Case Report

Metastatic medulloblastoma at diagnosis

ABSTRACT

Medulloblastoma is an aggressive tumor of the brain. It is the most common and the most malignant embryonal tumor of the pediatric central nervous system and a rare tumor of adults. We are reporting a rare presentation of adult classic subtype of medulloblastoma which was central in location with metastases in the suprasellar region at the time of diagnosis.

KEY WORDS: Adult medulloblastoma, histopathology, MR imaging, suprasellar metastases, treatment

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INTRODUCTION

Medulloblastoma was first described by Bailey and Cushing in the year 1925.[1] It is mainly a pediatric tumor but found rare in adults.[2]

It is uncommon for adult medulloblastoma to present with metastatic disease at the time of diagnosis.[3]

We report a case of adult medulloblastoma with large suprasellar metastases diagnosed on endoscopic biopsy of the suprasellar mass during endoscopic third ventriculostomy (ETV).

CASE REPORT

A 27-year-old male presented with symptoms of raised intracranial tension and bitemporal visual field disturbance for the past 2 months.

MRI showed a heterogeneously space occupying lesion involving vermis and adjoining part of cerebellum and medulla. Another lesion of similar intensity involving suprasellar region was seen. MR spectroscopy (MRS) of posterior fossa mass showed increased choline peak, decreased *N*-acetyl acetate (NAA) peak with increase choline to creatinine ratio (Ch/Cr) suggestive of neoplastic etiology [Figure 1].

The patient underwent ETV procedure along with an endoscopic biopsy of the suprasellar mass, which on histopathology showed a classic subtype of medulloblastoma. Tumor cells were positive for neuron specific enolase, synaptophysin, and negative for cytokeratin [Figure 2]. High expression of Ki67 was noticed. CSF was negative for malignant cells.

Following biopsy the patient received craniospinal radiation 36 Gy in 18 fractions followed by booster dose of 24 Gy in 12 fractions to suprasellar and posterior fossa, and further followed by cisplatin, CCNU, and vincristine-based chemotherapy for residual disease [Figure 3].

DISCUSSION

Our patient had a rare presentation of adult classic subtype of medulloblastoma which was central in location with metastases to the suprasellar region at the time of diagnosis disseminated probably through cerebrospinal fluid, whereas the presentation of adult medulloblastoma is characterized by localized lateral tumor location,

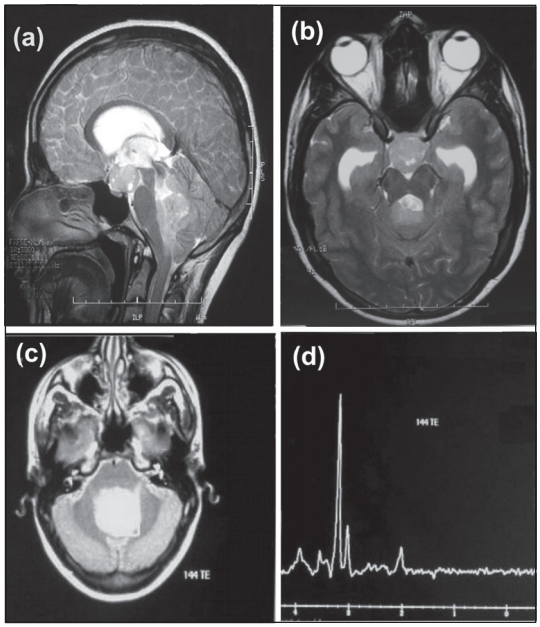

Figure 1: T2 sagittal and axial (a and b) showing posterior fossa mass and suprasellar mass. (c and d) MR spectroscopy of posterior fossa mass suggestive of malignant lesion

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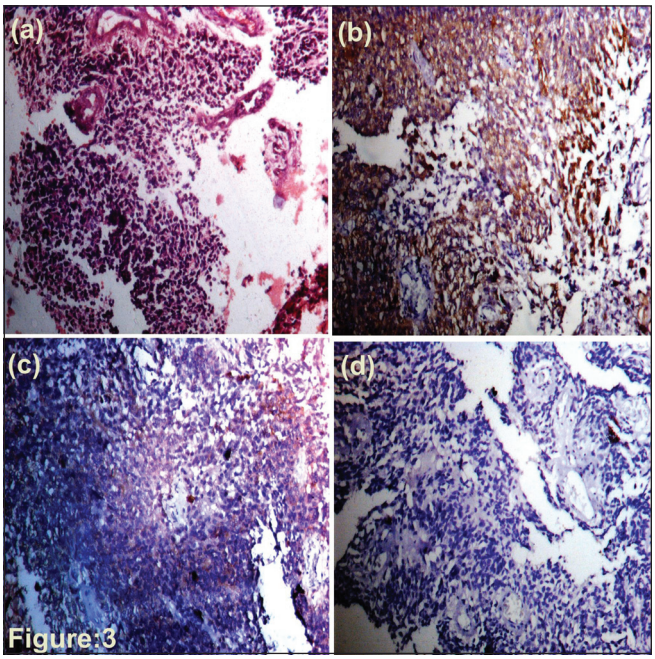

Figure 2: Microphotograph of endoscopic biopsy showing (a) small round cell tumor with cellular molding and rosettes in high power view (H&E × 125× digital magnification), (b) tumor cells are positive for neuronspecific enolase (c) synaptophysin, and (d) negative for cytokeratin (DAB × 525× digital magnification)

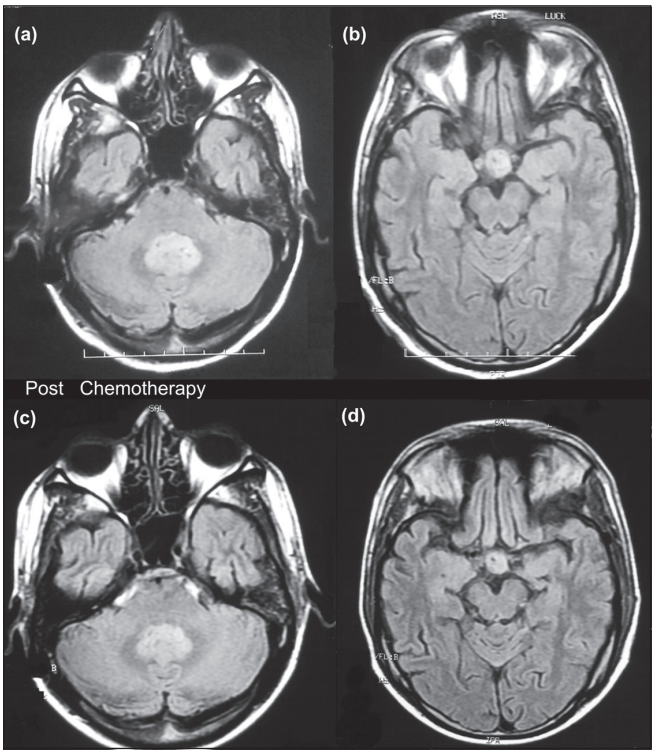

Figure 3: T1 axial (a–d) showing postradiotherapy and postchemotherapy residual disease in the posterior fossa and suprasellar region

desmoplastic or classic histology variant, and recurrence of disease after longer time periods.[3]

Traditionally, classical and desmoplastic variants which are more common in adults were considered favorable histology and degree of anaplasia was considered unfavorable.[4] However, recent evidence suggests that histopathology grading based on increasing anaplasia predicts clinical behavior of pediatric medulloblastomas and not in adult medulloblastoma, as the histopathologic spectrum of medulloblastoma in adults is different from that in children,[5] as seen in our patient the classic subtype of medulloblastoma had an aggressive presentation .

The criteria which determine the prognosis in adults include metastasis through CSF to the different regions of brain, nonradical surgery, and a postoperative performance status (PS) >2.

Recent advances in molecular biology and cytogenetics have contributed to the updated classification of medulloblastomas and refined our understanding of the new subtypes. Recognition of these variants becomes important because of their distinct biologic behavior and treatment strategies that may be different.

Gene expression analyses have revealed that expression of several marker genes, such as MYC, ERBB2, and TRKC are associated with poor prognosis.[6]

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