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Immature Intracranial Teratoma in Childhood - A Rare Clinical Case from Our Practice

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ABSTRACT

Immature teratomas are rare intracranial malignant tumors of childhood, predominantly localized in the pineal or suprasellar brain region. We present a rare clinical case of a 12-year-old boy with an immature intracranial teratoma who underwent apparently total tumor excision followed by local cranial radiotherapy. The presented clinical case focuses on the diagnosis and complex treatment of these rare non-germ cell brain tumors in childhood. A strict pathohistological diagnosis, serum and cerebrospinal fluid tumor marker testing (AFP and HCG), and MRI of the craniospinal axis are required.

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Introduction

Germ cell tumors (GCTs) are sporadic, constituting 0.3-0.6% of all primary central nervous system (CNS) tumors. They are categorized into germinomatous, nongerminomatous, and subtypes. Teratomas, usually delineated as a separate category, make up only 18-20% of all GCTs [1-3]. Intracranial teratoma usually found in young patients, accounting for about 0.3%–0.6% of all primary intracranial tumor and 0.5%–1.5% of all childhood brain tumors [4]. Intracranial teratomas, like other GCTs, have a predilection for the pineal and suprasellar regions [1,5]. They are more common in males than in females [6,7]. We present a rare clinical case of a 12-year-old boy with an immature intracranial teratoma who underwent apparently total tumor excision followed by local cranial radiotherapy (RT).

Clinical Case

This concerns a 12-year-old boy with complaints of headache, nausea and vomiting dating back to July 2024. An MRI performed in August 2024 revealed a tumor formation in the area of the third cerebral ventricle with evidence of occlusive hydrocephalus. MRI of the head - In the area of the dorsal part of the third ventricle and cranially from the mesencephalon, an oval tumor with a heterogeneous structure is visualized with the presence of solid and cystic components and calcifications. The cystic components have a high colloidal content with liquid levels. After intravenous administration of contrast material, the finding increases its signal intensity inhomogeneously. The lesion has axial dimensions of 31.5 mm/26.5 mm. Adjacently, it has a mass effect on the brain parenchyma and mesencephalon. Proximally, the ventral part of the third ventricle and both lateral ventricles are dilated with evidence of transependymal resorption. The basal cisterns are narrowed, the perineural spaces around both optic nerves are dilated. Pons, medulla oblongata and cerebellum have normal MRI images. Fourth ventricle is not dilated (Figure 1). The child was hospitalized in a neurosurgery clinic, where a ventriculoperitoneal shunt was placed at the first stage. After one week, due to visible growth of the lesion due to the cystic component to dimensions of 39.4 mm/ 31.3 mm, a visible total tumor excision was performed. Intraoperatively – A tumor with macroscopic characteristics of a germinoma was found, which was visibly totally excised. Histopathological examination / September 2024- Multiple materials with sizes of 1 to 20 mm were reported, in which a tumor was composed of structures of different pathogenetic origin - connective / presence of cartilage / and epithelial / glandular structures. Immunohistochemistry- Ki 67- 50% to 100% in some of the structures- The histological picture and the high proliferative activity point to the final diagnosis of immature teratoma. The child is referred to the Clinic of Pediatric Clinical Hematology and Oncology for assessment of the subsequent complex treatment. An MRI of the craniospinal axis was performed without any reliable evidence of residual tumor or leptomeningeal metastases. A lumbar puncture did not reveal tumor cells in the cerebrospinal fluid, nor did elevated tumor markers bHCG and AFP.

Given the histological result for an immature teratoma with high proliferative activity Ki 67- 50% to 100%, as well as the apparently total tumor extirpation performed, due to the high risk of recurrence, it was considered that the child should undergo postoperative local radiotherapy. In October 2024, the boy underwent postoperative intensity modulated radiotherapy (IMRT) using the VMAT technique in the area of the tumor bed with a daily dose (DD) of 2.16 Gy up to total dose (TD) of 54 Gy, as well as in the area of the lateral cerebral ventricles with a DD of 1.84 Gy up to total dose TD of 46 Gy (Figure 2 and Figure 3). The child tolerated the radiotherapy (RT) well, without neurological side effects. After 3 months, a control MRI of the brain was performed, which did not reveal pathological changes (Figure 4). Figure 5 shows the comparison of the MRI image 3 months after the completion of local IMRT with the MRI image before the operation.

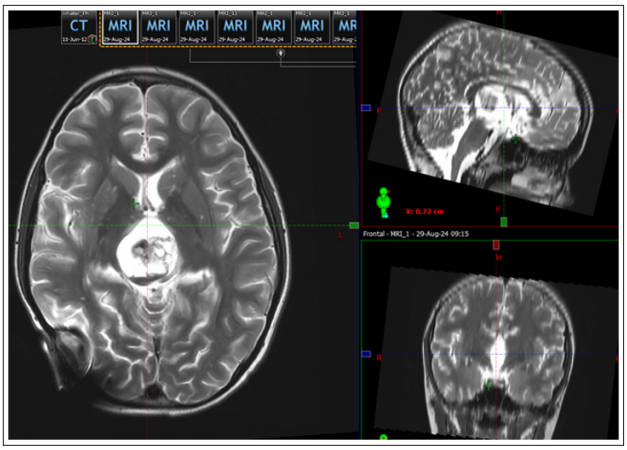

Figure 1: Cerebral MRI/ August 2024 - In the Area of the Dorsal Part of the Third Ventricle and Cranially from the Mesencephalon, an Oval Tumor Lesion with a Heterogeneous Structure is Visualized with the Presence of Solid and Cystic Components and Calcifications. The Cystic Components have a High Colloidal Content with Liquid Levels. After Intravenous Administration of Contrast Material, the Finding Increases its Signal Intensity Inhomogeneously.

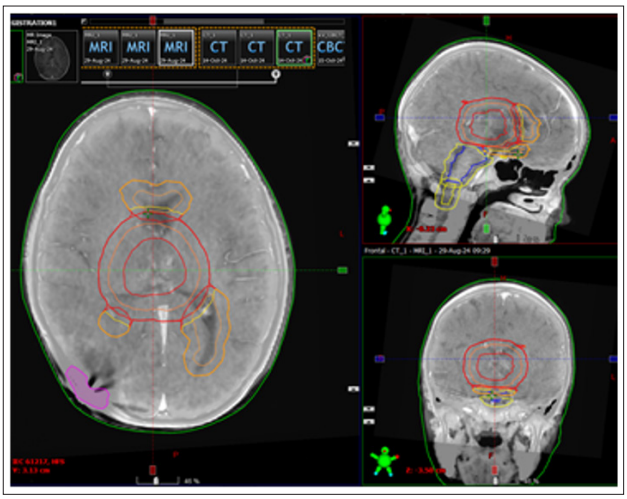

Figure 2: Delineation of the Clinical Tumor Volumes CTVp in the Area of the Primary Tumor and CTV ventr. in the Area of the Lateral Ventricles after Fusion of the Primary Preoperative MRI with the Planning CT.

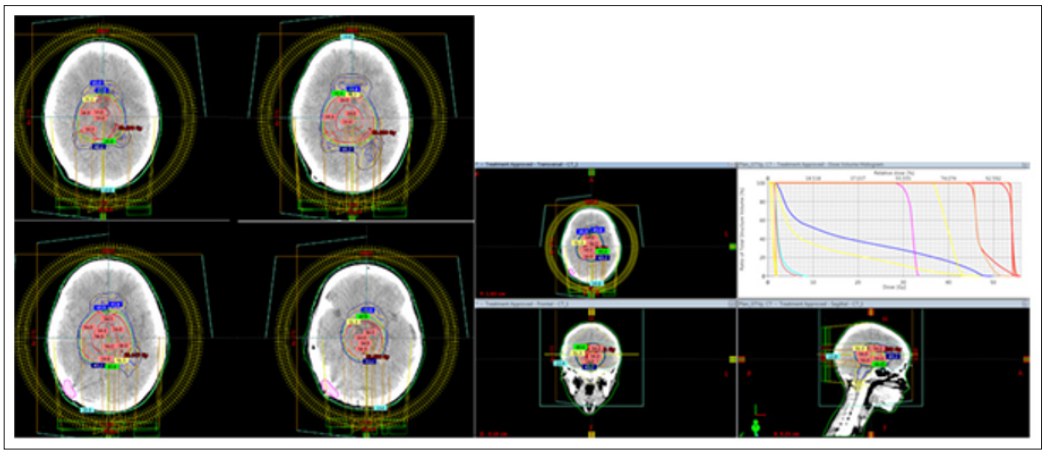

Figure 3: Postoperative IMRT using the VMAT Technique in the Area of the Tumor Bed /CTVp/ with a Daily Dose (DD) of 2.16 Gy up to Total Dose (TD) of 54 Gy, as well as in the Area of the Lateral Cerebral Ventricles / CTVventr./ with a DD of 1.84 Gy up to Total Dose TD of 46 Gy.

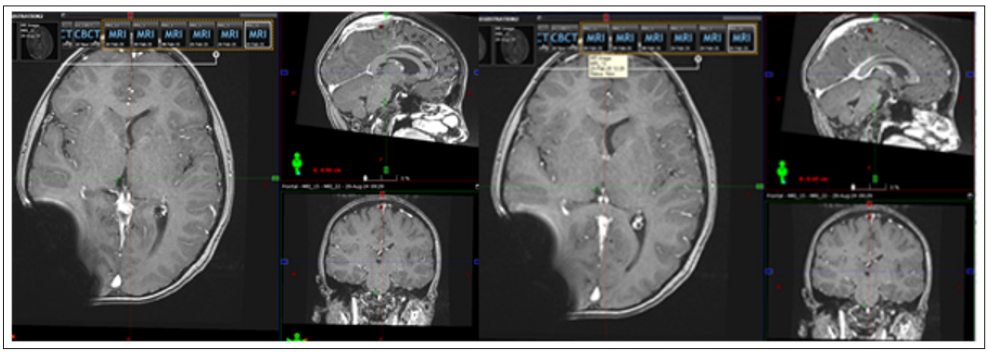

Figure 4: MRI from February 2025, 3 Months after the Completion of Local IMRT- Supratentorially and Infratentorially without Focal Changes. Ventricular System with Reduced Volume of the Right Lateral Ventricle on the Background of a Shunt. Normal width of the Third and Fourth Ventricle without Signs of Hydrocephalus. The Remaining Brain Structures without Changes.

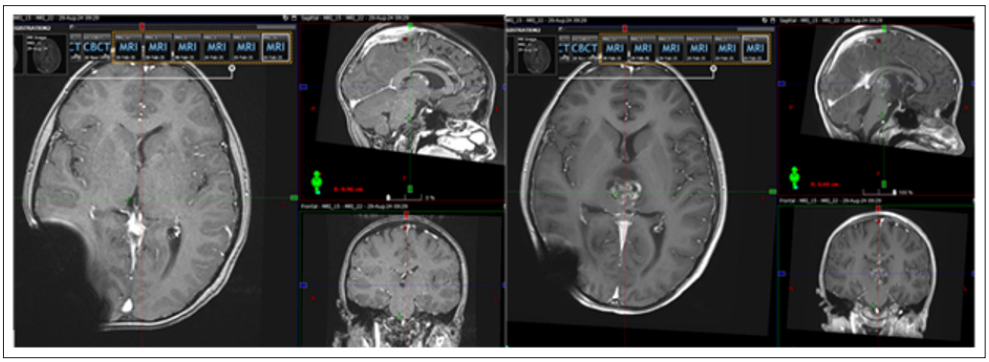

Figure 5: The Comparison of the MRI 3 Months after the Completion of Local IMRT with the MRI Image before the Operation.

Discussion

Intracranial GCT are a heterogenous group of rare cerebral neoplasms, including 5 different histological subtypes of increasingly malignant behavior: germinomas, teratomas, embryonal carcinomas, yolk sac tumors, and choriocarcinomas combined within mixed tumors in 25–32% of cases [8-11]. Approximately 90% of GCTs occur in patients younger than 20 years old, with a peak incidence at 11 years of age [12-14]. Nongerminomatous GCT's present as posterior third ventricular masses with hydrocephalus and midbrain compression [15]. Only germinomas and teratomas are likely to be encountered as pure tumor types [16,17].Immature teratomas, being composed of more primitive tissue, have a greater potential for malignant transformation and not surprisingly have lower survival rates within the 50%-70% range [18,19]. An immature teratoma is a germinal malignant tumor that is composed of three germ cell layers (the ectoderm, endoderm, and mesoderm), and it is histologically characterized by immature tissue, most frequently, neuroepithelial tissue [20]. In the presented clinical case, the tumor consists of various tissue structures, including connective and cartilage tissue, as well as glandular structures with high proliferative activity Ki 50-100%, which defines it as an immature teratoma with high malignant potential.

An immature teratoma is the only neoplasm with germ cells that are histologically graded depending on the immature neural elements, and this is a prognostic factor for overall survival [20]. Immature neuroectodermal tissues include islets and nests of neuroblasts, hypercellular and mitotically active immature glia, and primitive, melanic pigmented retinal tissue [21]. Based on quantity of immature neuroepithelium tumor cell differentiation is defined as: G0 - mature teratoma; no immature neuroepithelium; G1 - less than one lower power field (LPF) of immature neuroepithelium; LPF defined field at 4X magnification ; G2 - 1-3 LPFs ; G3 - more than 3 LPFs [22].

Intracranial teratomas, being a rare subtype of already rare intracranial GCTs, pose a set of diagnostic and treatment challenges [23]. The risk factors associated with a worse prognosis in cases of non-germinomatous GCT's are high alpha-protein levels (>1000 ng/ml) and residual tumor after all lines of treatment instituted (primary or second-look surgery, chemotherapy and radiotherapy [24]. The presence of metastasis and the absence of radiotherapy were factors associated with a worse prognosis [25]. Serum/CSF tumor markers can be measured to aid in diagnosis; yolk sac tumors secrete AFP, choriocarcinomas secrete human chorionic gonadotrophic hormone (HCG), and teratomas can secrete both AFP and HCG [23]. In the presented immature teratoma, we do not report high cerebrospinal fluid levels of tumor markers AFP and HCG.

On CT imaging of teratomas, calcification, cystic components, and/or fat are often noted [26]. As with all intracranial tumors, imaging requires MRI. Pineal region tumors have no pathognomonic imaging pattern. MRI and CT are complementary in diagnosis and are important to determine localization, extension, and meningeal spread [27].

On MRI, GCTs are described to be hypo- to isointense on T1W images and hyperintense on T2-weighted (T2W) images; it should be noted though that teratomas cannot always be reliably differentiated from other GCTs on imaging alone, including MRI [5,26]. All nongerminomatous GCT showed heterogeneous enhancement [28]. The immature teratomas and teratomas with malignant transformation demonstrated heterogenous, ring-like, intratumoral patchy enhancement on T1W images with contrast

[4]. MRI features tumours can facilitate correct diagnosis of GCT, including histological subtypes. After surgery the presence of even a small portion of immature or malignant tissue can predispose to recurrence [3]. In the dorsal part of the third ventricle and cranially from the mesencephalon, the MRI from August 2024 in the presented clinical case, reported an oval tumor with a heterogeneous structure with the presence of solid and cystic components and calcifications. The cystic components have a high colloidal content with liquid levels. After intravenous administration of contrast material, the finding increases its signal intensity inhomogeneously (Figure 1).

Immature teratoma is a malignant tumor requiring multimodal therapy for the best patient outcome [6,7,29]. Surgery may be performed as a first-line therapy when the results of noninvasive diagnostic investigations suggest the teratoma presence or after complex oncological therapy for removal of residual tumor, improving the prognosis of patients with mixed malignant GCTs [25]. Open surgery is important for all suspected teratoma since they frequently have mixed portion on different histologic tumors Complete resection of tumors and adjuvant chemoradiotherapy are known to have the highest 5-year survival rate, as presented in numerous previous case series and reports [30-34]. Non-germinoma malignant GCTs are best treated with neoadjuvant chemotherapy followed by RT given focally for local disease; craniospinal radiation therapy (CSRT) is reserved for patients with evidence of disseminated disease at the completion of induction chemotherapy [35]. Given the histological result for an immature teratoma with high proliferative activity Ki 67- 50% to 100%, as well as the apparently total tumor extirpation performed, due to the high risk of recurrence, it was considered that the child should undergo postoperative local RT (Figure 4). Due to the tumor localization in the area of the third ventricle, as well as the high malignant tumor potential with a high risk of cerebrospinal fluid metastases, in addition to the tumor bed, we also irradiated the lateral ventricles in order to prevent subsequent cerebrospinal fluid metastases (Figure 2). Figure 5 presents the MRI three months after completion of radiotherapy, which reported local tumor control. In such clinical cases, prolonged monitoring using MRI is required at six-month intervals, not only of the central nervous system, but also of the spinal axis.

Conclusion

Immature intracranial teratoma is a rare non-germ cell malignant brain tumor of childhood. Its localization primarily involves axial brain structures, namely the pineal and suprasellar regions. A strict pathohistological diagnosis, serum and cerebrospinal fluid tumor marker testing (AFP and HCG), and MRI of the craniospinal axis are required. In localized disease, maximally radical surgery is performed, followed by chemotherapy and/or local RT. Prolonged monitoring using MRI is required at six-month intervals, not only of the central nervous system, but also of the spinal axis.

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