2004) ⁷	22	M	Left temporal lobe	ST
Tseng Y-C et al (2004) ⁸	7 20	F F	Fourth ventricle Bilateral cerebral falx	IT ST
Tzerakis et al (2004) ⁹	68	M	Left frontal lobe	ST
Lim SC et al (2006) ¹⁰	62	F	Fourth ventricle	IT
Sparaco M et al (2009) ¹¹	30	M	Left CPA	IT
DiLuna ML et al (2010) ¹²	8	M	Medulla	IT
Shapey J et al (2011)¹³	40	M	Right CPA	IT
Margetis K et al (2011) ¹⁴	–	–	Cerebellar	IT
Chakraborti S et al (2012) ¹⁵	50	F	Fourth ventricle	IT
Wang M et al (2013) ¹⁶	22	M	Third ventricle	ST
Present study	21	F	Right Frontal lobe	ST

CPA- cerebellopontine angle; ST-supratentorial; IT- infratentorial

demarcated primary cystic masses with strong enhancement at their solid part and/or cystic walls. Intratumoral calcification or hemorrhage, commonly seen in ependymomas were also seen. However, these imaging features are still not specific enough to differentiate from other types of ependymomas or other primary cystic intracranial tumors such as astrocytoma, primitive neuroectodermal tumor, and choroid plexus papilloma. In conclusion, myxopapillary ependymoma can occur as a primary in the brain parenchyma or even extra-axial space.

Conclusion

This is the sixteenth reported case of histologically proven primary myxopapillary intracranial ependymoma and fourth case of intaparenchymal MPE. In conclusion, myxopapillary ependymoma can occur in the brain parenchyma or even extra-axial space so whenever a primary cystic intracranial mass is suspected, myxopapillary ependymoma should be always be a differential diagnosis.

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Competing Interests

None Declared

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