Case Report

Chiasmatic pilocytic astrocytoma in childhood with leptomeningeal dissemination at recurrence: A case report and review of literature

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ABSTRACT

Pilocytic astrocytoma is a benign variety of gliomas, which rarely spreads along the neuraxis. Leptomeningeal dissemination of pilocytic astrocytoma in children, either at the time of initial presentation or at recurrence, is a rare event. We report a case of chiasmatic pilocytic astrocytoma in a 4-year-old child with leptomeningeal dissemination at recurrence. A review of the literature is done regarding dissemination patterns, clinical picture, treatment, and outcome of pilocytic astrocytoma with leptomeningeal dissemination.

Key words: Chemotherapy, leptomening ealdissemination, pilocytic astrocytoma, radiation therapy

INTRODUCTION

Leptomeningeal dissemination (LMD) of primary central nervous system (CNS)tumors in children has been reported mainly in medulloblastomas, ependymomas, germ-cell tumors, primitive neuroectodermal tumors, high-grade gliomas and rarely in low-grade gliomas. Pilocytic astrocytoma (PA) is a benign variety of glioma, which rarelyspreads along the neuraxis.[1,2] LMD of pilocytic astrocytoma (PA) in children is much more uncommon. Dissemination patterns, clinical picture, treatment, and outcome are still poorly understood. We report a case of chiasmatic pilocytic astrocytoma in a 4-year-old child with leptomeningeal dissemination at recurrence.

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CASE REPORT

A 4-year-old Asian male child presented with complaints of headache and vomiting for last 3 months. On neurological examination, the child was irritable, the optic disks were atrophic, and there was no gross sensory motor deficit or cranial nerve palsy. Magnetic resonance image (MRI) of brain revealed a sellar and suprasellar mass lesion, which was hypointense on the T1-weighted image, hyperintense on T2-weighted image and intensely enhancing with gadolinium contrast [Figure 1]. Right frontal craniotomy and partial decompression was done by the subfrontal approach. Histopathological examination of the operated specimen showed fusiform cells with wavy fibrillary processes

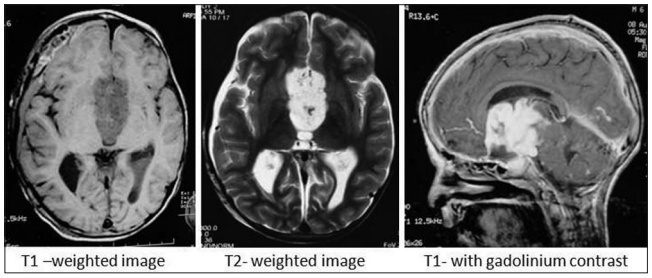

Figure 1: Magnetic resonance image of brain showing a sellar and suprasellar mass lesion, which was hypointense on T1-weighted image, hyperintense on T2-weighted image and intensely enhancing with gadolinium contrast

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and Rosenthal fibers [Figure 2]. As per the preoperative findings and histopathology, it was diagnosed as a case of chiasmatic pilocytic astrocytoma. The child received 54Gray/30 fraction radiation to the postoperative residual with three-dimensional conformal technique and kept on follow up. After 1 year, the patient presented with headache and vomiting not relieved by oral dexamethasone 4mg twice a daydosage. MRI of brain and spinal cord showed diffuse multiple leptomeningeal infiltration involving the posterior fossa [Figure 3] and cord. He was planned for palliative chemotherapy with Carboplatin (200mg/m^2 iv day 1-3) and Etoposide (100mg/m^2 iv day 1-3), every 3 weekly. Craniospinal irradiation was not given to avoid the risks of re-irradiation within 1 year, in this child. After receiving threecycles of chemotherapy, repeatedbrain MRI was done, which showed clear disease progression. The patient died 2 weeks after the third cycle chemotherapy.

DISCUSSION

Pilocytic astrocytoma (PA) is a benign variety of gliomas with an excellent prognosis. Most commonly, it arises from cerebellum, and less commonly from optic nerve, hypothalamus, and cerebral hemispheres. Although, it usually has a benign course, but rarely it can show a malignant behavior. Different patterns of presentation or recurrence, such as malignant transformation, multicentric disease or metastatic spread, may be found. Leptomeningeal dissemination (LMD) of PA, either at the time of initial presentation or at recurrence is a rare event, and till date only a few cases have been reported in the medical literature.[1,2]

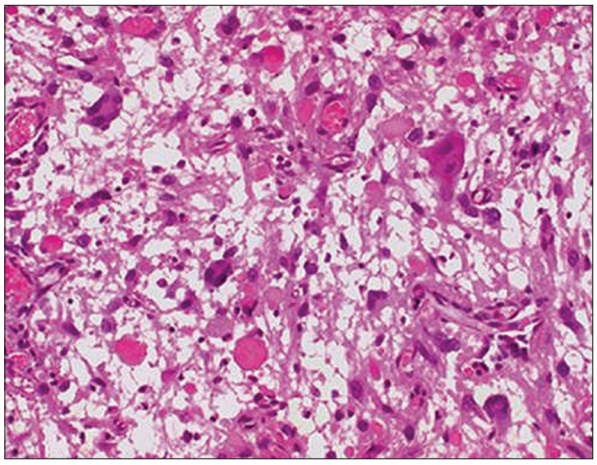

Figure 2: Photomicrograph of the histopathology of the operated specimen showing fusiform cells with wavy fibrillary processes and Rosenthal fibers (H and E, X20)

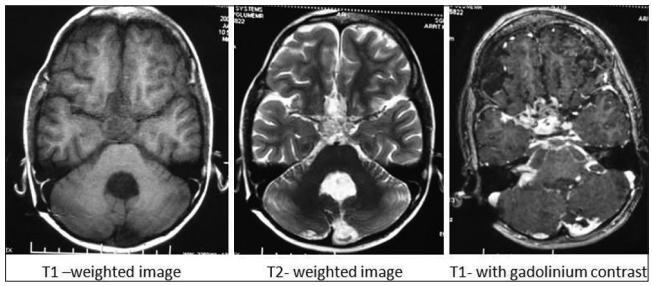

Figure 3: Magnetic resonance image of brain showing diffuse multiple leptomeningeal infiltration in the posterior fossa

It is reported that children with optic pathway gliomas should be considered for this possibility, particularly after the onset of new atypical neurologic symptoms.[3]

The incidence of LMD of PA is variable in different series. Mamelak, analyzing his series with 90 cases of PA, found an incidence of 12%.[1] The incidence of LMD in low-grade gliomas is around 3.7-5.3%.[2,4,5]

The outcome of patients with PA and LMD is not well known. Nevertheless, it is likely to be not as good as that of patients with localized recurrence or totally resected primary disease. However, it is not as bad as the LMD in high-grade gliomas. In a retrospective review to determine the patient, tumor, and treatment characteristics of patients withPA and LMD, Mazloom *et al*. found that the median survival for PA patients with LMD was 65 months. The 1-, 2-, and 5-year overall survival (OS) rate after the diagnosis of LMD was 81.1%, 75.7%, and 55.5%. Age, gender, primary site location, timing of LMD presentation (synchronous vsmetachronous), and LMD location did not significantly influence OS or progression free survival (PFS).[6]

The optimal treatment strategy for LMD of PA is still unknown. Craniospinal irradiation, palliative chemotherapy along with subtotal resection of the primary lesion, are the treatment options. Although, the use of chemotherapy for the treatment of LMD of PA, is better supported by some literature,[7-10] but in their retrospective review Mazloom *et al*. have not found any statistically significant difference in OS or PFS between the chemotherapy and radiotherapy groups.[6]

Clearly further controlled studies are needed to define the risks factors, optimal treatment of primary, and disseminated disease and prognosis of this rare pathological entity.

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