

Supra-Tentorial Cortical Ependymoma in an Adult: Case Report and Review of the Literature

Shaymaa Al Umran¹, Mahmoud Taha¹, Marwa Abdulkader²

¹Department of Neurosurgery, King Fahad Specialist Hospital, Dammam, KSA

²Department of Histopathology, King Fahad Specialist Hospital, Dammam, KSA

Email: dr.al-umran@hotmail.com

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Abstract

Supratentorial cortical ependymoma is a rare primary brain tumor in adults. Only 50 cases are reported in the literature including pediatric and adult patients. We report a 30-year-old male with sensory-type seizures who was found to have a right posterior frontal cortical lesion with large cystic component. The lesion was resected completely and histopathology and immunohistochemistry examinations confirmed anaplastic ependymoma WHO grade III. The patient received radiotherapy treatment post-surgery, and he was tumor free one year after treatment.

Keywords

Supratentorial Ependymoma, Sensory Seizures, Cortical Ependymoma

1. Introduction

Ependymomas are glial tumors derived from ependymal cells lining the ventricles and the central canal [1] [2]. They are most frequently seen in pediatrics and young adults. Ependymomas constitute 1.2% to 7.8% of all intracranial neoplasms [1]. According to World Health Organization (WHO) 2016 of CNS tumors, ependymomas are graded as grade II (low grade) and grade III (anaplastic or high grade) ependymomas [1]. They are usually located in the infra-tentorial compartment, and only one third of ependymomas are supra-tentorial [3].

Supra-tentorial cortical ependymomas (CE) are very rare type of ependymoma with a few reported cases in the literatures [1] [2]. They occur in the superficial cortex and have no connection with the ventricular lining [1] [4].

In this paper, we are reporting a rare tumor involving unexpected location of

ependymoma with limited reported similar cases between pediatrics and adults.

2. Case Report

A 30-year-old male, right handed, not known to have any chronic medical illness was referred to our hospital with five months history of left side episodes of numbness and heaviness sensation involving upper and lower limb. Each episode lasts for about 5 minutes. He had a total of 4 or 5 episodes. One month prior to his admission he started to complain of left sided upper and lower limbs weakness. On admission the patient was fully awake and orientated to time and place. His neurological examination showed mild increase in the left upper and lower limbs tone in comparison to the right side. Power was slightly reduced in the left side of the body.

MRI brain with contrast and fMRI was done and showed a right posterior frontal cystic mass lesion with enhancing mural nodule. The overall measurements were $4.5 \times 4 \times 4.8$ cm (**Figure 1(a)** and **Figure 1(b)**).

EEG was done and showed no epileptic activities.

Treatment

Patient underwent a right posterior frontal craniotomy under image guided. The cystic part of the tumor was drained first, and then the solid nodule was removed totally. The cystic part of the tumor has a gliotic wall except at the solid nodule which was grayish and slightly vascular and has a well-define capsule.

The initial frozen section came positive for low grade glioma.

Postoperative Course

Post operatively; the patient was doing well with no post-operative complications. He was watched post-operatively in ICU for 24 hours, then shifted to a regular ward. Post-operative MRI brain showed a gross total resection (**Figure 1(c)** and **Figure 1(d)**). His weakness improved within days from the surgery. The final pathology reported as anaplastic ependymoma (WHO grade III) (**Figures 2(a)-(d)**). The patient received focal radiotherapy at the operative site 6 weeks after surgery. We found no recurrence of the tumor after one year of follow up.

3. Discussion

Only 50 cases of supratentorial cortical ependymomas were reported in the literature, 17 cases were children (34%) and 33 were adults (64%) [1].

Supratentorial extra-ventricular Ependymoma more often found in adults than children, while posterior fossa extra-ventricular ependymoma found more in children [3].

The classic clinical presentation of CE as most superficial neocortical lesions are seizures and focal neurological deficits, and rarely may present with signs of raised intracranial pressure [1] [2] [4].

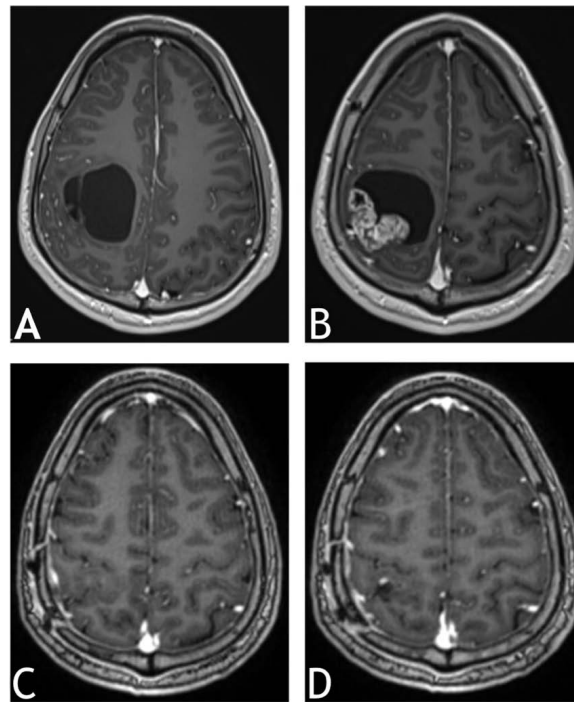


Figure 1. (a) Pre-operative axial T1 brain MRI with contrast showing intra-axial non enhancing cystic mass lesion at right anterior parietal area; (b) Pre-operative axial T1 brain MRI with contrast showing intra-axial complex mass lesion at right anterior parietal area with heterogenous enhancing solid component which is measuring $3.6 \times 1.3 \times 2.1$ cm in ML \times AP \times CC dimensions; (c) and (d) Post-operative axial T1 MRI brain with contrast showing a gross total resection of the lesion with tiny enhancing focus at the surgical bed.

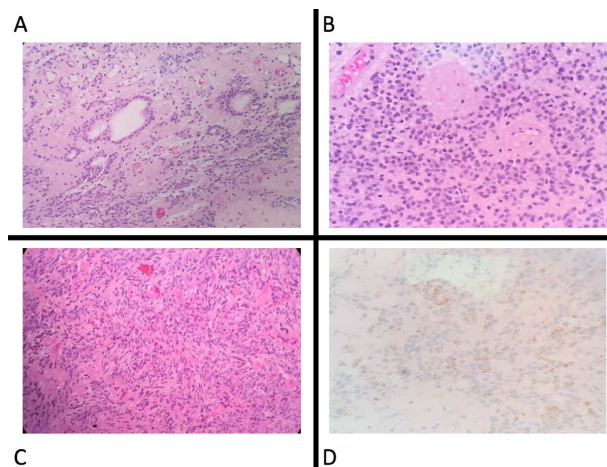


Figure 2. Pathology Specimens. (a) Demonstrating true ependymal rosette consisting of tumor cells arranged around well-defined lumens forming gland-like structures; (b) Demonstrating ependymal cells with spindle cell morphology; (c) Demonstrating perivascular pseudorosettes; (d) EMA dot pattern of immunoreactivity.

The pathogenesis of the extra-ventricular ependymomas remains uncertain. Several theories were suggested including: 1-cortical ependymomas may arise from embryonic remnants of ependymal tissues in the developing cerebral he-

mispheres. 2-Cortical ependymoma originated from cell type other than terminally differentiated ependyma [5].

Liu *et al.* [3] reviewed 45 cases of CEs and he found that almost half of them (24 cases) were grade II (low grade) and the rest were grade III (high grade). The average age was 20 years for the low grade CEs (range from 2 to 63 years old), and 33 years for high grade CEs (range from 1 to 70 years old). Male-to-female ratio was 1:1.1 for both groups [3]. Seventy-nine percent of the patients with low grade CEs presented with seizures while only 45% of the patients with high grade CEs presented with seizure. For both low grade and high grade CEs, the most common locations were frontal and parietal lobes. On MRI, there was no obvious surrounding edema for low grade CEs while there was marked brain edema in almost 60% of high grade CEs. The majority of tumors exhibit solid or mixed solid/cystic appearance. The solid component showed heterogeneous enhancement after administration of the contrast agent [3].

Radiologically, CE tends to be more commonly located in frontal and parietal areas.

larger in size at the time of diagnosis with a 94% of supratentorial ependymomas manifest with a size larger than 4 cm and often contain a mixed solid and cystic component as described by Roncaroli *et al.* [1] [2]. A plain CT of CE ranges from iso to hypo-dens. It has been found that a low grade CE rarely associated with brain edema. The radiological modality of choice for ependymoma is MRI. CE is hypo intense in T1 while hyper-intense in T2. Foci of signal heterogeneity within a solid neoplasm represent hemosiderin, necrosis, or calcification, which account 40% - 80% of cases. Post-contrast, it usual enhance in a heterogeneous pattern [4] [5].

The histopathological characteristics of CE are not different from the intraventricular ependymomas. The tumor cells are organized in perivascular pseudorosettes and to a less extent in true ependymal rosettes. The cytological features of ependymomas include round to oval nuclei with evenly dispersed stippled chromatin. Unusual morphological features like clear cells, spindle cells and giant cells, tanycytic, epithelioid can also be seen. For cases of anaplastic CEs glioblastoma and pleomorphic xanthoastrocytoma (PXA) should be considered in the differential diagnosis [1] [2] [4].

MIB-1, L1, and Ki-67 are known to be the most immunoreactive factors and are associated with high-grade ependymomas. However, a recent further markers have been found, including topo-II- α and p53 and murine double min 2 protein expression which are correlated with high-grade tumors and a poor prognosis. RELA fusion-positive ependymoma was found to be characteristically associated with supratentorial Ependymoma and related to a worse outcome [5].

Treatment is similar to the classical (intra-ventricular) ependymomas. Complete surgical resection is the treatment of choice followed by radiation therapy for both low and high grade CEs [1] [2]. Chemotherapy is given to children below the age of three after incomplete resection of the tumor to delay or avoid irradiation [1] [2].

Cortical ependymomas appear to have a relatively favorable prognosis [1] [2] [4].

Low grade CE showed a lower likelihood tumor recurrence and lower mortality [2] [5].

Tumor recurrence is higher in high grade CEs (12 patients out of 21) compare to 3 patients out of 24 in the low grade CEs during 48 months follow up [3].

4. Conclusion

Supratentorial cortical ependymomas are a very rare type of primary brain tumors with only 50 reported cases in the literature including adults and pediatrics. Patients usually present with seizures and the tumor exhibit solid or mixed appearance on imaging. Total resection followed by radiotherapy is the treatment of choice.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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