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Case Report

Pediatric Suprasellar Teratoma: A Rare Location of a Germ Cell Tumor

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Abstract

In this case report, we describe a solid cystic tumor in the sellar region associated with hydrocephalus. The most frequent tumors in the midline within the age group (craniopharyngioma, Rathke's pouch cyst, pituitary adenoma) were immediately included in the differential diagnosis; however, the intraoperative findings were not conclusive. A nearly total resection surgery was performed, reported as the choice in this location to avoid the risk of neurovascular and hypothalamic complications. The biopsy result was histologically compatible with a teratoma-type germ cell tumor with both mature and immature components. Intracranial teratomas are rare tumors, little studied, with a predilection for the midline, with inconsistent radiological characteristics and distinctive histological features in mature and immature patients. Due to this, the spectrum is variable and the prognosis is uncertain.

***Keywords:** Pediatric Suprasellar Teratoma; Germ Cell; Cystic Tumor*

Introduction

In the sellar region, germinoma is the most frequent germ cell tumor, representing 61.5% of cases, followed by teratomas and mixed germ cell tumors. (4) Congenital intracranial tumors are rare diseases that comprise 0.4-3.1% of all intracranial tumors, and teratomas represent 9-30% of them. (10) In our Health Institute specialized in pediatric patients, there are few reported cases (2 cases recorded in 5 years).

Despite the fact that teratoma can manifest anywhere along the midline, most occur in the pineal region and only a few cases of pediatric sellar teratomas are described in the literature. (3). Clinical manifestations are usually correlated with the location and size of the tumor. (1)

Although the craniospinal axis is a rare location, there has been considerable interest in this subset due to clinical heterogeneity and challenges in treatment. (7) Gross total resection may confer a survival benefit and prevent the development of more severe clinical complications, and the intracranial location of the tumor decides the surgical approach. (6) During surgery it is of utmost importance that the hypothalamus is not injured, in case of hypothalamic tumor invasion, a radical total resection should not be the main goal of surgery, but rather a near-total resection. (3)

In this article we present a case report of a ten-year-old boy who presented with a mixed teratoma in the sellar region with suprasellar extension and provide a review of the literature, details of the findings to arrive at the diagnosis, treatment and follow-up.

Case Presentation

A 10-year-old male patient with no medical history presented 10 months before admission with a moderate headache, which was accompanied in the last month by episodes of generalized tonic-clonic seizures. He was taken by his father for medical attention, where he was asked to undergo a brain magnetic resonance imaging (MRI) study with contrast, revealing an intracranial mass in the suprasellar region associated with hydrocephalus. Suddenly, his consciousness deteriorated, leading to a comatose state, so an external ventricular shunt was placed at his hospital of origin. After that, he was transferred to this hospital due to his level of complexity. He was admitted in poor general condition and poor neurological status. Brain MRI identified a solid-cystic suprasellar lesion with defined edges measuring 33 x 22 x 20 mm, covering the suprasellar region entering the third ventricle and subsequently to the pineal region, with compression of the optic chiasma and supratentorial hydrocephalus, with a high suspicion of a probable craniopharyngioma (Fig. 1 and 2). Tumor markers showed a slight elevation of alpha-fetoprotein (AFP): 51.13 ng/ml and normal value of human chorionic gonadotropin (b-HCG): 1.20 mIU/mL, so there was little suggestion of the possibility of a germ cell tumor.

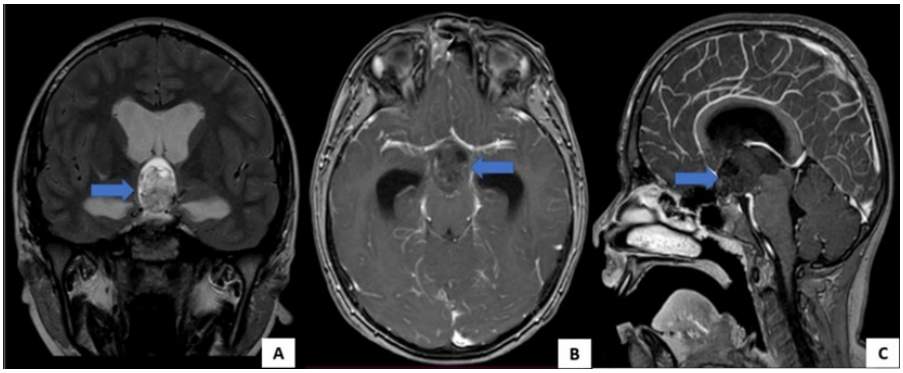

Figure 1. *Brain MRI (pre-surgical study) coronal (A), axial (B) and sagittal (C) section respectively shows a T2 sequence image of solid and cystic features at the suprasellar level with diameters of 33mm, 22mm and 20mm, in sequence Contrasted T1 shows minimal peripheral enhancement.*

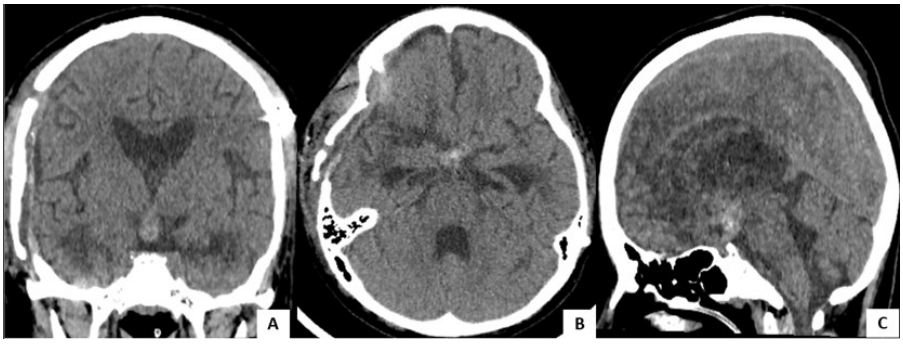

Figure 2. *Brain TEM (post-surgical study) in coronal (A), axial (B) and sagittal (C) sections, respectively, showing hyperdensity at the sellar level corresponding to small foci of bleeding.*

Intraoperative findings showed a tumor with a mostly solid consistency (85%) with few cystic areas with greenish content and a sparse sandy precipitate of a yellow-white color with regions of old bleeding strongly adhered to the nternal carotid and optic nerve with posterior extension to the upper portion of the midbrain. Total excision was achieved.

The anatomopathological result (Fig. 3 and 4) of the operative piece revealed mature teratoma in 95% and immature teratoma in 5%. Immunohistochemistry revealed S-100: positive, ki67: 30%. The immature component being the main risk for disease progression.

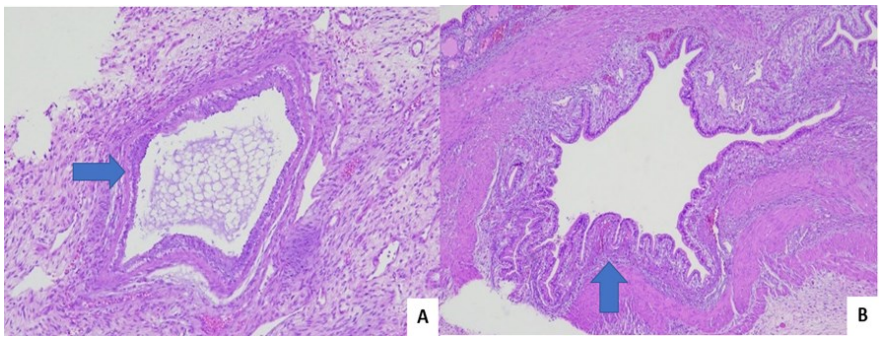

Figure 3. *Primitive-looking glandular elements covered by crowded columnar cells with clear subnuclear and apical cytoplasm (in imitation of the fetal intestine and respiratory mucosa), which suggests the diagnosis of Immature Teratoma A and B (HE, 10x).*

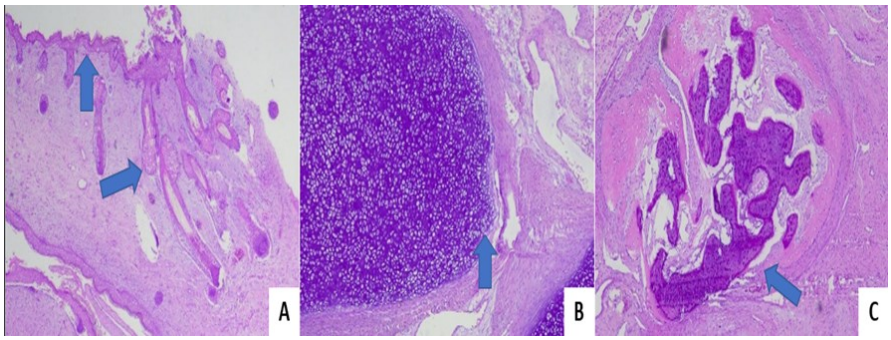

Figure 4. *Mature Elements: Keratinizing squamous epithelium with sebaceous glands and hair follicles A (HE, 2.5x), Islands of hyaline cartilage B (HE. 10x) and Bone tissue C (HE, 10x).*

During follow-up, the patient required a ventriculoperitoneal shunt, and panhypopituitarism was added, which worsened the prognosis. His state of consciousness improved slightly upon admission (Glasgow Coma Scale from 6 to 10 points), however, functionally he presented severe disability. The last brain tomography showed tumor recurrence 3 months after surgery.

Discussion

Intracranial germ cell tumors (IGCTs) are rare and account for 3% of all central nervous system (CNS) tumors in children and young adults. They arise primarily in the midline (3). Germ cell tumors of the sellar region often (6-13% of cases) present as synchronous lesions in association with pineal germinomas ("bifocal disease"). (4)

According to the WHO classification of CNS tumors, intracranial germ cell tumors comprise a heterogeneous group of neoplasms, divided into germinomas and non-germinomatous germ cell tumors (NGGTs). (6)

Intracranial teratomas are rare, and are usually found in the pineal region, followed by the suprasellar and hypothalamic regions. Teratomas comprise about 0.5% of all intracranial tumors. (9) Mature teratomas consist of fully differentiated elements derived from one or all three germ cell layers, whereas immature teratoma contains incompletely differentiated tissue. (3)

The composition of teratomas is distinctive, as they originate from all germ cell layers: endoderm, ectoderm, and mesoderm. Based on histological findings, teratomas are classified as mature, immature, and malignant, with incidences of 60%, 25%, and 15%, respectively. Immature teratomas consist of components resembling fetal tissue, often mixed with mature tissue elements. (5)

Teratomas are composed of a variable mixture of skin, skin appendages, smooth muscle, cartilage, and respiratory and gastrointestinal epithelium; Embryonic tissue may also be present (in an "immature" teratoma) and is neural in nature, often resembling a retinal bud. Due to the rapid growth of the tumor, the prognosis of teratomas is often poor, with a mortality rate of about 90%. (10) Both immature (fetal intestine and respiratory mucosa) and mature (keratinizing squamous epithelium with sebaceous glands and hair follicles) components were reported in the biopsy report.

The symptom complex of headache, bitemporal hemianopia, growth retardation, and diabetes insipidus strongly suggests the differential diagnosis of a sellar tumor, which is most commonly found in a craniopharyngioma but could be any other midline pathology in this region. (3) Our patient was admitted in poor general condition with symptoms of intracranial hypertension syndrome due to secondary acute hydrocephalus. The tumor per se was not clinically apparent.

Tumor markers such as β-hCG and AFP may be useful for the diagnosis of teratoma, as well as for distinguishing immature, mixed teratomas, and mature teratomas from immature or malignant components (2). Intracranial NGTs may show elevated serum and CSF markers, however, they are not usually elevated in mature teratomas. (3) No tumor marker study was reported in this case, as it was initially assumed to be a craniopharyngioma. However, there should always be clinical suspicion of a germ cell tumor.

Regarding radiographic characteristics, teratomas are mixed-density tumors, generally with cystic and solid components. In mature teratomas, teeth, fat, hair, or calcifications may occasionally be found. (3)

Surgery remains the first option for the treatment of intracranial immature teratomas. It is necessary to remove as much of the tumor as possible. (1)

Tumors originating in the sella turcica can be accessed either endoscopically endonasally transsphenoidally or via craniotomy depending on the extent of the tumor. In young children, the transsphenoidal approach may be more challenging due to incomplete pneumatization of the sphenoid sinus. (3) The patient underwent a pterional craniotomy with a transsylvian approach. 95% of the tumor was resected, minimizing hypothalamic manipulation.

Mature teratomas represent the only germ cell tumor entity that can be treated by resection alone, and they have a low recurrence rate. Other germ cell tumors, such as immature teratomas or mixed GCT teratomas, require multi-stage treatment including surgical resection, adjuvant or neoadjuvant chemotherapy and radiotherapy (3). The patient presented tumor recurrence of the immature component at 3 months with 1cm of growth. It was decided not to perform a reintervention and to follow the lesion with periodic images. A ventriculoperitoneal shunt was placed due to persistent hydrocephalus. His functional status was classified as ERm 5 points, his state of consciousness improved slightly reaching a Glasgow coma scale of 10 points.

Conclusions

A case was presented of a pediatric patient with mixed intracranial teratoma, a rare tumor with a benign and malignant spectrum, which leads to the analysis that the suspicion of germinal lesions of the midline is important, especially in the case of an immature teratoma. In this last case, the management becomes multidisciplinary, and the prognosis is grim. The surgical management is similar to other types of tumors of the sellar region, an almost total resection with minimal hypothalamic manipulation.

Conflict of Interest

The authors declare no conflict of interest.

Acknowledgement

None

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