Case Report

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Intracerebral Medulloepithelioma with Divergent Differentiation: A Case Report and Review of Literature

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Abstract

Cerebral Medulloepithelioma is an extremely uncommon tumor of childhood and so far 39 cases were reported in literature. We report a new case of cerebral intraventricular medulloepithelioma in an 11 year old female child which showed divergent line differentiation. We analyzed the histopathological and immunohistochemical features and differential diagnosis of this rare tumor. The prognosis and management issues in view of the available literature were discussed by the author.

Key words: Medulloepithelioma; Differentiation; Immunohistochemistry; Surgery; Prognosis.

Introduction

Medulloepithelioma is a rare, malignant embryonal brain tumour histologically characterized by papillary, tubular or trabecular arrangements of neoplastic neuroepithelium [1]. They mostly occur in young children between 6 months and 5 years. They are typically described in intraocular location and have relatively better prognosis. In contrast intracerebral tumours have high rate of recurrence due to their rapid subarachnoid spread and radioresistence of tumour cells. We report a cerebral intraventricular medulloepithelioma in an 11 year old female child.

Case Summary

An eleven year old girl presented to local ophthalmologist with presenting complaints of bi-temporal headache, blurring of vision, diplopia and vomiting. On examination found to have bilateral papilledema, on further evaluation, magnetic resonance images (MRI) showed lesion in right trigone of lateral ventricle, which was hypointense on T_1 and mixed intense on T_2 (solid and cystic), heterogeneously enhancing on contrast. **Operation**: Right parietal craniotomy and gross total excision of the tumor was done by superior parietal lobule approach. Grossly the tumor was greywhite to grey brown measuring 2x2x1cm with well defined margins from surrounding brain. Clear attachment with choroids plexus could not be defined. Gross total excision was done and the same could be confirmed on postoperative computed tomography (CT) scans. Patient recovered well without any neurological deficits after surgery. She was advised chemotherapy and radiotherapy but did not take further adjuvant treatment.

After 6 months the patient again presented with features of raised intracranial pressure with MRI showing recurrence of lesion which is hypointense on T_1 hyperintense on T_2 with partial suppression on flair involving the body of right lateral ventricle and also parieto occipital lobe with moderate perilesional edema and midline shift (Figure 1). Patient was taken up for surgery and gross total excision was done again by superior parietal lobule approach. Gross total excision was achieved again and patient was advised adjuvant treatment. Patient did not receive the therapy because of family issues and came back after 3 months with a large recurrence. Patient was again advised for surgery followed by chemoradiotherapy for which the parents refused and the patient expired after 10 months since the time of initial presentation.

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Pathological findings: Histopathology revealed a papillary lesion lined by psuedostratified epithelium showing frequent mitosis. In addition, there were areas showing divergent line differentiation into cartilage,skeletal muscle and squamous epithelium (Fig.2). Some blastemal cells were also seen. Immunostaining with pancytokeratin, EMA and vimentin was positive in papillary areas whereas desmin showed intense cytoplasmic expression in rhabdomyoblasts (Fig. 3). GFAP was negative and Ki 67 labelling index was high (>30%) Theblastemal cells were reactive for synaptophysin. The morphology and immunohistochemistry was consistent with the diagnosis of teratoidmedulloepithelioma. The tumor expressed INI-1 protein ruling out a possibility of atypical teratoidrhabdoid tumor.

Figure 1: Magnetic resonance images (MRI) showing hetrogenouslyhypointense lesion lesion on T_1 (A, axial cuts), hyperintense on T_2 (B axial cuts,C,Dcornal cuts) with partial suppression on flair involving the body of right lateral ventricle and also parieto occipital lobe with moderate perilesional edema and midline shift.

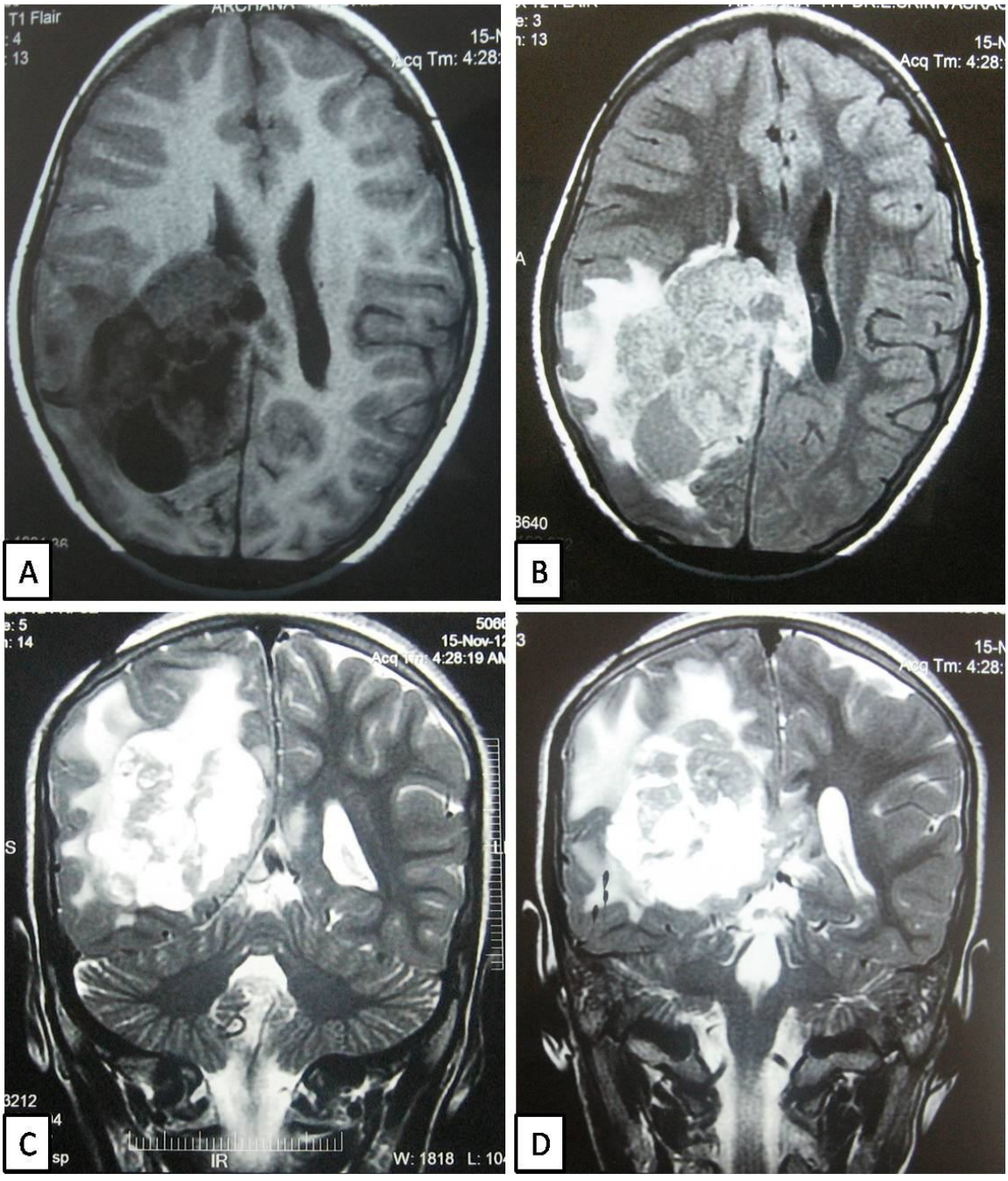


Figure 2: Photomicrograph of the tumor showing divergent differentiation. (A) (Haematoxylin and eosin [H&E] stain, original magnification x 20) papillary structures lined by pseudostratified epithelium. (B) (H&E stain, original magnification x 400) immature neuroepitheliumresting on basement membrane with mitosis located towards luminal surface. (C) (H&E stain, original magnification x 100) rhabdoid cells. (D) (H&E stain, original magnification x 100) lobule of primitive cartilage. (E) (H&E stain, original magnification x 100) sheet of undifferentiated blastemal cells. (F) (H&E stain, original magnification x 100) squamous islands.

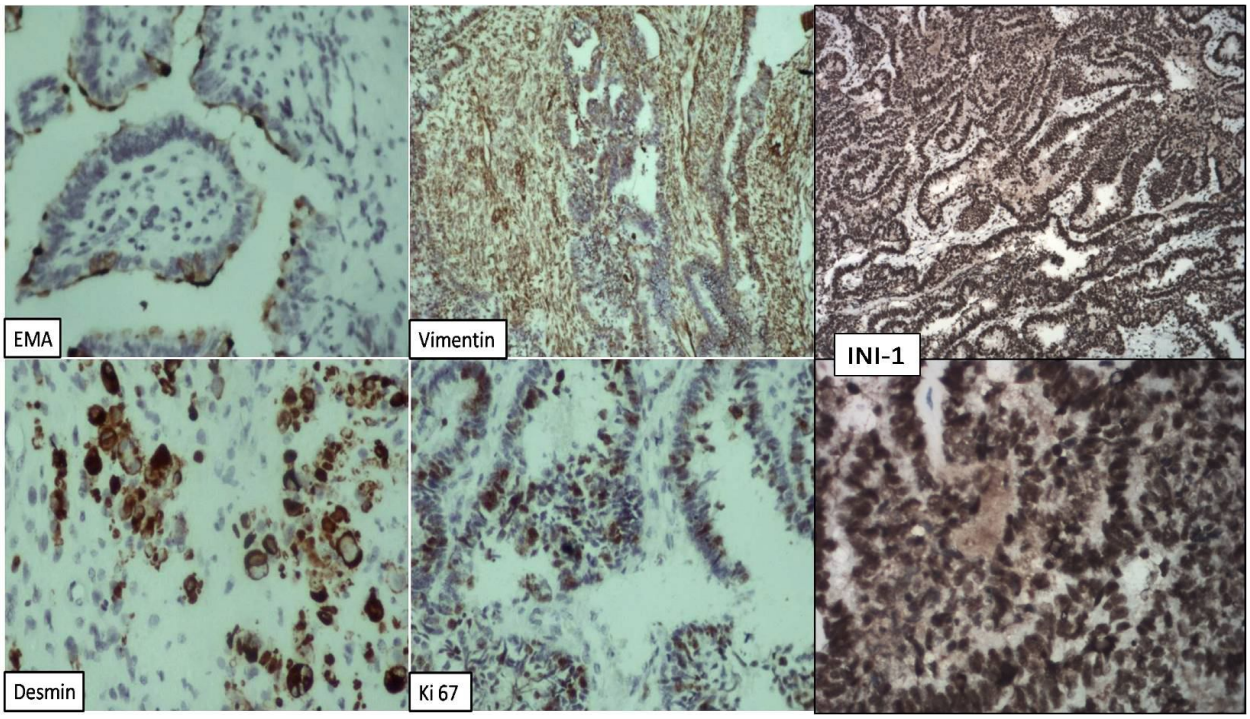
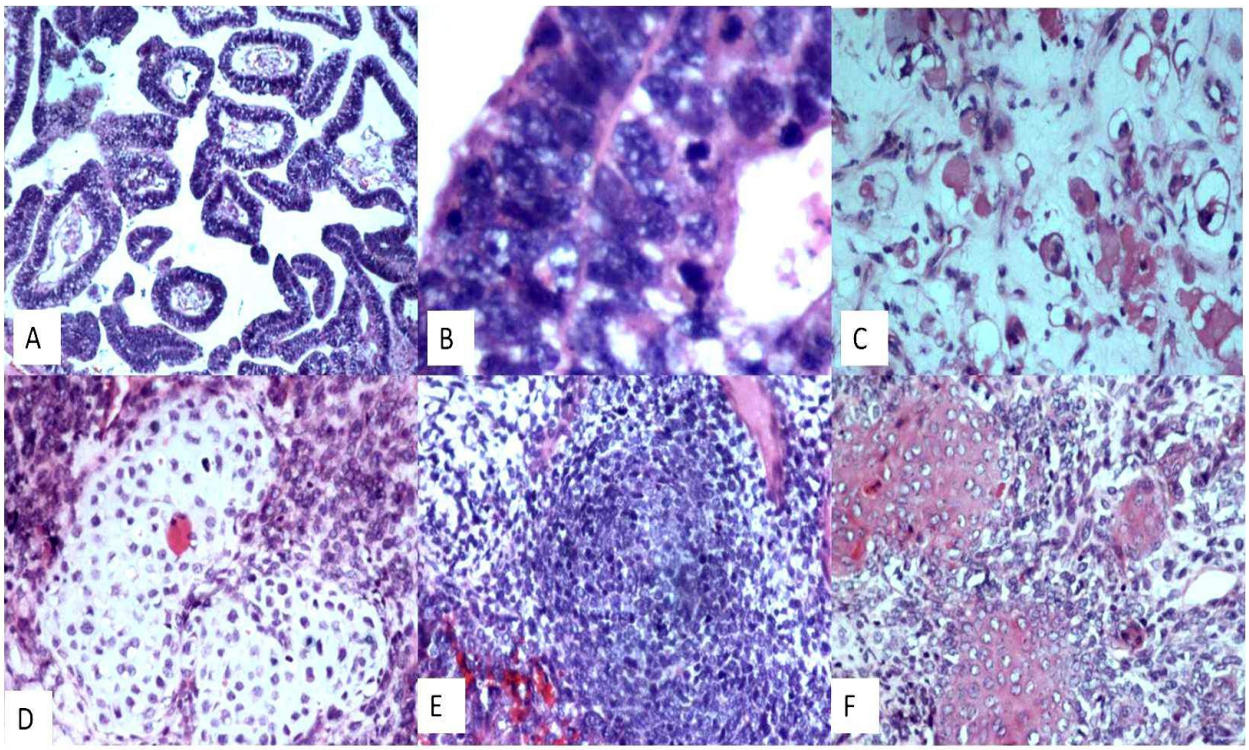


Figure 3: Photomicrograph of immunohistochemistry of the tumour. (A) (indirect immunoperoxidase, original magnification x 100) the tumor showing luminal cytoplasmic positivity with EMA (epithelial membrane antigen). (B) (indirect immunoperoxidase, original magnification x 20) neoplastic cells exhibiting vimentin cytoplasmic positivity. (C) (indirect immunoperoxidase, original magnification x 100) D) (indirect immunoperoxidase, original magnification x 100) rhabdomyoblasts showing intense cytoplasmic positivity with desmin.



Discussion

Cerebral Medulloepithelioma is an extremely uncommon tumor of childhood. It was first described by Bailey and Cushing in 1926[1]. It is a WHO grade IV tumor and is included under CNS PNETs[2]. They mostly occur in young children between 6 months and 5 years, however rare cases occurring beyond the first decade have also been reported [2,3,4,5,6]. Intracranial medulloepitheliomas have been described in hemispheres, brain stem and cerebellum. Periventricular location is most common in intracerebraltumours though Intraven tricularlocation is also described[7,8].

The postulated cell of origin of the medulloepithelioma is primitive neuroectodermal stem cell because of its ability to mimic the primitive neural tube and to differentiate along multiple lineages. Histopathology includes papillary tubular and trabecular arrangements [1]. These tumors show distinct neuroepithelium that is based on an outer basement membrane and characteristic architectural features that include long linear tubular, canalicular and papillary patterns. These are unique features and are not described in other central PNETs.

Medulloepitheliomas can show heteroplastic elements, like hyaline cartilage, rhabdomyoblasts, or glial tissue. Those with heteroplastic tissue are designated *teratoidmedulloepitheliomas*. The heterologous differentiation in the form of cartilage and rhabdomyoblasts was seen in this case. Nonteratoid and teratoidmedulloepitheliomas are benign or malignant [9]. However the present case showed features of malignancy in the form of brisk mitosis and necrosis.

Varied histological patterns of medulloepithelioma can raise a broad differential diagnosis including medulloblastoma, ependymoblastoma, neuroblastoma, choroid plexus carcinoma and immature teratoma. However choroids plexus carcinomas are not known to show divergent differentiation which was very frequent in our case. Immature teratoma can be excluded on the basis of absence of tissue of foetal appearance from other germ layers [10]. Also immature teratomas usually show some mature tissues as well which was not seen in the present case.

The immunehistochemical profile of these tumors includes reactivity for nestin and vimentin particularly in the neuroepithelial component. Our case was also immunoreactive for vimentin which is the first intermediate filament protein demonstrated in early developmental stage of neural tube [11]. Immunoreactivity reflects the pattern and degree of differentiation of the tumor. Ki-67 labelling is variable within these tumors with areas of low (1–3%) labeling adjacent to those with extremely high labelling (>50%) [1]. Our case had a high ki 67 labelling (30%). This high proliferative rate might be responsible for early recurrence and rapid increase in size of the tumor resulting in short survival.

Intracerebral medulloepitheliomas have dismal prognosis as compared to intraocular tumors. Till date only 39 cases have been reported and with different treatment protocols used in each case and the wide range of survival, the benefit of each treatment is not known [12,13,14]. But a look at the available cases where patient came in good performance scores and gross total excision was achieved the survival was beyond 6 months. The present case also recurrence occurred after 6 months of gross total resection and gross total resection of the recurrent lesion also gave another 3 months of symptom free survival. The recent case reports and small case series published where aggressive chemoradiotherapy was used the survival ranged from 2 months to 60 months [14,15,16,17]. In the case report by Moftakar et al, the patient survived more than 5 years. Previously only 2 more cases were available in the literature where the patient survived more than five years after surgery and chemoradiotherpay [18]. Our cases and the other cases available in the literature suggest that good performance score at the presentation with gross total resection seem to have a favorable outcome. The survival advantage of chemoradiotherpy still needs to be ascertained. Adding to that is the adverse effect of these therapies in the growing children thereby affecting the quality of life during the short term survival offered by surgery. Most medulloepithelioma patients die within a year of diagnosis, usually with cerebrospinal fluid dissemination but rarely with systemic metastases [10]. Age, gender or differentiation of the tumor does not seem to affect the prognosis [14]. The factors affecting prognosis include location of tumor, and extent of tumor resection [11,14,19,15,16,17,18]. Supratentorial location and exclusive intraventricular location of the tumor and negative cerebrospinal fluid have a better prognosis [14,19].

Conclusions

In conclusion, intracranial medullopeithelioma is a rare aggressive tumor in the age group of 6 months to 5 years. Gross total resection with good performance score at presentation seem to have

favorable outcome. The role of highly toxic chemoradiotheraphy with all its adverse effects in the growing age is doubtful.

Disclosure

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

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