A Rare Aggressive Tumor of The Sellar Region: Atypical Teratoid/rhabdoid Tumor

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Introduction

- Atypical teratoid rhabdoid tumor (AT/RT) is a rare, highly malignant central nervous system neoplasm.
- ATRT is extremely uncommon in adults, with a male predominance and a mean diagnostic age of 29 years.
- It is most commonly supratentorial with only 9 confirmed adult cases localized to the pineal region.

Herein, we report a metastatic AT/RT case in an adult who displayed an aggressive clinical course and complicated with pituitary apoplexy.

Clinical Case

- 55-year-old woman → headache, polyuria polydipsia, visual impairment in the left eye.
- Brain MRI: 12x20 mm pituitary mass.
- **Within weeks, the patient complained of worsening.** Physical examination: **Visual acuity was decreased in both eyes with limited temporal vision.**
- Follow-up pituitary MRI demonstrated in Figure-1 (the mass has grown more quickly before) and she had panhypopituitarism and hypernatremia secondary to diabetes insipidus → prednisone 7.5 mg/day and desmopressin 60 µg/day.
- The patient's headache intensified → pituitary MRI: **haemorrhage in tumor** → Transsphenoidal surgery (After complete resection of the tumor, bleeding occurred in the pons and mesencephalon).
- Pathology: Atypical teratoid rhabdoid tumor (Figure 2).
- Spinal and cranial MRI : multiple metastatic lesions on the thoracic vertebras (Radiotherapy was applied).
- She was followed in the intensive care unit, then the patient passed away.

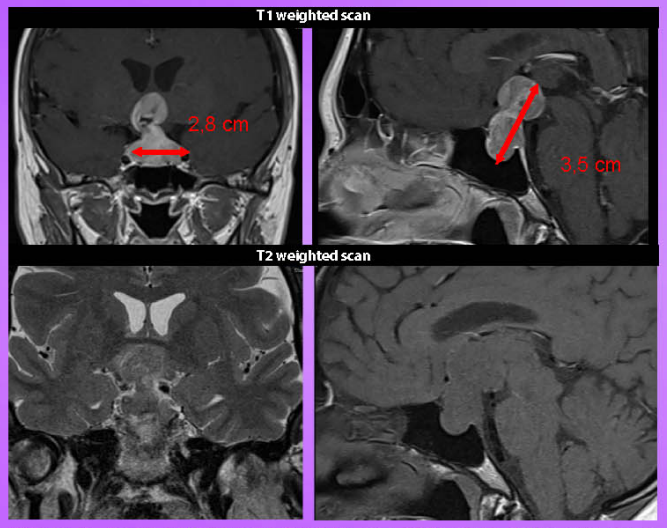

Figure 1. Large sellar mass (2.5X3,5 cm) with suprasellar extension and heterogeneous enhancement without cavernous sinus infiltration, however there is an anterior and posterior indentation to the optic chiasm.

Conclusion

- AT/RT is a tumor of the central nervous system which is rare in adults and in sellar region.
- AT/RTs are highly aggressive tumors and require early intervention.
- Imaging methods may not be distinctive in the diagnosis of the disease therefore, these tumors should be suspected in the rapid clinical course.
- Treatment guidelines for ATRT in an adult population have not been established, should be treated multimodally.

CONTACT INFORMATION

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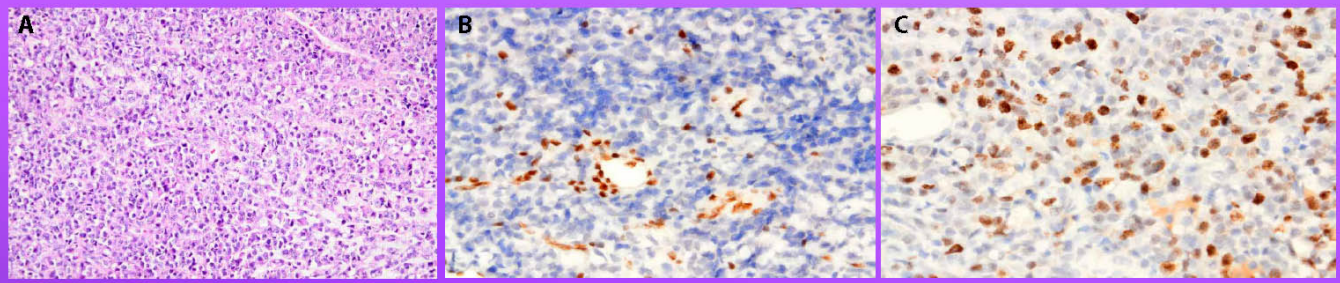

Figure 2. -A-EMA (+), SMA (+), CD34 (+), TTF-1(+), SALLA-4(+), P53 (10-15%); Braküri(-) GFAP (-) S100(-) IDH1 (-) B-Poorly differentiated, malignant neoplasm with loss of INI expression C- Ki-67 LI: 20%