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Case Report

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A CASE REPORT: GRADE III EPENDYMOMA IN 74 YEARS OLD FEMALE

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ABSTRACT

Grade III anaplastic ependymomas are a rare type of brain tumors. Ependymomas have distinct subtypes depending on their localization, with some types being linked to the patient's age. These tumors are infrequently found outside the ventricular system. Commonly originate from the ependymal cells within the ventricles but it may also extend to cerebral hemispheres or infratentoriel. We present a rare case of grade III ectopic ependymoma in a 74-year old female with progressive onset of a sensory deficit involving the left side of the body, starting at the left lower limb with gradual extension to the upper limb. The anatomopathological results are in favor of an ependymoma grade III subtype for which surgical resection with complementary radiotherapy was considered as the first choice of treatment. **Aim:** To study the clinical features, imaging findings, and treatment. **Conclusion:** Grade III ependymomas are rare subtype of CNS tumors that seem to be diagnosed more often. The clinical progressions and management of such patients are quite variable. The significance of adjunctive therapy is continuously debated yet surgical intervention is the main choice of treatment.

KEYWORDS: Ependymoma, Anaplastic Ependymoma, and Neuropathology.

INTRODUCTION

Anaplastic Ependymoma is a slow growing soft glial-based neuroepitheliel tumor arising from ependymal cells. Ependymomas may arise along the entire neuraxis, whether found inside ventricles, cerebral hemispheres, or occasionally within the central canal in the spinal cord. Grade III Ependymomas represent about 9% of all neuroepithelial tumors. Most commonly seen in children between 1 to 2 years of age, comprising 12% of all pediatric CNS tumors.[1] Ependymomas usually present as lobulated heterogeneous masses of 2-6 cm in size with about 60% of them infratentorial, no gender preferences with manifestation at all ages.[2] Infratentorial ependymoma arise most often in children while supratentorial types seen commonly in adults. Anaplastic ependymoma is the most aggressive subtype and according to the World Health Organization's (WHO) classification of central nervous system tumors, anaplastic ependymoma is classified as grade III. Ependymomas located outside the ventricles may be labeled ectopic, cortical, or extraventricular ependymomas.[3]

CASE REPORT

A 74y- old female with known history of HTN, presented with progressive onset of a sensory deficit involving the left side of the body, starting at the left lower limb with gradual extension to the upper limb. Later she had a motor deficit on the lower limb with extension to the upper member sparing the face. After seen in emergency, a cerebral scan was demanded followed by a cerebral MRI with contrast revealing an intra-axial mass measuring 42 mm long, located in the right central region (retrorolandic area), presenting a very heterogeneous signal globally in a discrete isointense signal T1, hyper-signal FLAIR (*figure 1*). After injection of gadolinium, this lesion appears well delimited with an intense and heterogeneous enhancement. On the Diffusion sequences, there is an increase in rCBV at 8-fold associated with a discrete rupture of the blood-brain barrier. On the spectroscopic sequence, tumor profile finding indicated an elevation of choline (ratio Choline / Naa> 1) with an increase in lipids.

Patient was admitted under care of neurosurgery. She was treated for a para-sagittal parietal lesion due to progressive hemiparesis with major sensory disorders in the lower limb. Patient in supine position and under general anesthesia, a sponge under the right shoulder with head slightly turned to the left in Mayfield head holder. Using the neuro-navigation system and 3D T1 MRI with Gadolinium injection for localizing. Before the surgical draping, cutaneous sterilization of the skin and then subcutaneous infiltration with adrenaline xylocaine. A parietal inverted U-shaped incision and then dragging

the scalp downwards. A parasagittal parietal flap at the level of the median line with hinged dural opening and traction. We find a part of the cortical lesion that flushes behind the post-central groove.

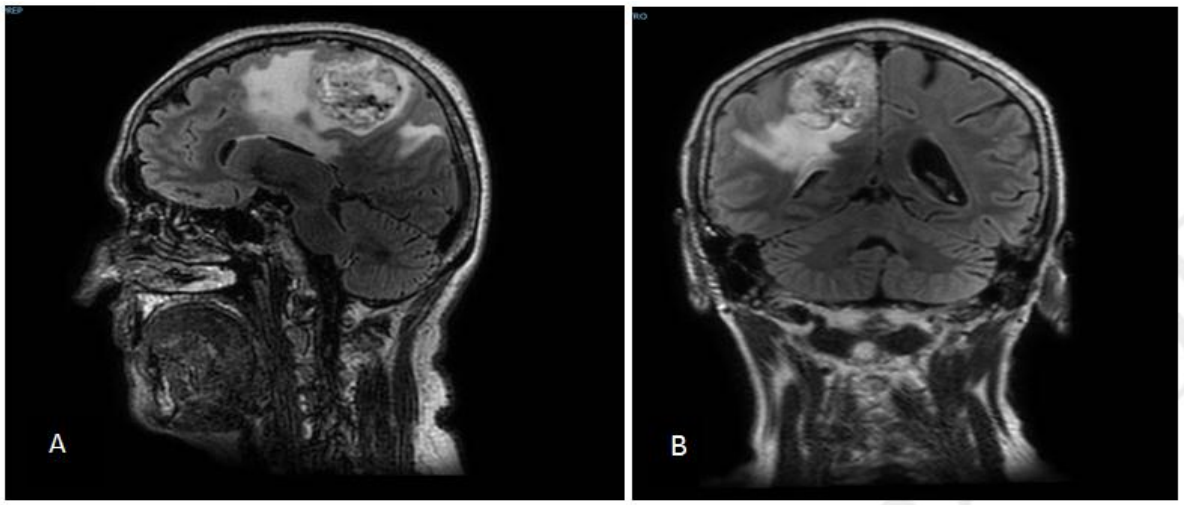

Figure. 1: (A) Showing a preoperative MRI sagittal flair T2 sequance of parietal lobe lesion, (B) Coronal sequence showing the lesion in cerebral cortex of the upper right hemisphere.

A complete macroscopic surgical resection performed. The Operative follow-up is generally favorable even if there is still persistence of a sensory deficit predominant in the left lower limb. He walks with the help of a cane, benefits from physiotherapy owed to sensory disorders that show gradual improvement.

Follow-up and outcome: The histological and immunohistochemical characteristics observed are in favor of an anaplastic grade III ependymoma with a clear cell component and territories of anaplasia (*figure 2*). Proliferation of tumor cells with presence of abnormal mitoses observed in the histological studies. Moreover, elevated mitotic activity with relative cell density with marked cytonuclear atypies and combined tumor necrosis regions. *H&E* (a) staining showing highly cellular lesion with increased mitotic activity (original magnification ×400) and multiple immunolabelings were performed showing tumor proliferation through *GFAP* (b), *OLIG2* (e), and *ATRX* being positive with the *EMA* (c) focuses on a few tumor cells. The *Ki67* (d) proliferation index is high in the anaplasia territories of up to 20-30%. These features permit us to classify this tumor as grade III according to the international histopronostic classification guidelines of World health Organization of CNS tumors. The general neurological condition is completely preserved; with no complain of any neurologic symptoms.

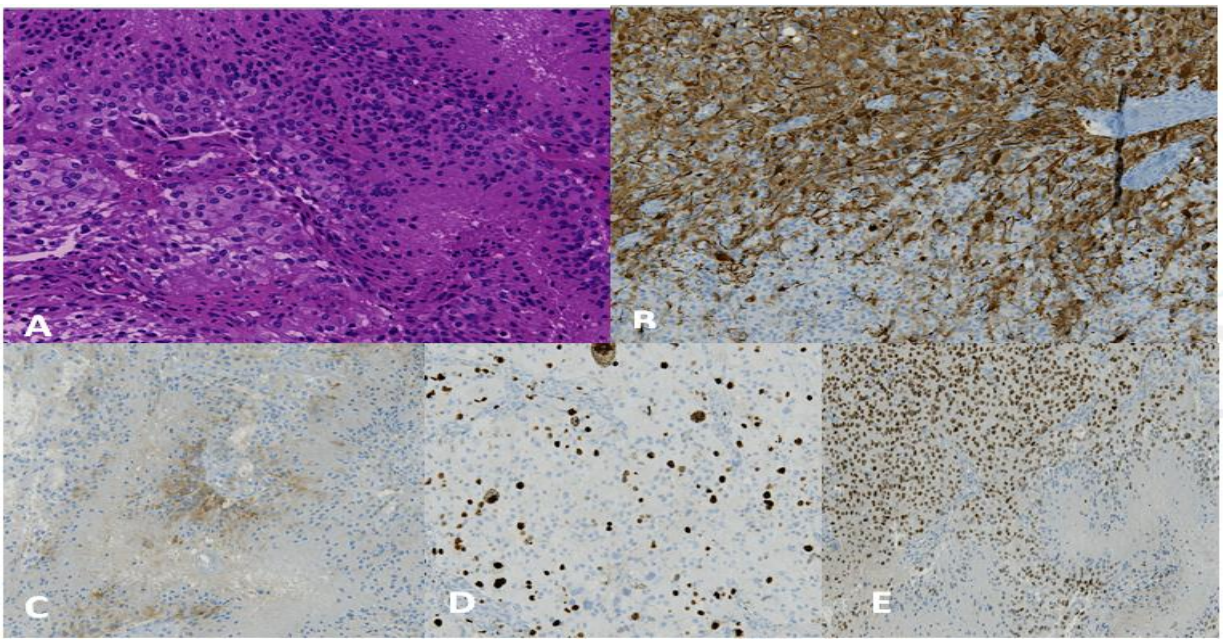

Figure. 2: Microscopic images of Histopathology staining with x400 magnification; the different staining types (A) HE (b) GFAP (c) EMA (d) Ki67 (e) Olig2.

A Post-op CT (figure 3) showed large anterior frontal pneumoencephaly with minor edema of 10 mm thickness within the excision cavity of the right central region. There continues to be significant vasogenic edema, stable compared to MRI. A bony window is seen in the right fronto-parietal region due to the craniotomy. Stability of the mass effect with no temporal or tonsillar involvement and enhancement in the ventricular system. The general neurological condition is completely preserved; with no complain of any headache or any other neurologic symptoms. After multidisciplinary meeting, complementary radiotherapy has been agreed without chemotherapy.

DISCUSSION

Intracranial ependymomas are commonly located in in children and are relatively rare in adults [4]. In adults, supratentorial ependymomas are usually found in lober hemispheres with cortical installation [5]. Some authors state that these tumors form in the lateral ventricle and expand toward the cerebral tissue describing them as supratentorial extraventricular anaplastic ependymoma. These anaplastic ependymomas don't present with any distinct clinical appearance, generally neurological signs such as vomiting and headache in relation to increase intracranial pressure within the nervous system.

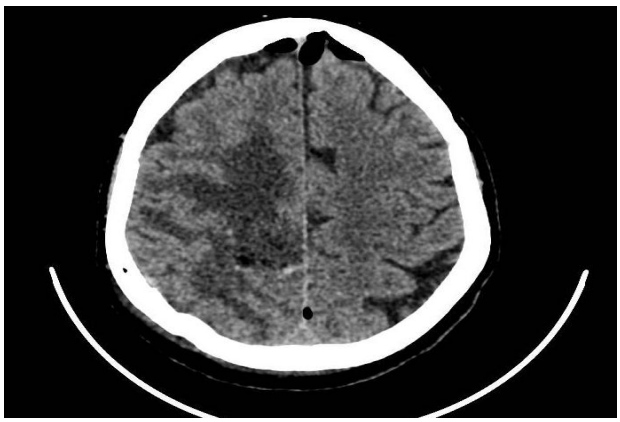

Figure. 3: post-op CT finding shows edema within the excision pathway and near the central region, with slight pneumoencephaly in the parietal lobe of the right hemisphere.

Due to pathophysiology of the nervous system, clinical features only present as the lesion begins to occupy more and more cerebral space. Occasionally the clinical features develop only once the lesion enlarges to cortical surface.[6] There are no classic radiological images for these tumors; glial tumors are not straightforwardly differentiated from one type to another.[3,7] However, clear observation of focal signal heterogeneity with necrosis is seen these tumors. In the literature, the embryonic ependymal parenchymal remnant during brain development may be related to the formation of such tumors.[1,8] Others suggest that extraxial ependymomas such ovarien localization and CNS subtypes are from different origins with distinctive morphology.[9,10]

CONCLUSION

Anaplastic Ependymoma just like other cortical tumors may generally cause seizures and other focal neurological deficits. Differential diagnoses are made according to immunohistochemical findings. Considerable awareness is needed by performing more studies of such conditions to assist in determining the cause and for establishing a histopathological based therapeutic plan for anaplastic ependymomas.

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