Intracranial teratoma is an extremely rare disease that accounts for approximately 0.3%–0.6% of all primary intracranial tumors [1]. Moreover, intracranial immature teratomas are much rarer, malignant, and have a poor prognosis, requiring complicated treatments [1-6]. Due to its histologic nature, it is mostly located in the deep midline of the intracranial hemisphere from the pineal gland to the suprasellar area [1,3-5,7-9]. Therefore, the tumor may cause various complications, and the deep location of the tumor makes surgery challenging, resulting in incomplete resection in many cases, and requiring multimodal treatments [10].

We present our experience with a fast-growing pineal gland immature teratoma in a 4-year-old child, who presented with obstructive hydrocephalus and abducens nerve palsy, which was treated with total surgical resection of the tumor. In addition, we aimed to determine the appropriate treatment modality for intracranial immature teratomas by reviewing the literature and investigating the prognosis.

**Keywords**  
Immature teratoma; Brain neoplasms; Hydrocephalus.

**CASE REPORT**

A 4-year-old boy, who presented with drowsiness after a general tonic-clonic seizure, was admitted to the emergency department of our institute. Brain CT showed acute obstructive hydrocephalus and associated diffuse interstitial edema caused by a tumor in the pineal gland region, blocking the cerebral aqueduct (Fig. 1A). The patient showed drowsy mentality with lateral gaze impairment of right eye, indicating abducens nerve palsy. An emergency extra-ventricular drainage catheter insertion surgery was performed as a temporary measure due to the persistent drowsiness. The patient’s mental status recovered after drainage but the abducens nerve palsy did not. A brain MRI was performed to identify the tumor and it demonstrated a 2.5-cm-sized pineal gland tumor consisting of heterogeneous enhancement with calcific foci and cysts (Fig.
There were no remarkable changes in serum alpha-fetoprotein (AFP) and beta human chorionic gonadotrophin (bHCG) levels.

The patient underwent an endoscopic third ventriculostomy with tumor biopsy. Histopathological examination revealed a mature teratoma with cystic epithelium surrounded by mesenchymal fibrotic tissue.

The patient was determined to undergo surgical resection of the tumor. A repeated MRI for preoperative evaluation revealed improved hydrocephalus, but a much increased solid portion of the tumor, which was 3.5 cm in size, compared to 2.5 cm at the initial scan 2 weeks prior, indicating a faster than expected growth (Fig. 2). The patient underwent surgery using the occipital transtentorial approach, and gross total resection of the tumor was performed (Fig. 3). The symptoms including seizure and abducens nerve palsy resolved after the total removal of the tumor (Fig. 4). The final histopathological diagnosis revealed primitive neuronal epithelial tissue, indicating immature teratoma (Fig. 5). Standard chemotherapy was subsequently initiated (Table 1) [10], and no recurrence was observed.
Fig. 2. Preoperative sagittal (A) and axial (B) MRI showing improved hydrocephalus after endoscopic third ventriculostomy but much increased solid portion of the tumor.

Fig. 3. Intraoperative microscopic view and postoperative MRI. A: Intraoperative microscopic image after retracting the right occipital lobe. The tumor (black arrow) anterior to deep venous system (white arrow) was noted. B: Microscopic view after gross total removal of the tumor. Right occipital lobe (black arrow), deep venous system (white arrow), and incised tentorium (white arrowhead). C and D: Postoperative MRI demonstrating no residual tumor.
A Case of Intracranial Immature Teratoma

Intracranial teratoma is a rare type of non-germinomatous germ cell tumor (GCT), 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 [8]. Among the three histological types of intracranial teratoma (mature, immature, and teratoma with somatic-type malignancy), immature teratoma is known as the highly malignant, fast-growing intracranial tumor, thus presenting poor outcome and challenging for clinical diagnosis and treatment [1,3–5,8–10]. The tumor is usually located at the midline of the intracranial hemisphere, similar to other GCTs, and MRI findings of heterogeneous enhancement, mixed signals due to different tissues, intratumoral cyst, calcifications, and fat signals may indicate teratoma. There are some rare case reports of teratomas found in non-midline locations such as the lateral ventricles, posterior fossa, basal ganglia, and cavernous sinus [8,11–14]. Serum AFP and bHCG are the most commonly used tumor markers in GCTs. Although it may not be accurate, elevated serum AFP may suggest a higher likelihood of immature teratomas than mature teratomas [8,9,11].

It is a well-known fact that the most important feature for predicting prognosis is the histological type of the malignant GCT [4–6,9,15]. Open surgery is important for all suspected teratoma since they frequently have mixed portion on different histologic tumors [10]. The endoscopic approach may be a great option due to the ability to achieve tissue access under good visual control and the option to perform additional third ventriculostomy to resolve obstructive hydrocephalus caused by the tumor [9].

Many pathological conditions may be complicated by the deep midline location of the tumor. Obstructive hydrocephalus and increased intracranial pressure due to obstruction of the cerebrospinal fluid (CSF) tract, such as the foramen of Monro and cerebral aqueduct, cranial nerve palsies, hearing or visual problems due to compression of the tectum of the midbrain or optic chiasm, and endocrine problems due to hypothalamic

| Table 1. Details of the chemotherapy applied [10] |
|-----------------------------------------------|
| Chemotherapy regimen* | Dose |
| A course | | |
| Carboplatin | 450 mg/m³, day 1–2 |
| Etoposide | 150 mg/m³, day 1–3 |
| Bleomycin | 15 mg/m³, day 3 |
| B course | | |
| Cyclophosphamide | 2,000 mg/m³, day 1–2 |
| Etoposide | 150 mg/m³, day 1–3 |
| Bleomycin | 15 mg/m³, day 3 |

*A/B/A/B, every 3 weeks

observed on the 6-month postoperative MRI scan.

DISCUSSION

Intracranial teratoma is a rare type of non-germinomatous germ cell tumor (GCT), 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 [8]. Among the three histological types of intracranial teratoma (mature, immature, and teratoma with somatic-type malignancy), immature teratoma is known as the highly malignant, fast-growing intracranial tumor, thus presenting poor outcome and challenging for clinical diagnosis and treatment [1,3–5,8–10]. The tumor is usually located at the midline of the intracranial hemisphere, similar to other GCTs, and MRI findings of heterogeneous enhancement, mixed signals due to different tissues, intratumoral cyst, calcifications, and fat signals may indicate teratoma. There are some rare case reports of teratomas found in non-midline locations such as the lateral ventricles, posterior fossa, basal ganglia, and cavernous sinus [8,11–14]. Serum AFP and bHCG are the most commonly used tumor markers in GCTs. Although it may not be accurate, elevated serum AFP may suggest a higher likelihood of immature teratomas than mature teratomas [8,9,11].

It is a well-known fact that the most important feature for predicting prognosis is the histological type of the malignant GCT [4–6,9,15]. Open surgery is important for all suspected teratoma since they frequently have mixed portion on different histologic tumors [10]. The endoscopic approach may be a great option due to the ability to achieve tissue access under good visual control and the option to perform additional third ventriculostomy to resolve obstructive hydrocephalus caused by the tumor [9].

Many pathological conditions may be complicated by the deep midline location of the tumor. Obstructive hydrocephalus and increased intracranial pressure due to obstruction of the cerebrospinal fluid (CSF) tract, such as the foramen of Monro and cerebral aqueduct, cranial nerve palsies, hearing or visual problems due to compression of the tectum of the midbrain or optic chiasm, and endocrine problems due to hypothalamic

Fig. 4. Pre- and postoperative eyeball motion of the patient. A: Preoperative eyeball motion of the patient presenting right abducens nerve palsy. B: Postoperative eyeball motion presenting resolution of the palsy.

Fig. 5. Histologic findings of the tumor (H&E stain, ×100). A: Immature cartilage with high cellularity is seen. B: Primitive neuronal epithelial tissue and multilayered neural crest forming tissue are seen, suggesting immature teratoma.

Table 1. Details of the chemotherapy applied [10]

| Chemotherapy regimen* | Dose |
|-----------------------|------|
| A course              |      |
| Carboplatin           | 450 mg/m³, day 1–2 |
| Etoposide             | 150 mg/m³, day 1–3 |
| Bleomycin             | 15 mg/m³, day 3 |
| B course              |      |
| Cyclophosphamide      | 2,000 mg/m³, day 1–2 |
| Etoposide             | 150 mg/m³, day 1–3 |
| Bleomycin             | 15 mg/m³, day 3 |

*A/B/A/B, every 3 weeks
or hypophysis dysfunction may be consequences of the tumor location. Interestingly, our case presented with a very fast growing tumor, which enlarged from 2.5 cm to 3.5 cm over 2 weeks. The patient had abducens nerve palsy, which did not resolve after drainage of CSF but resolved after the total resection of the tumor. The cause may be the pathologic stretching of the nerve as a result of the linear force of the mass effect due to the rapidly growing tumor above the midbrain, and not from the hydrocephalus [16]. Although the complications can be treated by surgical decompression and resection of the tumor, the overall 5-year survival rate of immature teratomas is poor according to the published literature [1,4-6,15]. This is thought to be the result of the malignant histologic nature of the tumor, high recurrence rate, and poor complete resection rate of the surgery due to the tumor's deep and vascular-rich location [4-6,15]. 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 [4,6,9,11]. Huang et al. [4] demonstrated a significant increase in the survival curve of patients who underwent gamma knife surgery after partial removal of immature teratomas. The details of the reported cases in the published literatures are summarized in Table 2. However, these results involved only a small number of cases and some are included as part of studies on intracranial GCT, since immature teratomas are extremely rare. Moreover, some case reports and case series have reported relatively higher survival rates (70%–100%) when total surgical resection of the tumor is performed [6,9]. Larger studies should be performed to determine the exact outcome and most effective treatment of the pathology.

In conclusion, we present a rare case of a fast-growing pineal gland immature teratoma in a 4-year-old child, which was successfully treated with complete surgical resection of the tumor through an occipital transtentorial approach. Intracranial immature teratomas may present with various serious pathologic symptoms and are difficult to treat due to the deep location and highly malignant nature of the tumor. Complete surgical removal and adjuvant chemoradiotherapy are the effective treatments of choice for this pathology.

### Ethics Statement

This study was approved by the Institutional Review Board of Keimyung University Dongsan Medical Center (2022-02-049). Written informed consent was obtained from the patient for publication of this Case Report and any accompanying images.

### Availability of Data and Material

The datasets generated or analyzed during the study are available from the corresponding author on reasonable request.

### ORCID iDs

Seung-Bin Woo [https://orcid.org/0000-0002-3038-1364](https://orcid.org/0000-0002-3038-1364)  
Chang-Young Lee [https://orcid.org/0000-0001-6046-2571](https://orcid.org/0000-0001-6046-2571)  
Chang-Hyun Kim [https://orcid.org/0000-0001-5401-5660](https://orcid.org/0000-0001-5401-5660)  
Young San Ko [https://orcid.org/0000-0002-6668-0905](https://orcid.org/0000-0002-6668-0905)
A Case of Intracranial Immature Teratoma

El Kim  
https://orcid.org/0000-0002-7664-6030
Ye Jee Shim  
https://orcid.org/0000-0002-5047-3493
Sang Pyo Kim  
https://orcid.org/0000-0003-0948-2408
Sae Min Kwon  
https://orcid.org/0000-0001-9720-6037

Author Contributions
Conceptualization: Sae Min Kwon. Data curation: Seung-Bin Woo. Funding acquisition: Sae Min Kwon. Investigation: Young San Ko, El Kim. Methodology: Chang-Hyun Kim. Project administration: Chang-Young Lee. Resources: Ye Jee Shim, Sang Pyo Kim. Supervision: Sae Min Kwon. Writing—original draft: Seung-Bin Woo. Writing—review & editing: Sae Min Kwon.

Conflicts of Interest
The authors have no potential conflicts of interest to disclose.

Funding Statement
This work was supported by the National Research Foundation of Korea (NRF) grant funded by the Korean government (MSIT) (No. 2020R1G1A1013289).

REFERENCES
1. Goyal N, Kakkar A, Singh PK, Sharma MC, Chandra PS, Mahapatra AK, et al. Intracranial teratomas in children: a clinicopathological study. Childs Nerv Syst 2013;29:2035–42.
2. Georgiu C, Opincariu I, Cebotaru CL, Mirescu ŞC, Stănoiu BP, Domşa TA, et al. Intracranial immature teratoma with a primitive neuroectodermal malignant transformation - case report and review of the literature. Rom J Morphol Embryol 2016;57:1389-95.
3. Abdelmuhdi AS, Almazam AE, Dissi NA, Albastaki UM, Pierre-Jerome C. Intracranial teratoma: imaging, intraoperative, and pathologic features: AIRP best cases in radiologic-pathologic correlation. Radiographics 2017;37:1506-11.
4. Huang X, Zhang R, Zhou LF. Diagnosis and treatment of intracranial immature teratoma. Pediatr Neurosurg 2009;45:354-60.
5. Lee YH, Park EK, Park YS, Shim KW, Choi JU, Kim DS. Treatment and outcomes of primary intracranial teratoma. Childs Nerv Syst 2009;25:1581-7.
6. Ogawa K, Toita T, Nakamura K, Uno T, Onishi H, Itami J, et al. Treatment and prognosis of patients with intracranial nongerminomatous malignant germ cell tumors: a multistitutional retrospective analysis of 41 patients. Cancer 2003;98:369–76.
7. Ferraz ST, Valera ET, Brassesco MS, Santos de Oliveira R, Carlos dos Santos A, Sagüiro FP, et al. Intracranial teratoma in children: the role of chromosome 21 trisomy. Neuropathology 2014;34:197-200.
8. Liu Z, Lv X, Wang W, An J, Duan F, Feng X, et al. Imaging characteristics of primary intracranial teratoma. Acta Radiol 2014;55:874-81.
9. Noudel R, Vinchon M, Dhellemmes P, Litré CF, Rousseaux P. Intracranial teratomas in children: the role and timing of surgical removal. J Neurosurg Pediatr 2008;2:331-8.
10. Han JW, Koh KN, Kim JY, Back HJ, Lee JW, Shim KW, et al. Current trends in management for central nervous system germ cell tumor. Clin Pediatr Hematol Oncol 2016;23:17-27.
11. Zhang X, Wang H, Hong F, Xu T, Chen J. “Bones in the medulla oblongata?” — A case report of intracranial teratoma and review of the literature. Front Pediatr 2021;9:628265.
12. Osborn AG, Preece MT. Intracranial cysts: radiologic-pathologic correlation and imaging approach. Radiology 2006;239:650-64.
13. Beschoner R, Schittenhelm J, BuedtTman E, Ritz R, Meyermann R, Mittelbronn M. Mature cerebellar teratoma in adulthood. Neuropathology 2009;29:176-80.
14. Li Q, You C, Yan X, Chen N, Zhou L, Xu J. Mature cystic teratoma (dermoid cyst) in the sylvian fissure: a case report and review of the literature. J Child Neurol 2012;27:211-7.
15. Hoffman HJ, Otsubo H, Hendrick EB, Humphreys RP, Drake JM, Becker LE, et al. Intracranial germ-cell tumors in children. J Neurosurg 1991;74:545-51.
16. Zyal IM, Bozkurt G, Bilginer B, Gülsen S, Ozcan OE. Abducens nerve palsy in a patient with a parasagittal meningioma—case report. Neurol Med Chir (Tokyo) 2006;46:98-100.