Radiology Case Reports 17 (2022) 3015–3018

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journal homepage: www.elsevier.com/locate/radcr

Case Report

Intracranial bifocal germinoma☆,☆☆

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ARTICLE INFO

Article history:
Received 7 May 2022
Revised 25 May 2022
Accepted 28 May 2022

Keywords:
Bifocal germinoma
Suprasellar tumor
Pineal tumor
Hypophysis
Epiphysis intracranial germ cell tumour

ABSTRACT

Primary intracranial germ cell tumors are rare, often affecting children and young patients. Germinomas are the most common type of germ cell tumors. We present the case of a 10-year-old child, who was admitted with decreased visual acuity, asthenia, polyuro-polydipsic syndrome, and gait disorder. His biological assessment showed an hypocortisolemia and diabetes insipidus. Imaging found a bifocal process in the suprasellar and pineal region, suggestive of a bifocal germinoma. Imaging data and the positivity of human chorionic gonadotrophin in the cerebrospinal fluid were in favor of the diagnosis of bifocal germinal tumor confirmed by biopsy. Currently the patient is hospitalized in pediatric oncology department.

The diagnosis of germ cell tumors is based on imaging, tumor marker assays, and biopsy. They are treated by radiation therapy alone or in combination with reduction chemotherapy, and surgery for tumor residues. The location of the tumor usually makes surgery difficult.

Introduction

Primary germ cell tumors of the central nervous system are rare tumors of extraneural origin, secondary to aberrant migration of primordial germ cells during embryogenesis. Histologically they are similar to tumors found in the gonads.

They are classified into who main groups with different prognostic significance and therapeutic implications: germinomas (or seminomas) which are the most frequent (45%) of germ cell tumors, they may include syncitiotrophoblastic cells which explains the possibility of low human chorionic gonadotrophin (hCG) secretion. And non-seminomatous germ cell tumors, which include embryonal carcinoma, yolk sac tumor that secretes alpha-fetoprotein, choriocarcinoma, ter-

☆ Competing Interests: All authors declare no conflict of interest.
☆☆ Abstract

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https://doi.org/10.1016/j.radcr.2022.05.080

atoma (immature and mature), and mixed germ cell tumors [1].

They are most often diagnosed in children and young adults. These tumors represent about 0.9% of pediatric tumors, 3% to 5% of childhood brain tumors, and 28.7% of germ cell tumors [2]. They most often arise in the pineal and/or supra-sellar region. Supra-sellar germ cell tumors usually affect hormonal function, vision may also be affected, if the tumor is close to the optic nerve. Children with germinoma have a 90% chance of recovery.

Case report

A 10-year-old child, without any notable pathological history, who was admitted for the management of a decrease in visual acuity that goes back to 1 year ago, the symptomatology was aggravated by asthenia with a severe polyuro-polydipsic syndrome quantified at 168 cc/kg/day, and gait disorder, in a context of apyrexia, with conservation of the general condition.

Clinical examination on admission found a child in good general condition, apyretic, normotensive (blood pressure at 110/60 mm Hg) without orthostatic hypotension. Diuresis estimated at 7 cc/kg/h, weight at 41.5 kg; height at 133 cm; head circumference at 51 cm; heart rate at 86 b/min, respiratory rate at 21c/min and who saturates properly at ambient air.

The neurological examination showed ataxic walking, cutaneous hyperesthesia with deep sensibility disorder, without sensory level, or motor deficit, with hyperactive tendon reflex, and oculomotor paralysis concerning vertical movements (a Parinaud's syndrome).

The ophthalmological examination noted a visual acuity of 3 of 10 in the right eye, and 2 of 10 in the left eye. The photomotor reflex was decreased in both eyes with pupils in spontaneous semi-mydriasis. Schirmer's test was normal 10 mm at 5 minutes. Posterior segment examination found stage 2 of bilaterally papilledema. Examination of the external genitalia noted a stage 1 according to Tanner's classification. The rest of the somatic examination was unremarkable.

Biologically, the obvious causes of polyuro-polydipsic syndrome were ruled out, including normal kalemia, normal glycated hemoglobin, and correct kidney function. Urinary osmolarity was at 64,073 mosm/L. A pituitaryogram showed a low 8 hour cortisolemia level at 1.7, and a TSH at 2,320 mu/L.

A bHCG tumor marker assay came back positive at 15.54 mIU/mL at the cerebrospinal fluid (CSF). Plasma bHCG and alphaFP were negative. CSF immunohistochemistry of Ac anti Placental Alkaline Phosphatase and Ac anti Glial fibrillary acidic protein were negative.

A radiological assessment (cerebral computed tomography and magnetic resonance imaging [MRI]) revealed the presence of 2 lesions, one in the supra-sellar region, well limited, with regular contours, isodense in spontaneous contrast, homogeneously enhanced after contrast, measuring 10 × 12 mm in antero-posterior and transverse diameter, and coming in contact with the optic chiasma (Fig. 1). And the second lesion at the level of the pineal gland, well limited, with lobulated contours, discretely hyperdense in spontaneous contrast, which contains a macrocalcification, and measures 8 × 10 mm of antero-posterior and transverse diameter (Fig. 2), in favor of a bifocal germinoma. MRI also showed cerebro-medullary dissemination, in the form of multiple small lesions in hypersginal T2 and Fluid Attenuated Inversion Recovery (FLAIR), millimetric enhanced after contrast at the levels of the central gray nuclei, internal capsules, and cerebral barter. A nodular enhancement of the ependymal walls of the frontal horn of the lateral ventricles, and pia-matter.

Biopsy confirmed the diagnosis. The patient was put on hormone replacement therapy, based on hydrocortisone 10 mg (1 cp in the morning, 3/4 cp in the evening, 17 mg/d), and Desmopressin 60 microg 1 cp/d. the patient also received 5 days of solumedrol bolus 500mg/m2/

Imaging, positivity of Bhcg elevated in the CSF, and biopsy allowed to retain the diagnosis of bifocal germinoma, metastatic since the CSF was infiltrated by suspicious cells, and cerebro-medullary dissemination on MRI. The patient was put under primary chemotherapy.

Discussion

Most germ cell tumours (90%) occur in patients under 20 years of age, mostly in early adolescence, 2 to 3 times more frequent in boys than girls. They occur mainly in the pineal region and are usually germinomas. Non-seminomatous germ cell tumors and germinomas occur with the same frequency in the supra-sellar region, without gender predominance at this level [1].

Bifocal tumors in the pineal and suprasellary regions are not considered as metastases, and are most often germinomas. Uni- or bifocal tumors located in these sites should evoke a germ cell tumor and indicate a determination of alphafetoprotein (αFP) and hCG markers in blood and CSF. Localization in the thalamus, hemispheres, and multiple sites are rare (less than 10%). Meningeal involvement is sometimes visible on MRI or CSF analysis.

Clinical signs depend on the size and location of the tumor. The tumor can manifest itself either acutely by causing hydrocephalus that leads to intracranial hypertension by obstruction of the aqueduct of sylvius for tumors of the pineal region. Subacutely or chronically in the supra-sellar region, by a neuro-ophthalmological symptomatology (decrease in visual acuity, Parinaud's syndrome, diplopia, visual field alteration) by compression and / or invasion of the optic chiasma, Or neuroendocrine symptomatology (growth retardation, early or delayed puberty, decreased thyroid function, fertility problems, hyperprolactinemia and / or diabetes insipidus, or panhypopituitarism) [1,3].

The biological assessment to be performed in case of suspicion of primary germ cell tumor is:

__Specific tumor markers: αFP which indicates the presence of a yolk tumor contingent, total hCG and / or β hCG in the blood and CSF, whose value between 0.5 mIU/mL and 50 mIU/mL points to the presence of a secreting germinoma contingent, and to the presence of a choriocarcinoma contingent if total hCG >50 mIU/mL.

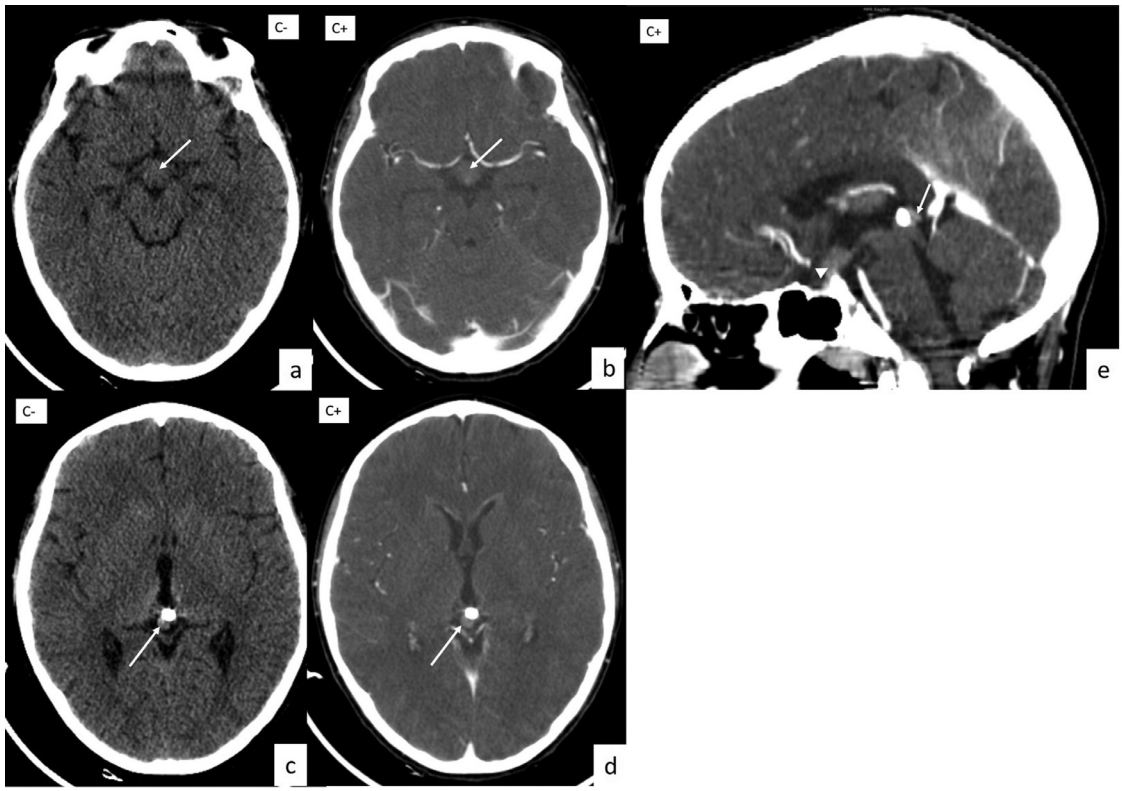

Fig. 1 – Axial non-injected (A,C), axial injected (B,D), and injected sagittal reconstruction (E) CT scan: showing suprasellar lesion (A,B,E), pineal gland lesion (C,D,E).

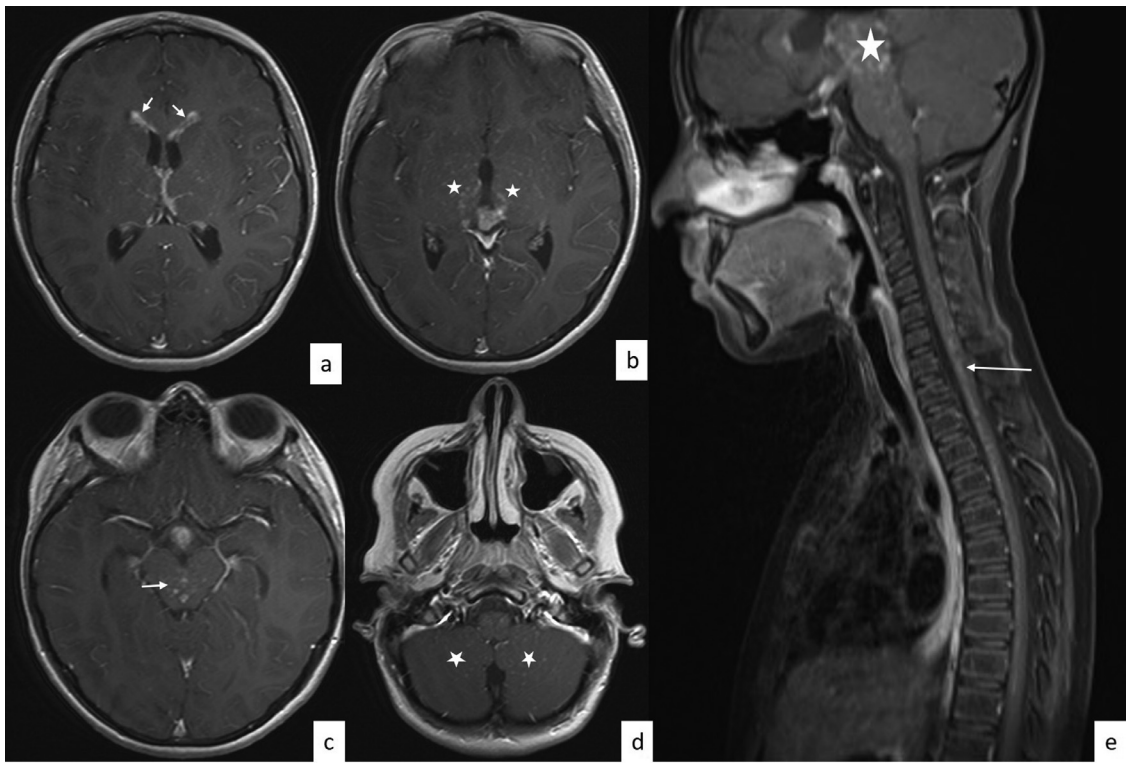

Fig. 2 – Axial (A,B,C,D), and sagittal (E) injected T1 MRI showing cerebro-medullary dissemination.

The positivity of αFP or ßHCG in the blood or CSF confirms the germinal and secreting nature of the tumor, histological proof is not necessary. For non-secreting tumors, histological evidence is essential by biopsy or even surgery if it is not risky, except in case of bifocal lesions because the vast majority are germinomas.

_ An endocrine assessment in search of a pituitary deficit.

_ Brain and spinal cord imaging: Primary germ cell tumors develop in the midline (pineal region, and/or supra-sellar in uni- or bifocal form) or in the thalamus. Tumors of the basal ganglia are often cystic and/or hemorrhagic masses, and are accompanied by homolateral brain atrophy. Germinomas are often well limited, homogeneous and enhance after injection, sometimes they are hemorrhagic or present calcifications. Teratomas have a heterogeneous appearance with sometimes intratumoral cysts and calcifications, and enhance inhomogeneously. Some of these lesions may disseminate, hence the need to look for meningeal localizations and to perform a spinal MRI that should explore the entire axis including the cul de sac.

The MRI characteristics of supra-sellar germinomas, such as T2 hypointensity and mild enhancement of the solid component, the presence of cystic lesions, and the lower apparent diffusion coefficient compared to other tumors (lymphoma, pinealoblastoma, medulloblastoma, high-grade glioma) allow the diagnosis to be made. The extension to the pituitary gland, to the pituitaite stalk and the presence of a synchronous tumor in the pineal gland are orienting elements [4]. A magnetic resonance imaging classification of supra-sellar tumors describes 5 types: a tumor of the floor of the third ventricle which extends dorsally to the cavity of the third ventricle with or without a visible pituitary stalk (Type Ia, Ib), ventral extension to the suprasellar region and/or the sella turcica (Type III), in both directions (Type II), small lesions on the floor of the third ventricle which extends to the pituitary stalk (type IV) and tumor localized in the pituitary stalk (Type V) [2].

A complete radiological assessment which includes a testicular ultrasound, thoraco-abdomino-pelvic scanner is necessary to rule out metastasis of an extracranial germ cell tumor. A neuro-ophthalmological assessment. And a fertility preservation consultation before starting chemotherapy.

In case of hydrocephalus or intracranial hypertension, endoscopic ventriculo-cysternostomy or ventriculo-peritoneal shunt, or even external shunt is performed. Biopsies are proposed only if the diagnosis is not established after analysis of tumor markers in plasma and CSF. They make it possible to eliminate differential diagnoses such as gliomas and pineal parenchymal tumors. Given the sensitivity to chemotherapy and radiotherapy, initial excisional surgery is not recommended. Excision is reserved for mature teratomas and residue after chemotherapy in non-seminomatous tumours.

For the diagnosis PALP assay is not in routine practice. β-hCG may help in the diagnosis and monitoring of germinomas. The presence of malignant cells beyond 14 postoperative days indicates the existence of metastases.

In metastatic forms, some authors recommend craniospinal radiotherapy without chemotherapy. Certain studies report that patients do not fully recover after radiotherapy (endocrine and psychomotor sequelae), and that pituitary insufficiency may even worsen, requiring lifelong hormone replacement therapy [5,2].

Conclusion

Intracranial bifocal germinoma is rare, it affects young people. Diagnosis can be made based on biology and imaging. The biopsy may be necessary if the biology is negative and the appearance on the imaging is atypical. The treatment is based on the combination of chemotherapy and radiotherapy

Patient consent statement

The patient has given his consent for the publication of this data.

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