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CASE REPORT

Giant intracranial teratoma in a pediatric patient: a case report and literature review

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Abstract

Purpose Primary intracranial non-germinomatous germ cell tumors, including teratomas, are rare in pediatrics and often pose diagnostic and therapeutic challenges due to their heterogeneous origin and histopathology. This case report aims to contribute to the literature by presenting a rare giant intracranial mature teratoma in a pediatric patient, highlighting its clinical presentation, radiological features, surgical management, and outcomes.

Methods We retrospectively reviewed the clinical, radiological, intraoperative, and histopathological data of a 4-year-old boy who presented with imbalance, gait disturbance, and lower extremity weakness. Preoperative serum tumor markers and neuroimaging (MRI and CT) were analyzed, followed by surgical resection and postoperative follow-up.

Results Imaging revealed a large heterogeneous tumor extending from the lateral and third ventricles to the cerebral hemispheres, characterized by cystic, fatty, and calcified components suggestive of teratoma. Serum AFP and β-HCG were normal. Complete tumor resection was achieved via a transcortical intraventricular approach. Histopathology confirmed a mature teratoma. Postoperatively, the patient exhibited transient motor weakness and decreased verbal response but returned to baseline within 1 year without neurological sequelae.

Conclusion Intracranial mature teratomas, although rare, can be successfully managed with complete surgical excision. Accurate radiological and pathological evaluation is crucial for diagnosis and treatment planning. Complete resection minimizes recurrence and complications, leading to favorable neurological outcomes in pediatric patients.

Keywords Dermoid cyst · Germ cell tumor · Mature teratoma

Abbreviations

AFP	Alpha-fetoprotein
B-HCG	Beta human chorionic gonadotropin
MRI	Magnetic resonance imaging
CT	Computed tomography

Introduction

The primary non-germinomatous germ cell tumors are rare neoplasms, constituting 1–3% of all pediatric intracranial tumors. They are histopathologically classified into five subgroups: embryonal carcinoma, yolk sac tumor, choriocarcinoma, teratoma, and mixed germ cell tumors. Teratomas, such as in this case, originate from three different embryonic germ cell layers and are predominantly localized in the midline structures, particularly from the pineal gland region of the cranium. However, as in our case, it may be uncertain whether the tumor originated from the midline extending to cerebral hemispheres or from hemispheres extending to midline structures. Because they may be localized outside the midline structures, intracranial teratomas are classified into three subgroups histopathologically: mature teratoma, immature teratoma, and teratoma with somatic-type malignancy. While mature teratomas are described with well-differentiated three germ layers, immature teratomas have more primitive components, and teratomas with somatic malignancy, which is the malignant transformation of a teratomatous component into a non-germ cell malignancy (rhabdomyosarcoma type). The immature teratoma exhibits a high degree of malignancy, rapid growth, and a poor prognosis, in contrast to the mature one. This case report covers a pediatric patient diagnosed with a giant intracranial mature teratoma. We aim to contribute to the literature for such extremely rare

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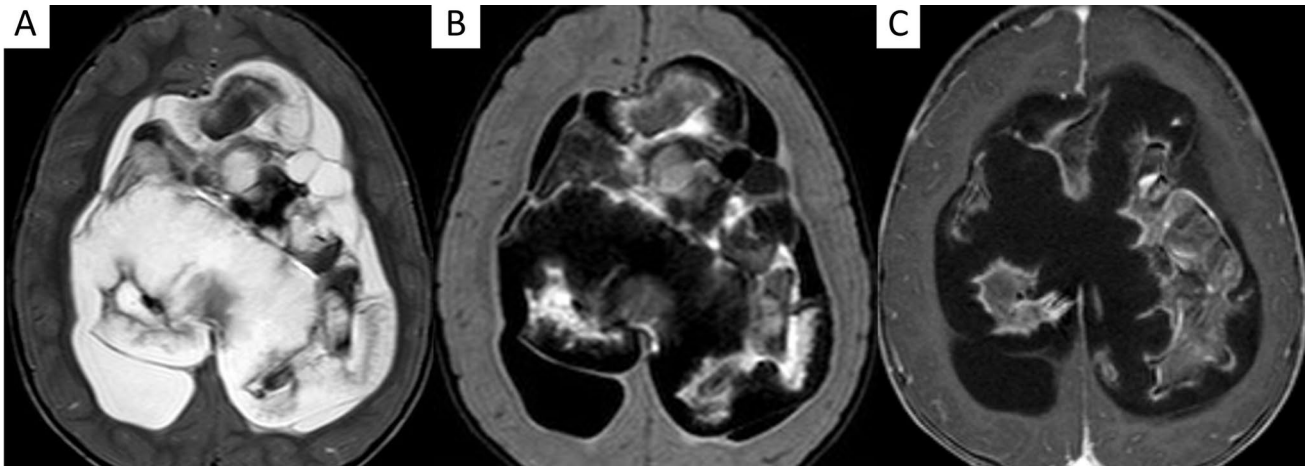


Fig. 1 **A** Axial T2-weighted and (**B**) T2-weighted FLAIR MR sequences demonstrating a giant intracranial mass extending from the lateral and third ventricles into the cerebral hemispheres, characterized by a hyperintense edematous appearance. **C** Axial gadolinium-enhanced T1-weighted MR sequence revealing a heterogeneous mass with cystic, fatty, and calcified components, consistent with the diagnosis of a teratoma

giant intracranial teratomas in terms of diagnosis, classification, pathological, clinical, and radiological features, and treatment and management.

Materials and methods

In this case, the complaints, clinical symptoms, neurological examination findings, radiological and intraoperative images, and histopathological diagnosis were examined by retrospective analysis from our clinic's electronic database.

Case report

A 4-year-old boy presented to our clinic with imbalance, gait disturbance, and motor weakness in the lower extremities. During preoperative investigations, AFP (alpha-fetoprotein) and β-HCG (human chorionic gonadotropin) were found in the normal range. On MRI (magnetic resonance imaging) and CT (computed tomography), a giant tumor extending from both the lateral and third ventricles to the cerebral hemispheres was seen. The radiological imaging was compatible with teratoma/dermoid cyst by hyperintense edematous appearance on T2/T2 FLAIR MRI and heterogenous contrast enhancement with cystic, fatty, and calcified components (Fig. 1). Following all investigations, surgical intervention was planned, and the tumor was completely resected (Fig. 2). During surgical resection by transcortical intraventricular approach, bones, cartilage, hair follicles, and adipose tissue components of the tumor were exposed in furtherance of the preoperative diagnosis (Fig. 3). Moreover, histopathologically it was reported as mature teratoma. During the postoperative period, the patient experienced approximately 40% motor weakness in all extremities and a decrease in verbal response, producing only incomprehensible sounds. The patient was discharged with recommendations for physical therapy and was monitored through routine follow-ups. By the end of the first postoperative year, neurological examination revealed no sequelae, and the patient returned to their preoperative baseline.

Discussion

Intracranial teratomas, in other words, dermoid cysts as a subtype of germ cell tumors, are rare, dysembryogenetic cysts that originate from ectodermal inclusions of primitive pluripotent cells [26]. These developmental abnormalities occurring between the third and fifth week of embryogenesis progress gradually and constitute approximately 0.5–1.5% of all infant brain tumors. So, many fetal cases have been reported in the literature, including both stillbirths and live births. Teratomas are predominantly localized in the midline, particularly in the cisternal space. Therefore, it is believed that they originate from the pineal gland, involving the third ventricle, and may cause hydrocephalus [1]. They may also be localized outside the midline structures, such as the basal ganglia and cerebellopontine angle [8]. Moreover, teratomas may appear as craniofacial lesions with intracranial extension. In the present review, we excluded fetal and neonatal cases diagnosed only by autopsy (6/45), cases of craniofacial lesions with intracranial extension (6/45), and cases with lesions located outside the lateral ventricles

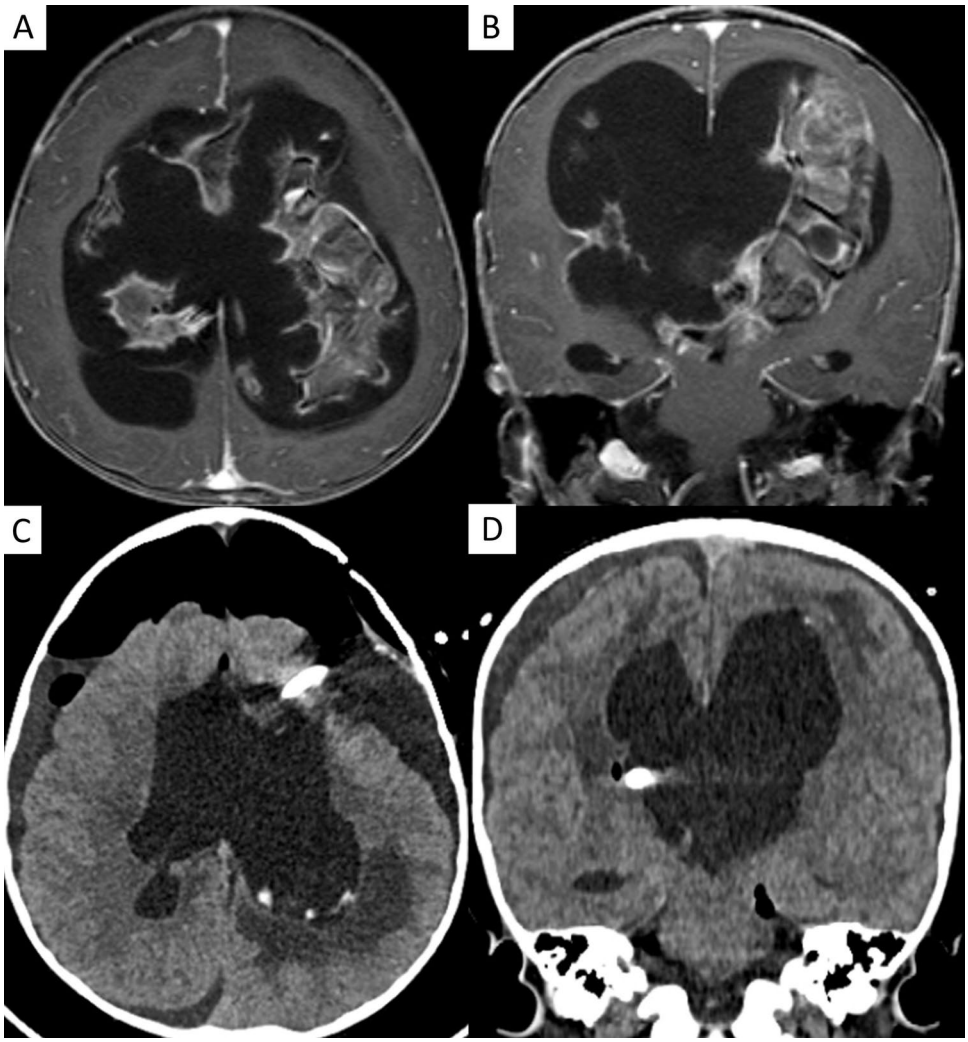


Fig. 2 **A** Preoperative gadolinium-enhanced axial T1-weighted and (**B**) coronal MR sequences demonstrating a giant intraventricular teratoma with heterogeneous contrast enhancement. **C** Postoperative axial and (**D**) coronal CT scans showing complete tumor resection via a transcortical intraventricular approach, along with the placement of a ventricular drainage catheter

(20/45) from a total of 45 reported pediatric intracranial giant teratomas. Ultimately, 13 cases, including the present one, were analyzed concerning demographic features, clinical presentation, treatment modalities, surgical approach, follow-up duration, neurological outcome, and prognosis (Table 1) [10, 12, 17, 20–22, 24, 27–29, 32]. In the present case, whether the tumor originated from the midline extending to cerebral hemispheres or originated from hemispheres extending to midline structures is uncertain. They can be pathologically identified by a capsule of simple epithelium underlayed by collagen. They comprise multiple tissues such as cartilage, bones, hair follicles, sebaceous glands, and apocrine glands [2]. Intracranial teratomas are classified into different subgroups regarding histopathological, radiological, and clinical features. Histopathologically, they can be examined as mature teratomas with well-differentiated three germ layers, immature teratomas with more primitive components, and teratomas with somatic malignancy, which is the malignant transformation of a teratomatous component into a non-germ cell malignancy (rhabdomyosarcoma type) [1, 13, 18]. Unlike the mature type, immature teratoma shows a high degree of malignancy, rapid growth, and poor prognosis [3, 30]. The information about immature teratomas' genetic and molecular etiopathogenesis is still insufficient. It is known that DNA methylation levels in mature and immature teratomas are similar, but there are some aberrant methylation levels in immature teratomas [19]. However, they can be classified into intact and ruptured cysts. Intact cysts grow insidiously, expand within the subarachnoid space, and envelop critical neurovascular structures [16]. It commonly presents with headaches, cranial nerve compression symptoms, and seizures [25]. Ruptured dermoid cysts may manifest with meningitis, acute hemiparesis, arachnoiditis, ventriculitis, hydrocephalus, and cerebral ischemia [4–6, 11]. Intracranial teratomas are diagnosed by radiological imaging such as computed tomography (CT) or MRI (magnetic resonance imaging) in addition to clinical symptoms. On MRI, they generally appear as



Fig. 3 Macroscopic appearance of the surgically resected tumor fragments, demonstrating components such as bone, cartilage, adipose tissue, and hair follicles

fatty tissue due to their liquid cholesterol content. But their intensity changes depending on fat content [2]. In a word, fatty components may be absent, or other elements such as hair or debris might predominate [9]. On CT, calcifications of the tumor can be seen easily. In addition to radiological imaging, serum levels of AFP (alpha-fetoprotein) and B-HCG (beta-human chorionic gonadotropin) help diagnose. The treatment of teratomas is complete surgical resection as far as possible, as well as adjuvant chemotherapy and radiotherapy, especially for immature teratomas. Thus, the potential complications can be prevented due to rupture or growth, like meningitis, dissemination, or compression on adjacent structures [31]. However, in some cases, the cyst may adhere strongly to adjacent critical structures, and consequently, residual lesions may be left. After these subtotal resections, the recurrence risk increases, resulting in more surgical interventions. So, complete surgical resection whenever feasible is a milestone for managing intracranial teratomas [4, 14, 23, 25, 31]. The prognosis of teratomas is associated with the degree of tumor differentiation because undifferentiated embryonic tissues have a worse prognosis [7, 15].

Conclusion

Radiological and hormonal examinations are essential to diagnose rarely observed intracranial teratomas. However, there is no standard treatment and management algorithm because of the different tissue components and low sensitivity of non-germinomatous germ cell tumors to chemoradiotherapy. But after surgical resections, the recurrence risk of the tumor and the possibility of complications during the normal clinical course decreased. As a result, complete surgical resection whenever feasible is critically essential for managing intracranial teratomas. In addition, they generally show a good prognosis after surgery and chemoradiotherapy in comparison to untreated controls.

Table 1 Demonstrating studies on giant ıntraventricular teratomas in children, with an extensive review of the literature

Author and year	Age/sex	Location	Clinical presentation	Surgical approach/extent of resection	Treatment	Histological features	Adjuvant therapy	Length of follow-up	Neurological outcome	Prognosis
Ventureyra et al. (1983)	6 d, M	Right lateral ventricle, 3rd ventricle	Macrocephaly	Frontoparietal craniotomy, intraventricular, total resection	Surgery	Teratoma	-	4 months	SIADH and seizures (both controlled), increased ICP (resolved with/VPS)	Good (no recurrence)
Oi et al. (1989)	37 wk premature infant, F	Lateral and 3rd ventricles	Preterm C-section, macrocephaly, hydrocephalus	Bifrontal craniotomy, transcortical, subtotal resection	Surgery	Teratoma	CT	7 months	Developmental delay	Intermediate
Nanda et al. (1991)	6 d, F	Extensive involvement	Macrocephaly	Temporal, subtotal resection	Surgery	Teratoma	-	3 days	Expired	Poor (mortality)
Selcuki et al. (1998)	8 y, M	Right lateral ventricle	H/A, N/V, coma, anisocoria, decorticate posturing, papilledema	Parietooccipital craniotomy, transcortical, total resection	Surgery	Teratoma	-	24 months	Sensorimotor aphasia	Good (no recurrence)
Selcuki et al. (1998)	2.5 mo, F	Lateral and 3rd ventricles	Nausea, bilateral CN VI palsy, papilledema	Frontal craniotomy, transcortical intraventricular, total resection	Surgery	Teratoma	-	12 months	Bilateral CN VI palsy	Good (no recurrence)
Sinha et al. (2001)	2 mo, M	Left lateral ventricle, trigone	Macrocephaly, seizures	Temporoparietal craniotomy, transcortical, near total resection	Surgery	Teratoma	-	2 years	Bilateral subdural effusion	Good (no recurrence)
Phi et al. (2005)	20 d, F	Extensive involvement (intraventricular)	Macrocephaly, hydrocephalus lethargy	Transcortical, intraventricular, total resection	Surgery (×2)	Teratoma	-	76 months	Developmental delay, mental retardation	Good (no recurrence)
Erman et al. (2005)	3 d, F	Extensive involvement (right parietooccipital and lateral ventricles)	N/V, seizure	Parietooccipital craniotomy, total resection	Surgery	Teratoma	-	3 weeks	No postop complication, ex (pneumonia)	Poor (mortality)

Table 1 (continued)

Author and year	Age/sex	Location	Clinical presentation	Surgical approach/extent of resection	Treatment	Histological features	Adjuvant therapy	Length of follow-up	Neurological outcome	Prognosis
O'Grady et al. (2012)	16 y, F	Lateral ventricles	H/A, N/V, dizziness, disorientation, seizure	Transcallosal, subtotal resection	Surgery	Teratoma	-	6 months	Postoperative hydrocephalus (resolved with/VPS); improved dysphasia, short-term memory and mobilization with/rehabilitation	Good (no recurrence)
Li et al. (2014)	3 y, F	Right lateral ventricle	H/A	Frontal craniotomy, total resection	Surgery	Teratoma	-	?	?	Good (no recurrence)
Zhu et al. (2016)	11 mo, M	Lateral and 3rd ventricles	Lower limb weakness, hydrocephalus	Transcallosal interforniceal, total resection	Surgery	Teratoma	-	2–5 months	No postop complication	Good (no recurrence)
Demir et al. (2020)	17 y, M	Lateral and 3rd ventricles, leptomeningeal dissemination	H/A, N/V, obstructive hydrocephalus	Interhemispheric transcallosal, subtotal resection	Surgery	Teratoma (mixed germ cell tumor)	CT/RT	5 months	No postop complication	Good (no recurrence)
Present case	4 y, M	Lateral and 3rd ventricles	Imbalance, gait disturbance, lower limb weakness	Transcortical intraventricular, total resection	Surgery	Teratoma	-	18 months	No neurological deficit	Good (no recurrence)
Total	13 patients									

CN cranial nerve, *CT* chemotherapy, *H/A* headache, *ICP* intracranial pressure, *N/V* nausea–vomiting, *SIADH* syndrome of inappropriate antidiuretic hormone, *RT* radiotherapy, *VPS* ventriculoperitoneal shunt

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Data availability No datasets were generated or analysed during the current study.

Declarations

Conflict of interest None declared.

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