

Rhabdoid Meningioma: Case Report of a Rare Pathological Entity

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Abstract

Rhabdoid meningioma is a rare aggressive variant of meningioma. Histopathologically, it is particular and distinctive type having abundant eosinophilic cytoplasm with eccentrically placed nuclei. It has been included in the revised WHO classification of tumours of the CNS as a subtype of meningiomas with high risk of recurrence, more aggressive growth and poor survival, corresponding to WHO grade III.

Keywords: Meningioma, Rhabdoid, Aggressive, WHO Grade III.

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INTRODUCTION

Rhabdoid meningioma is an unusual variant of meningioma classified as a grade III subtype under the new World Health Organization (WHO) classification of brain tumors. This particular histological subtype of meningioma was first described in 1998 as an unusual variant of meningioma. It is associated with rapid growth and known for its aggressive behavior, high incidence of recurrence, and poor prognosis than others.

Here, we are reporting a rare case of rhabdoid meningioma in a middle-aged male patient and describing the clinical and pathological features of this entity.

OBSERVATION

A 40-year-old patient with no significant pathological history, came to the consultation for intracranial hypertension symptoms, balance disorders and left V2 - V3 neuralgia. The clinical examination found a GCS=15/15 and a statokinetic cerebellar syndrome (lateralised on the left). Ocular fundus examination objectified bilateral papillary oedema.

Brain MRI showed an extra-axial lesion straddling the temporal lobe and the posterior brain fossa. It had low signal intensity on T1-weighted images (Figure 1), while on T2-weighted images; its signals were hyper intense after injection of

Gadolinium. The lesion pushes back the brain stem and cerebellar hemisphere. This radiological aspect was suggestive of a V neuroma.

Left subtemporal approach was selected to operate the patient and revealed an extra-axial, whitish, non-bleeding lesion. Brain CT at the end of the procedure did not show post-operative bleeding.

Histopathological examination, including immunohistochemistry, of the tumour fragments concluded to a rhabdoid meningioma: it had large tumour cells, with eccentric nuclei with a small prominent nucleolus, surrounded by an abundant eosinophilic cytoplasm (Figure 2). It expressed vimentin and EMA on immunohistochemical study with a Ki67 proliferation index evaluated at 45% (Figure 3).

Our patient was then sent to the radiotherapy department for additional therapy. The evolution was spectacular both clinically (disappearance of intracranial hypertension symptoms and V neuralgia) and radiologically (brain MRI made at a regular rate showed total removal of meningioma and the absence of recurrence) (Figure 4).

DISCUSSION

Rhabdoid meningioma was first observed by Kips and Parry in 1998. It is a recently described entity

(only about thirty have been reported in the literature) [1-3].

It is classified as WHO Grade III, which implies a high rate of recurrence, and aggressive behaviour either by its malignant local growth or by its metastatic dissemination [1, 4].

In general, rhabdoid meningioma occurs on average around age 50 years with a female predominance [1, 3]. The incidence is 0.75 to 1% of all meningiomas.

It has no preferential location [1]. The average survival is 48.3 months (this varies according to the quality of removal) [1, 5]. It can be primitive from the outset or constitute a recurrence of benign meningioma [6].

Histopathologically, the cells are similar to those found in other locations of tumors of rhabdoid appearance: namely, a large eccentric nuclei with a prominent nucleoli and an abundant eosinophilic cytoplasm [1, 6].

In immunohistochemical analysis, rhabdoid meningioma expresses the following antibodies: EMA, progesterone receptors (PR) and vimentin. The Ki67 proliferative index marker is generally high [1, 7].

The treatment is not codified. However, surgery remains the ideal treatment and external radiation therapy is considered in case of recurrence of the lesion and/or in case of incomplete removal. Proton therapy seems to be a promising alternative. Chemotherapy and radiosurgery have not proven their effect on this tumor [1, 5, 8].

CONCLUSION

Rhabdoid meningioma – histologically diagnosed - is a recently described and very rare variant of meningioma. Complete excision along with dural attachment, appropriate histological diagnosis and grading, and adjuvant radiotherapy are imperative for proper management.

Conflicts of interest

The authors do not declare any conflict of interest.

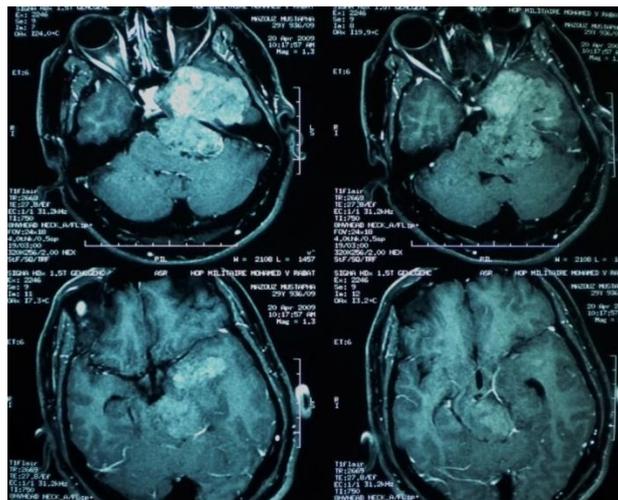


Fig-1: Brain MRI showing the location and extent of the tumor

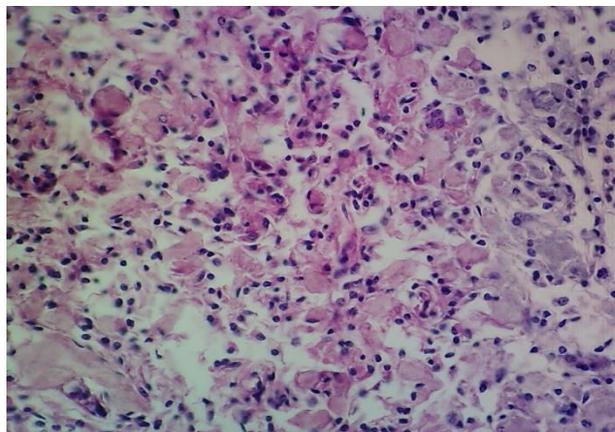


Fig-2: Tumor cells are large, displaying abundant eosinophilic cytoplasm and eccentric nuclei (HE, Gx400)

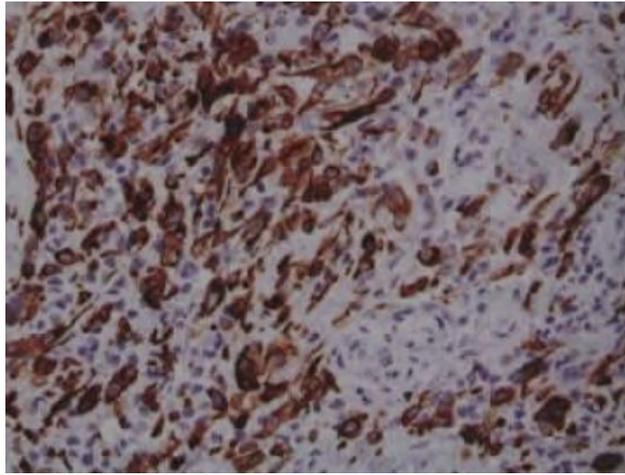


Fig-3: Positive immunostaining for Vimentin.

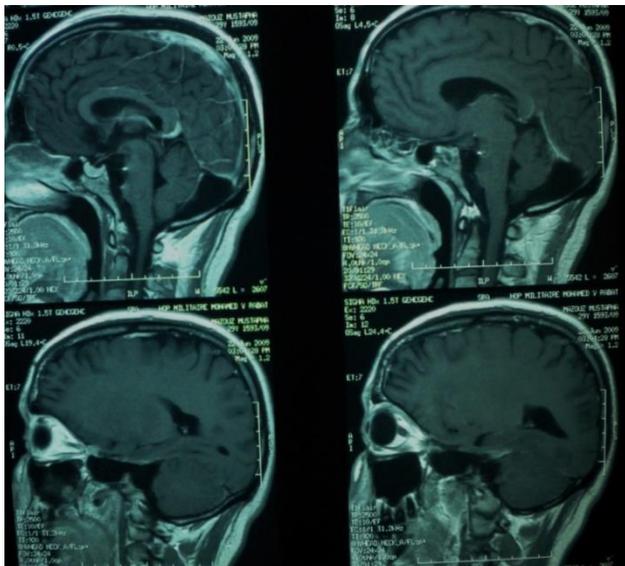


Fig-4: Control brain MRI after one month of surgery

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