THERATOID RABDOID/ATYPICAL TUMOR IN AN ADULT

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BACKGROUND

Pituitary apoplexy is an ischemic or hemorrhagic phenomenon that usually appears on a pituitary adenoma, although it may occur on other less frequent tumors in adulthood.

CLINICAL CASE

We report the case of a 43-year-old female, having clinical presentation consisting in headache for a period of 3 months as well as vomiting, diplopia and left ptosis prior to admission. An image of a sellar pituitary tumor (20x23mm) with suprasellar growth (with obliteration of the parasellar tank and cavernous sinus) compressing the optic chiasm was seen by magnetic resonance imaging (MRI) confirming the presence of invasive pituitary macroadenoma with subacute bleeding (Fig 1) Analytical data showed hypopituitarism with partial data. A tumor resection by transsphenoidal approach was perfomed, suspecting pituitary apoplexy. The hystopathological finding indicated rhabdoid tumor/teratoid atypical (WHO grade IV). The evolution was satisfactory until the ninth postoperative day when the patient started with a headache, vomiting and impaired level of consciousness. A CT scan was performed and confirmed a large isointense mass of about 7,2cm in temporal and frontal rights lobes and suprasellar region, collapsing the right lateral ventricle. The patient finally died at 27 days after surgical intervention.

Fig. 1: MRI pituitary. Coronal section. T1 sequence with contrast. Day 1. Sellar tumor (20x23 mm) with suprasellar growth and subacute hemorrhage.
Fig 2: CT scan with contrast. Sagital section. Day 30 (20 days after surgery): Isodense sellar tumor (7.2 cm) with infiltration of the sellar region, cavernous sinus, mass effect and enhances intensely with intravenous contrast.

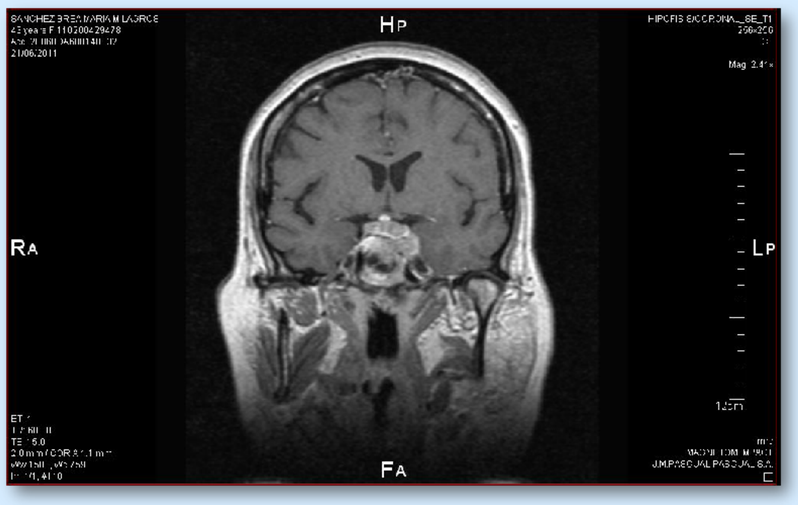

FIG 1.

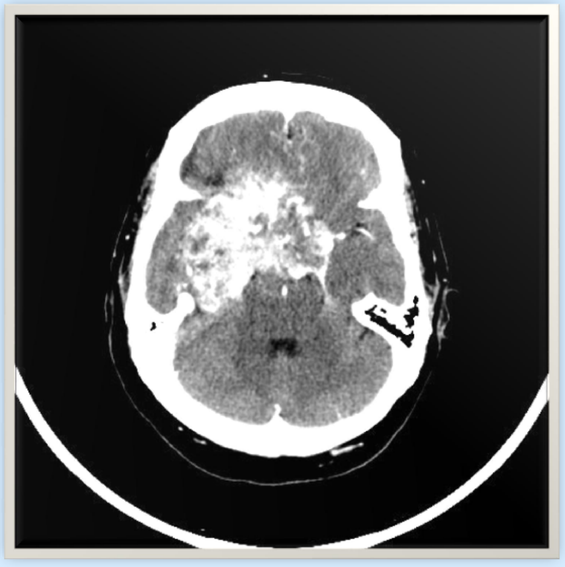

FIG 2.

DISCUSION

Even though pituitary apoplexy usually occurs in the context of a pituitary adenoma, it can also occur over other tumors. Teratoid/atypical rhabdoid tumor is a rare tumor of the CNS in adults and its located in the pituitary, which can make the diagnosis difficult, We need an adequate immunohistochemical evaluation, because the loss of expression of INI1 is pathognomonic for this tumor. Because of its low incidence, there are no specific therapeutic management protocols for the adult population. The prognosis is ominous in a variable time.

AUTOR (Year)	AGE/SEX	LOCALIZATION	TRACKING TIME	OUTCOME
Kuge et al. (2000)	32 years/F	Sellar and suprasellar	28 months	Death
Raisanen et al. (2005)	20 years/F	Sellar and suprasellar	28 months	Survival
Raisanen et al. (2005)	31 years/F	Sellar and suprasellar	9 month	Death
Arita et al. (2008)	56 years/F	Sellar and right cavernous sinus	23 months	Death
Las Heras and Pritzker (2010)	46 years/F	Sellar	-	-
Schneiderhan et al. (2011)	61 years/F	Sellar, suprasellar and left parasellar	3 months	Death
Schneiderhan et al. (2011)	57 years/F	Sellar and left parasellar	6 months	Survival
Hae Gi Park et al (2014)	42 years/F	Sellar and suprasellar	27 months	Survival
Present Case	43 years/F	Sellar, suprasella and right parasellar	35 days	Death

Table 1. Summary of teratoid/atypical rhabdoid tumors reported in the literatura. (Adapted from Hae Gi Park et al, 2014)

CONCLUSION

Our case shows the rapid progression of the disease, and is one of the TTRA cases in adults with more aggressive behavior and poor survival from diagnosis, as published in the literature so far.

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