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Double Localization of a Cerebral Germinoma: Case Report

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Abstract **Case Report**

Germinal cell tumors are embryonic tumors located mainly in the gonads, and their cerebral localization is rare, representing less than 1% of intracranial neoplasia. Clinical expression can be polymorphous, while imaging provides a highly suggestive radiological semiology. We report the case of a 31-year-old patient presenting with diabetes insipidus evolving for one year and associated with decreased visual acuity. Cerebro-medullary MRI revealed a tumoral process in the pineal region associated with thickening of the pituitary region. Tumor markers (β HCG and AFP) in plasma and CSF were negative, confirming the diagnosis of pure germinoma without the need for biopsy. Treatment is mostly based on radiotherapy, and sometimes on a combination of radiotherapy and chemotherapy. The latter was our therapeutic choice, with a good clinical and biological evolution.

Keywords: Cerebral Germinoma, Tumor, Gonads, MRI, Diabetes.

INTRODUCTION

Germ cell tumors are embryonic tumors located mainly in the gonads. Their cerebral localization is rare, representing less than 1% of intracranial neoplasia. They can be pure (with a good prognosis), or mixed secretory or non-secretory (with a poorer prognosis) [1]. They most often affect adolescents and young adults, with a clear male predominance. The contribution of imaging and tumor markers is considerable, and may even obviate the need for biopsy, which can sometimes prove difficult [3]. These are chemo- and radiosensitive tumours with an excellent prognosis [2].

OBSERVATION

D.A, aged 34 with no particular pathological history, presented for one year with diabetes insipidus associated with a decrease in visual acuity evolving in a context of conservation of general condition. Clinical examination revealed a hemodynamically and respiratorily stable OMS:1 patient with a Glasgow score of 15/15, with no sensitivomotor deficits and a slight drop in visual acuity to 9/10 ODG with no papilledema. The rest of the clinical examination revealed no endocrine or neurological signs.

Radiological findings: cerebral MRI revealed a nodular lesion in the pineal region measuring 12.2*10.8*9.3 mm, associated with thickening of the pituitary stem suggestive of a germinoma.

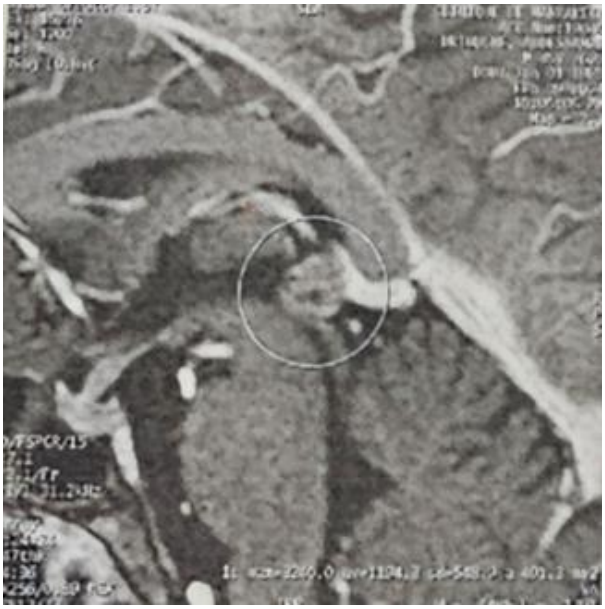

Figure 1: T1-weighted sagittal cross-section brain MRI after gadolinium injection, showing pineal localization

Serum and CSF tumor marker assays (β HCG and AFP) were negative, as was CSF cytology, confirming the diagnosis of a pure germinoma without the need for biopsy, which the surgeons considered difficult. Medullary MRI and thoraco-abdomino-pelvic CT scans revealed no medullary or distant extension.

The patient underwent initial chemotherapy according to the following protocol: courses 1 and 3: D1: carboplatin 600mg/m²/d; D1 to D3: etoposide 100 mg/m²/d, courses 2 and 4: D1 to D5: ifosfamide 1800 mg/m²/d; D1 to D3: etoposide 100 mg/m²/d. Cures are spaced 3 weeks apart: J1=J21, followed by ventricular radiotherapy at a dose of 24 Gy with a 16 Gy boost on the initial tumors, with a complete response on follow-up imaging.

DISCUSSION

Germinomas are among the malignant germ cell tumours, most frequently affecting adolescents and young adults, with a clear male predominance[1]. Strictly extraneural in origin, but developing along the midline, particularly in the pineal and suprasellar regions, primordial germ cells, precursors of germ cells, localized in the extraembryonic mesoderm, migrate along the dorsal mesentery to reach the genital ridges which will give rise to the gonads. On their migration path during embryogenesis, they may develop in the pineal region, as at the anterior end of the 3rd ventricle, where they appear macroscopically months or years after the onset of diabetes insipidus. This is the hypothesis of poor cell migration and incorporation [4]. The tumor is most often found in the pineal region or in the suprasellar and hypothalamic region (35% of cases); intrasellar localization is possible but rare, and the association of intrasellar germinoma with pineal germinoma has already been reported [5]. Germ cell tumours can be pure, accounting for 2/3 of cases (pure germinomas), or mixed, associating several tissue components (mature or immature teratoma, hCG-secreting choriocarcinoma, alpha-fetoprotein-secreting yolk sac tumour or embryonal carcinoma) [7].

The clinical expression is polymorphous and associates an intracranial hypertension syndrome, occulomotor disorders and in particular Parinaud's syndrome due to compression of the superior colliculi, as well as an endocrine syndrome due to infundibular compression explaining diabetes insipidus, or potomania due to stimulation of the hypothalamus [13]. Tumor markers (HCG, AFP, PALP) are routinely tested in blood and CSF to rule out choriocarcinoma, embryonal carcinoma or endodermal sinus tumors. Finally, the presence of secondary bone and lung localizations should raise the possibility of pineal metastasis [6].

Cerebral imaging provides considerable diagnostic support. Cerebral CT scan reveals a well-limited, homogeneous, round or lobulated, iso or hyperdense mass in the pineal or suprasellar region, enhancing after injection of contrast medium [12]. MRI is essential for differentiating between a normal pineal gland and a pineal tumour, bearing in mind that a pineal gland measures between 5 and 10 mm in its long axis and 1 to 4.5 mm in thickness. Germinoma usually appears as a well-limited mass, isointense in T1 and iso or hyperintense in T2, taking up gadolinium homogeneously [8].

Diagnosis of certainty is based on histological data, but the new diagnostic means represented by tumor markers and modern imaging can lead to sufficient presumption to initiate treatment based on radiotherapy and chemotherapy. Indeed, several authors have concluded that medial multifocal localizations are associated only with germinomas, making biopsy unnecessary [9]. The detection of tumor markers in blood or CSF is systematic, as their positivity is sufficient to diagnose a less-differentiated secretory form, so that any elevation of αFP excludes the pure form, and requires more intensive therapy. The absence of tumour markers means that malignant secretory germ cell tumours can be ruled out. Nevertheless, some pure germinomas may be accompanied by very low ßhCG secretion, with concentrations of less than 50 IU/l in both serum and CSF (8-15% of cases, depending on the series), suggesting the presence of syncitiotrophoblastic cells in the germinoma, as has been described in extra-cranial forms. In the few patients presenting with moderate ßhCG elevation in serum or CSF, biopsy is of no diagnostic interest, but may be of therapeutic value, enabling the identification of various non-secreting tumour contingents (particularly teratomatous), whose presence may alter prognosis and therapeutic management [11]. Dissemination of this tumor is mainly via the lepto-meningeal route, but it may also disseminate into the cerebral parenchyma, or into the ventricular cavities and sub-arachnoid spaces; the initial extension work-up should therefore include cytological and marker analysis of the CSF, as well as spinal cord MRI [10].

Despite high cure rates, the current treatment of intracranial germinoma remains controversial. Intracranial germinomas can be cured by radiotherapy: overall survival after craniospinal radiotherapy ranges from 86% to 97%. The optimal dose is controversial, with the majority of authors opting for a dose of 40 grays [12]. Intracranial germinomas are sensitive to chemotherapy, in particular cyclophosphamide and platinum salts, but have a high recurrence rate (as in our patient's case), hence the interest in complementary radiotherapy. New therapeutic strategies have therefore been developed, using chemotherapy as a first step in order to avoid or reduce the volume of irradiation [14]. Some publications have reported an increased relapse rate for germinomas with moderate ßhCG concentrations compared with those with no ßhCG concentrations, implying regular follow-up of our patient to watch for late relapses that may occur in this entity. Bifocal forms should be considered and treated as locoregional and non-metastatic forms, as their evolution is identical [15].

CONCLUSION

Germinomas with a double pineal and suprasellar localization are still rare, representing only 5-

10% of germinomas, with only a few cases reported in the literature. Clinical expression is polymorphous, and imaging provides a highly suggestive radiological semiology.

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