# Cerebral Medulloepithelioma: A Rare Aggressive Brain Tumor in Child – A Case Report

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Indian J Neurosurg 2016;5:55–58.

**Abstract**

Medulloepithelioma of the central nervous system (CNS) is a rare primitive neuroectodermal tumor (PNET) usually occurring in early childhood. It is a highly aggressive tumor with poor outcomes despite aggressive treatment. The ideal treatment appears to be gross total surgical resection, followed by radiation therapy. We report a case of a cerebral medulloepithelioma in a child with an aggressive tumor growth despite the gross total resection, who is surviving 1 year after diagnosis.

**Keywords**

- cerebral medulloepithelioma
- tumor
- brain
- child

## Introduction

Cerebral medulloepithelioma is a rare, highly malignant embryonal brain tumor, commonly seen in pediatric age group and carrying a very poor prognosis with a reported median survival of only 5 months with a propensity for progression, recurrence, and dissemination, despite therapy.[1] We report a case of cerebral medulloepithelioma in a 9-year-old child who had an aggressive tumor growth after first surgery, and after second surgery, additionally received postoperative radiation therapy, and is now following up, 1 year after diagnosis, without any clinical evidence of tumor recurrence.

## Case Report

A 9-year-old boy, presented with history of progressively worsening headache and weakness of left half of body for the last 5 days. The physical examination revealed the child to be conscious but disoriented and having left hemiparesis with papilledema bilaterally. There was a positive history of contact with tuberculosis in the family. The routine preoperative investigations were within normal limits, with the exception of erythrocyte sedimentation rate (ESR), which was 46 mm. Computed tomographic (CT) scan and contrast magnetic resonance imaging (MRI) of the brain were suggestive of a large well-defined heterogeneously enhancing mass lesion in the right frontal and parietal region, with gross perilesional edema and severe mass effect (►**Figs. 1** and **2 A–D**). Craniotomy was done and gross total resection of the tumor mass was achieved. Peroperative, the tumor was well encapsulated with a clear plane with the normal brain parenchyma. The tumor was grayish yellow, caseous, firm in consistency, with no involvement of the overlaying dura. Per-operative impression was of a tuberculoma. Postoperatively, the child recovered, hemiparesis improved, and he was discharged on post-op day 6, on five-drug antitubercular therapy.

The histopathology was suggestive of a highly cellular tumor, composed of nests, tubules, and trabecular arrangement of malignant cells, lined by pseudostratified epithelia, resembling primitive neural tube (►**Figs. 3A and B**). Immunohistochemistry was positive for pan-cytokeratin, nestin, and vimentin (►**Fig. 3C**), suggestive of medulloepithelioma.

The patient, however, was lost to follow-up; hence he could not receive adjuvant radiotherapy.

The child was again brought to casualty after 3 months of the initial surgery in a state of altered sensorium. Plain CT

**received**
July 28, 2015
**accepted**
November 23, 2015
**published online**
February 24, 2016

DOI http://dx.doi.org/10.1055/s-0036-1572375.
ISSN 2277-954X.

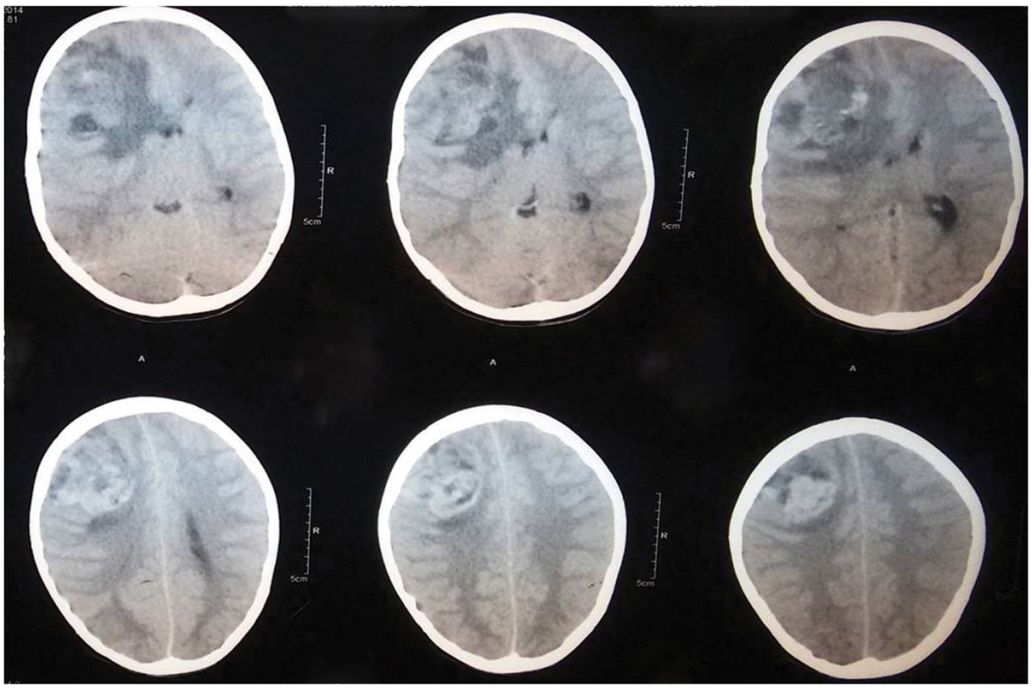

**Fig. 1** Plain CT scan at admission.

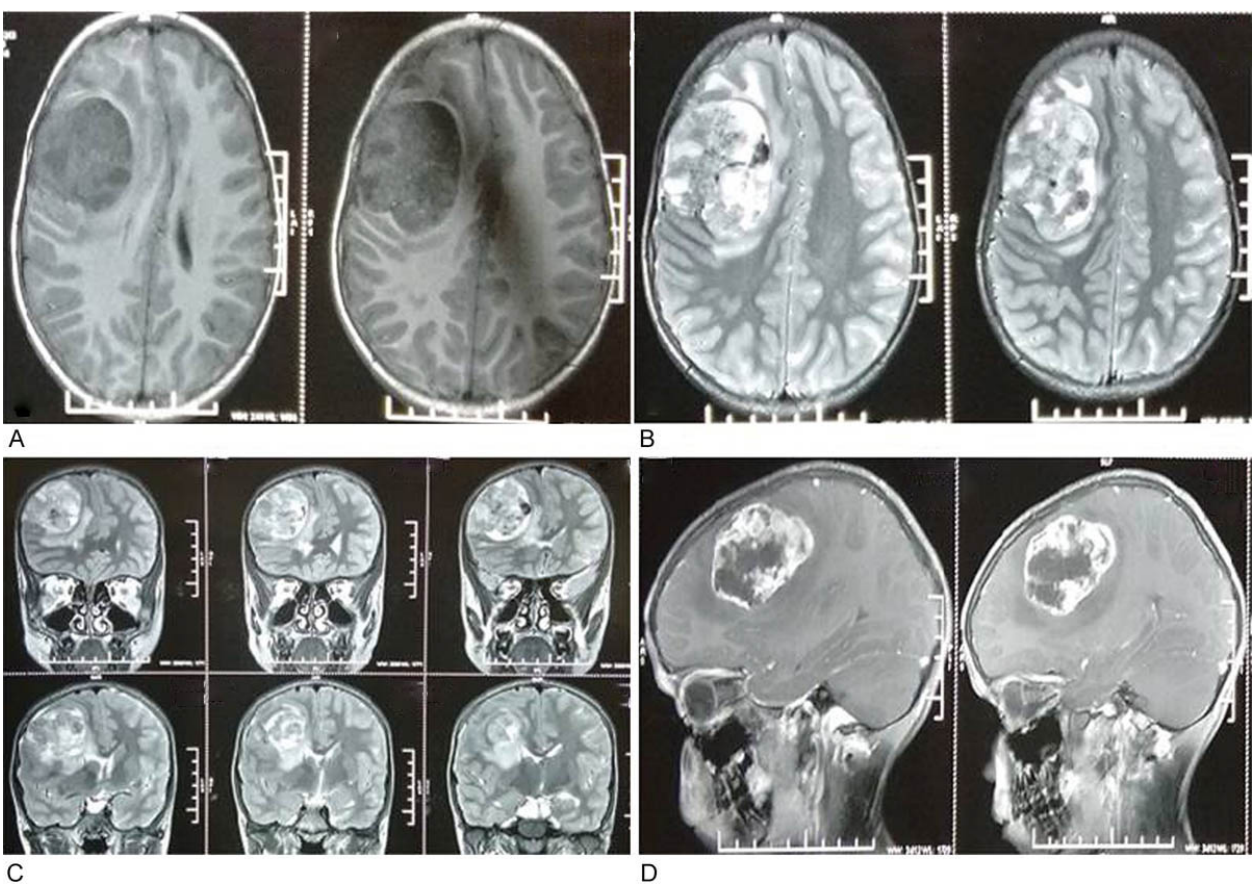


**Fig. 2** MRI (**A**) T1 axial. (**B**) T2 axial. (**C**) T2 coronal. (**D**) Contrast.

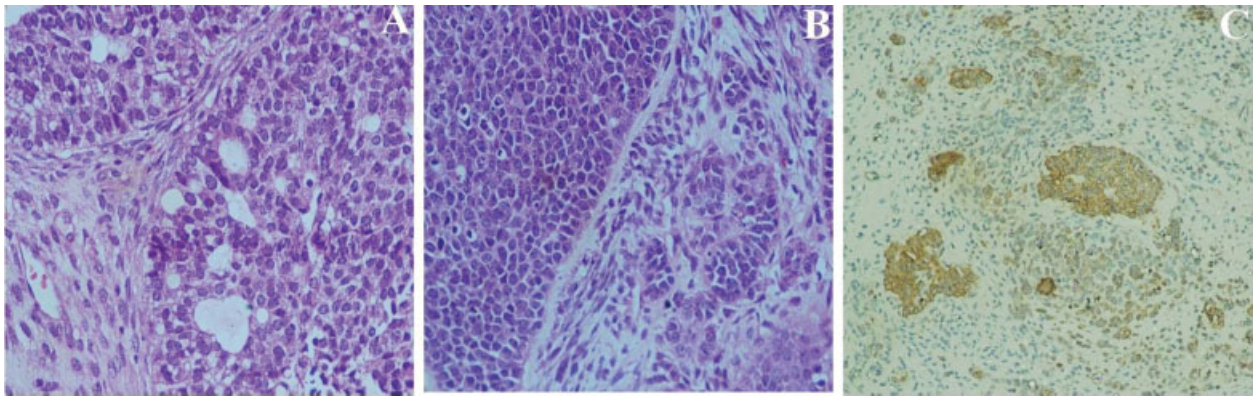


**Fig. 3** (**A**) Tubules appreciated in tumor islands. (**B**) Solid tumor islands with adjacent cellular stroma. (**C**) Immunohistochemistry: Pan-cytokeratin–positive cells.

scan and contrast MRI done at that time was suggestive of an extensive tumor recurrence along with severe mass effect (►**Fig. 4**). He was reoperated and gross total resection achieved. MRI of the spine and lumbar cerebrospinal fluid (CSF) examination, done to rule out any dissemination of tumor, were unremarkable.

The histopathology report was once again suggestive of medulloepithelioma. He recovered well and received postoperative radiation therapy. The child attended the outpatient clinic 1 year after the diagnosis and was clinically not having any neurologic deficits and was attending school.

## Discussion

The cerebral medulloepitheliomas were first described by Bailey and Cushing in 1926, as a separate entity of tumors.[2] They are categorized as rare, highly malignant primitive neuroectodermal tumors (PNETs) (World Health Organization [WHO] grade IV). So far, only nearly 45 cases of cerebral medulloepitheliomas have been reported in the literature, with four cases reported from India.[3–5] They usually manifest in the first decade of life[6,7] and are occasionally seen in older patients,[8] with an equal incidence in males and females.[1] The

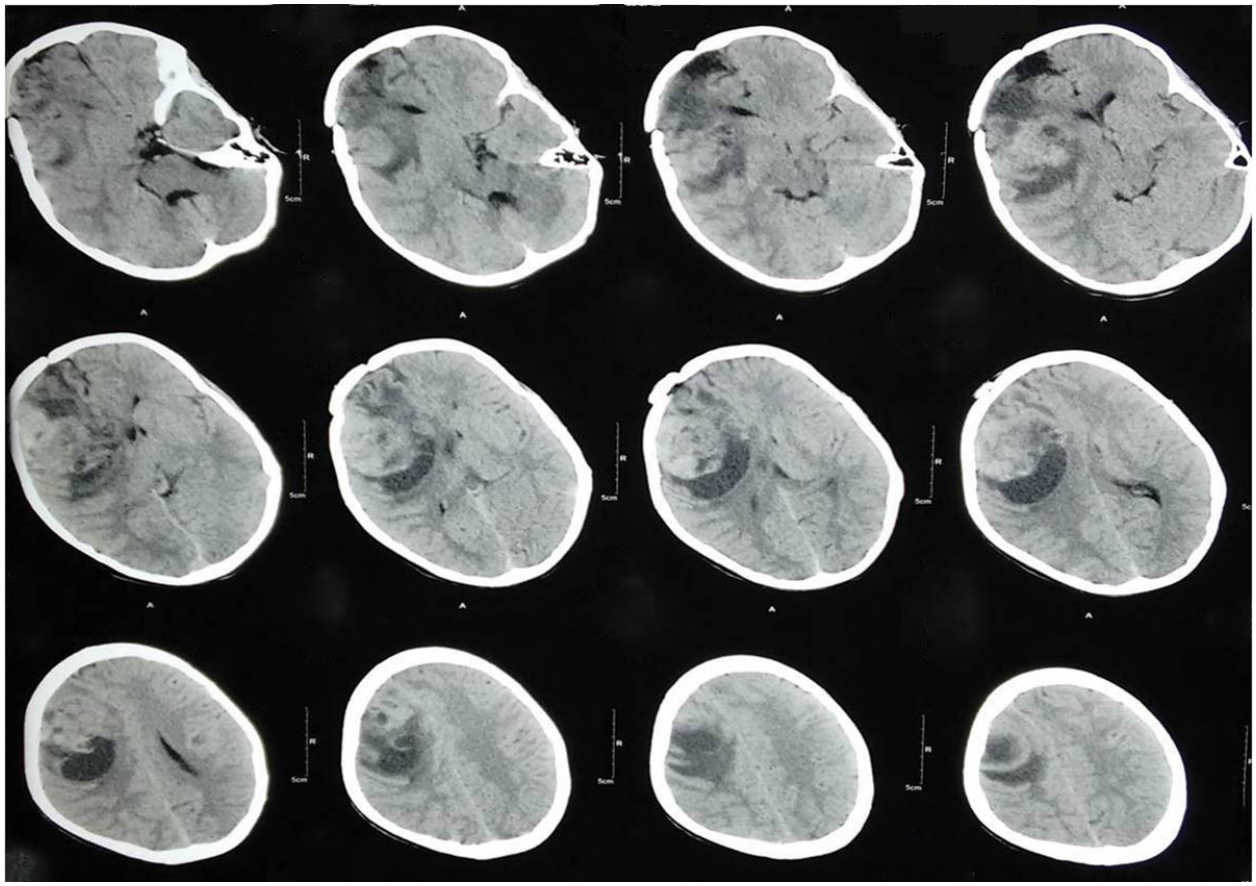

**Fig. 4** Plain CT scan showing recurrence of tumor.

natural history is of an aggressive type and the outcome in most cases is fatal despite treatment.

On CT scan, these tumors appear well circumscribed, isodense to hypodense to brain parenchyma with heterogeneous contrast enhancement, and on MRI, they are hypointense on $T_1$ and hyperintense on $T_2$ with no contrast enhancement.[1,9] Our patient, however, showed a heterogeneous contrast enhancement on MRI, which is also reported by some other authors in their reports.[4,10]

On histopathology, the most distinguishing feature is the arrangement of pseudostratified columnar cells around a lumen, resting on a continuous basement membrane, mimicking a primitive neural tube. Immunohistochemically, these cells are positive for nestin and vimentin and positivity for cytokeratin indicates ependymal differentiation.

The ideal treatment of cerebral medulloepitheliomas is a gross total resection of the tumor followed by radiation therapy of the craniospinal axis with a local boost to cranium.[1] The role of radiation therapy is clearly well established, with most of cases reported having a fairly long survival after postoperative radiotherapy.[1,8] In our case also, the tumor initially recurred within 3 months without adjuvant radiotherapy; however, the child is tumor free after 1 year of second surgery and postoperative radiotherapy. Few authors have advocated the use of chemotherapy, but its efficacy in the treatment of medulloepithelioma remains to be determined.[10] Only one case has been reported in the literature who survived more than 7 years after surgery, without any adjuvant therapy.[11]

## Conclusion

Medulloepitheliomas must be considered as a differential diagnosis, whenever a PNET is suspected, particularly in children in their first decade. The approach toward management should be rather an aggressive one, with screening of craniospinal axis for dissemination of tumor and a gross total surgical resection of the tumor, followed by radiation therapy. The establishment of the utility and efficacy of chemotherapy in the management warrants further study.

Conflict of Interest
The authors report no conflict of interest concerning the materials or methods used in this report or the findings specified in this paper.

Financial Disclosure
None.

Permission Requests
None.

Contributors' Statement
The study was first conceived by Dr. Rohit Bansil, Dr. S. Bhaskar, and Dr. Aman Kindra. The pathological diagnostic help was provided by Dr. P. Agarwal. The manuscript was prepared by Dr. Rohit Bansil, Dr. Naseem Mansoori, and Dr. Sumit Sinha.

Acknowledgments
We thank Mr. Shanu and Mr. Akshay for technical assistance.

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