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ABSTRACT

Pilocytic astrocytoma (PA) is a subset of gliomas characterized by a benign course with an excellent prognosis and rarely metastasizing or spreading along the neuraxis. We report a case of a 56-year female with intraventricular dissemination of pilocytic astrocytoma in an adult and discuss the clinical significance diagnosis and management including the peculiar pattern of dissemination of the pilocytic astrocytoma. The course of the disseminated disease may not be as good as that of patients with localized recurrence or totally resected primary disease and can vary from rapid progression to prolonged stabilization.

INTRODUCTION

Pilocytic astrocytoma (PA) is subset of gliomas characterized by a benign course with an excellent prognosis and rarely metastasizing or spreading along the neuraxis. 1-14 Dissemination of a pilocytic astrocytoma is a rare event that occurs in 2-4% of cases particularly in children. 6, 8, 11 We report a case of intraventricular dissemination of pilocytic astrocytoma in an adult and discuss the clinical significance diagnosis and management including the peculiar pattern of dissemination of the pilocytic astrocytoma.

CASE REPORT

A 56-year old female patient presented with progressively increasing headache, vomiting and blurring of vision without any focal neurological deficits or seizures. Her general and systemic examination was unremarkable. Neurologically she was conscious, alert and oriented to time, place and person. Higher mental functions were normal. She had bilateral papilloedema and other cranial nerves were normal. Motor and sensory examination was normal. Her general and systemic examination was normal. The patient underwent MRI brain and it showed minimally contrast enhancing bilateral thalamic lesions, with intraventricular spread in third ventricle and the aqueduct causing obstructive hydrocephalus (Figure 1, 2 and 3). The patient underwent

Keywords

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endoscopic exploration of the third ventricular lesion that could be excised as well as the lesion in the aqueduct also could be excised. Part of the lesion in the fourth ventricle was left alone as it was densely adherent to the surrounding structures. Intra-operatively external ventricular drainage was (EVD) was inserted and was kept in situ for 5 days till the CSF was acceptably clear. She later underwent right ventriculo-peritoneal shunt surgery.

Histopathological examination of the section and additional made serial deeper section showed few fragments bits of cellular neoplasm of astrocytic origin. The tumour composed admixture of compact cellular and oedematous loose-knit tissue showing scattered protoplasmic astrocytes having fairly uniform round to oval or slightly elongated nuclei with delicate - open chromaWr pattern and barely discernible cytoplasm. The fibrillary background showed few microcytic spaces and vascular proliferation comprising few ectatic congested blood vessels Coupie of foe show doubtful Rosenthal fibers (Figure-4). No granulomas were seen. Histopathological features were consistent with - 'pilocytic astrocytoma'. Section shows a tumor of moderate cellularity with cells amidst fibrillary background. Microcystic change, hyalinized vasculature were seen. No mitosis, necrosis, microvascular proliferation seen. Rosenthal fibres and eosinophilic granular bodies were seen. The tumor cells express GFAP & P 53 (focal) and were Immunonegative for synaptophysin 8i EMA. The Mib-1 labeling index was approximately 1%. Low grade astrocytic neoplasm suggestive of pilocytic astrocytoma. In post-operative made she did well as she was conscious, with no focal neurological deficits.

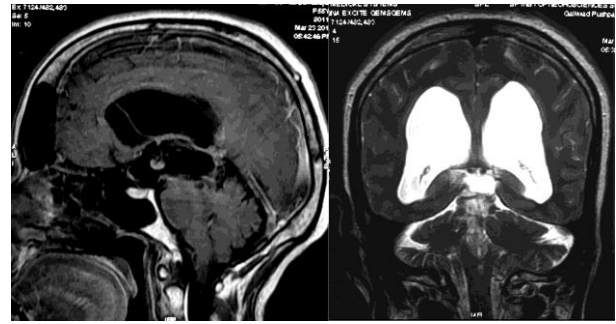


Figure 2. MRI brain T1W sagittal images and T2W coronal image showing the blockage of aqueduct by tumour.

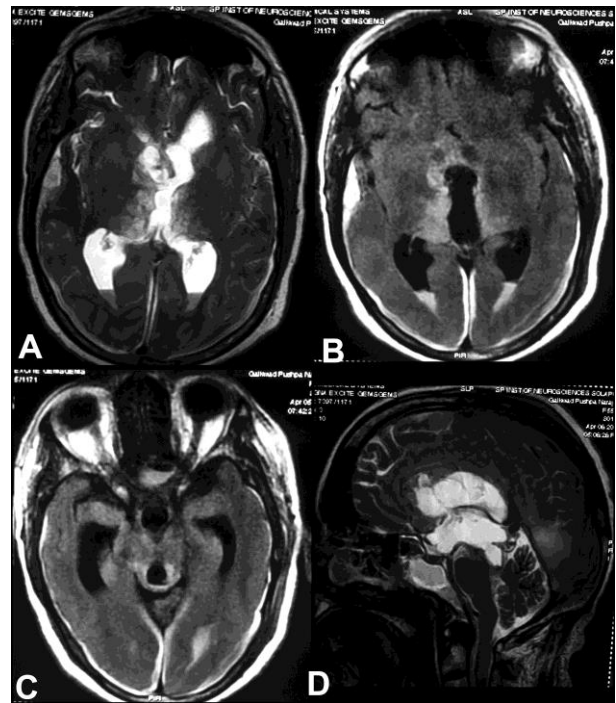


Figure 3. Post-operative MRI T2W and FLAIR axial images showing although the tumor could be removed from upper part of the aqueduct but still the lower part is closed.

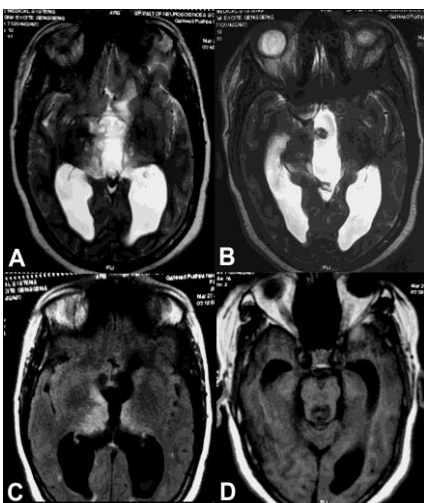


Figure 1. MRI brain T1W, T2W and Flair axial images showing bilateral thalamic tumor with blockage of the aqueduct and associated hydrocephalus.

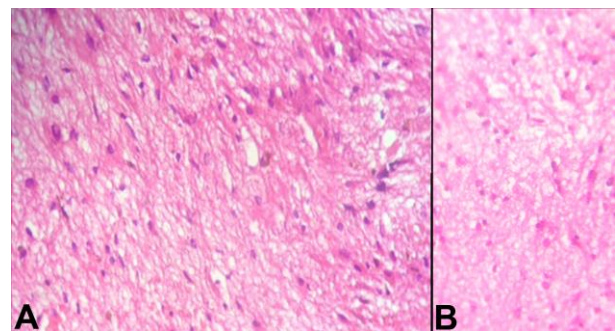


Figure 4. Histopathological examination of the section and additional made serial deeper section showed few fragments bits of cellular neoplasm of astrocytic origin. The tumour composed admixture of compact cellular and oedematous

loose-knit tissue showing scattered protoplasmic astrocytes having fairly uniform round to oval or slightly elongated nuclei with delicate - open chromatin pattern and barely discernible cytoplasm. The fibrillary background showed few microcystic spaces and vascular proliferation comprising few ectatic congested blood vessels. Couper of foie show Rosenthal fibers.

DISCUSSION

Dissemination of primary central nervous system neoplasms along the neuraxis is commonly associated with high grade lesions i.e. medulloblastoma, germ cell tumors and malign glial tumors.^{1-5,11,15-18} Dissemination of low-grade gliomas has been documented only in few cases.^{1-14, 19} Usually the site of dissemination is spinal^{1, 6, 8, 11, 16, 19-23} but in rare cases it can be intraventricular metastasis leading to hydrocephalus.²¹ Clinically it can occur after a long postoperative period or may be the first sign of disease or of relapse.^{1, 6, 8, 11, 16, 19-25} The clinical presentation ranges from asymptomatic cases to hydrocephalus, meningismus, worsening of focal deficits, new onset of neurological deficits and onset of seizures.^{8, 11, 19, 24} The tumour spread in these patients is via the CSF and it is accepted that low grade astrocytomas in proximity to ventricles or CSF cisterns are more likely to spread than deeply located tumors.^{21, 26, 27} Several mechanisms have been hypothesized to explain the spread of intracranial tumors by CSF pathways including malignant transformation, cellular anaplasia, surgical manipulation, natural history, multiplicity and presence of cell adhesion molecules (CD44 adhesion molecule as it may play a role in astrocytic invasion and adhesion).^{7, 28} It has been suggested that the tumor mass located in the floor of the third ventricle may breach the ependyma, invading the ventricular cavity and thus resulting in to ependymal or leptomeningeal seeding.^{2, 3, 5, 8, 9}

Pilocytic astrocytoma is a benign tumor that corresponds to histological Grade I^{13, 29-31} and associated with 20-year survival rates of greater than 90% in patients who undergo total excision of the lesion.^{8, 13, 29, 30} Presently there is not much known about the optimum treatment and course of disseminated low-grade astrocytomas.^{1, 2, 14, 16, 32} Probably hydrocephalus, biopsy and partial resection may also be additional favorable factors, although this remains unproven.^{8, 14} It has been suggested that the total resection must be performed as often as possible and no adjuvant therapy should be carried out for low grade gliomas,

^{13, 14} however the treatment of the disseminated tumor, remains controversial.^{14, 20, 25} The course of the disseminated disease may not be as good as that of patients with localized recurrence or totally resected primary disease and can vary from rapid progression to prolonged stabilization.^{1, 2, 14, 16, 25, 32, 11, 20, 21}

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