Germinoma occurring 9 years after surgical resection of a mature pineal teratoma: a case report

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European Journal of Medical Case Reports
Volume 5(12):334–336
https://doi.org/10.24911/ejmcr/173-1621160240

ABSTRACT

Background: Mature teratomas are common tumors in the pediatric population. They can arise in the central nervous system and often require surgical resection. They are classified as non-germinomatous germ cell tumors and their recurrence are extremely rare.

Case Presentation: We are reporting the case of a 6-year-old boy who was diagnosed with mature pineal teratoma after he presented with signs of acute hydrocephalus. Histopathology did not reveal any other germ cell tumor component. He underwent a complete resection of the mass, with no adjuvant chemotherapy. Nine years post-operatively, magnetic resonance imaging results showed recurrence of the pineal tumor as a germinoma. Chemotherapy and radiotherapy resulted in significant shrinkage of the mass and resolution of the clinical symptoms.

Conclusion: This case illustrates the possibility of the late recurrence of a germinoma after the complete removal of a mature intracranial teratoma. It also raises the issue of whether mature teratomas should be treated with adjuvant therapy after surgical resection to prevent their recurrence as another germ cell tumor.

Keywords: Germinoma, mature teratoma, germ cell tumor, pineal teratoma, case report.

Received: 16 May 2021 **Accepted:** 13 October 2021 **Type of Article:** CASE REPORT **Specialty:** Pediatric oncology

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Background

Teratomas account for 2% of all intracranial tumors in children, and they usually arise in midline structures such as the pineal gland, basal ganglia, and the thalamus [1]. While teratomas are part of the non-germinomatous germ cell tumors, they are often studied as a distinct category consisting of immature and mature teratomas [2]. Immature teratomas are poorly differentiated and often derive from neural epithelial tissue. Mature teratomas are well differentiated and derive from ectodermal, mesodermal, and endodermal origins [3]. Complete surgical resection is the definitive treatment of mature teratomas in the central nervous system, with a 10-year survival rate of 93% [4,5].

This report documents the case of a patient with germinoma that recurred 9 years after the surgical removal of a mature teratoma in the same pineal region.

Case Presentation

We are reporting the case of a 6-year-old boy with no pertinent past medical history who presented to the emergency room (ER) with headache and diplopia. A brain CT scan showed a pineal mass with obstructive hydrocephalus for which a ventriculo-peritoneal shunt was installed. Magnetic resonance imaging (MRI) was done and showed a mass in the third ventricle extending to the pineal gland. The patient underwent a complete resection of the pineal tumor via the posterior interhemispheric transcallosal route. Histopathological findings revealed a mature pineal teratoma characterized by bronchial and squamous epithelial cells with no evidence of germinoma or any other germ cell tumor components. The tumor was extensively sampled to confirm that there were no areas of immature tissues or malignant transformation. The postoperative period was significant for left crural paresis that resolved on its own. The patient was followed with serial imaging, which were all normal. Seven years post-operatively, the patient was diagnosed with diabetes insipidus, followed by panhypopituitarism.

A routine MRI 9 years after the initial presentation was performed and showed normal findings. Five months later, the patient presented to the ER with a continuous headache of 2 days duration. An MRI was done and showed a suprasellar mass with thickening of the pituitary stalk, extending to the third and left lateral ventricles, which

explained his panhypopituitarism. A biopsy by neuroendoscopy was positive for beta-human chorionic gonadotropin (hCG), CD99, and CD177 tyrosine-protein kinase Kit (c-KIT); these results were in favor of a germinoma. The samples from the initial tumor (mature teratoma) were investigated again to confirm that no immature or malignant tissues were initially present and missed. The patient was lost to follow-up and presented 1 month later with somnolence and altered levels of consciousness. MRI results showed a newly developed large pineal lesion with obstruction of the aqueduct of Sylvius and of the third ventricle. Serum levels of carcinoembryonic antigen, alpha-fetoprotein, and hCG were within normal ranges.

The patient was immediately started on rescue chemotherapy regimen (Cisplatin 20 mg/m^2/day and Etoposide 100 mg/m^2/day for 5 days). He also received radiation therapy, with a total dose of 3,960 rads to the pineal region (22 sessions) and 3,060 rads to the total neuraxis (17 sessions), before completing the rest of his chemotherapy. MRI results showed important shrinkage of the tumor with normalization of the clinical symptoms. The patient remained stable for 10 years on hormonal replacement therapy, requiring serial imaging and blood testing.

Discussion

Current guidelines for the management of mature teratomas recommend complete surgical resection without any adjuvant therapy (chemotherapy or radiotherapy) [6]. Recurrence of such tumors after successful resection is significantly rare. However, if recurrence does occur, it might mean that remnants from the primary mature teratoma reactivated into a germ cell tumor [7]. This is supported by the fact that mature teratomas sometimes consist of different malignant tissues rather than a single "pure" cell origin [7]. A study by Sawamura et al. [8] showed that in case a mature teratoma was suspected of having immature components the patient might benefit from a chemotherapy regimen with cisplatin.

To the authors' knowledge, this is the second study in the literature to report the late occurrence of a germinoma after resection of a mature pineal teratoma. The only other study to document this incidence was published by Sakakura et al. [7], which described the recurrence of a germinoma 11 years after the complete resection of a mature pineal teratoma. Apart from that, a few other studies reported the emergence of a germ cell tumor after total resection of a teratoma: Mano et al. [9] described the development of a germinoma 21 years after resection of an immature teratoma, while Utsuki et al. [10] documented the recurrence of a yolk sac tumor 7 years after resection of a mature teratoma.

In all four case reports discussed above, including our case, no adjuvant treatment was given to the patients after their initial surgical treatment, and in all these cases, the tumor recurred several years afterward. Although mature teratomas are typically considered radio-insensitive, those case reports raise the issue of whether mature teratomas should be treated with adjuvant therapy after surgical resection to prevent the delayed recurrence of a germ cell tumor. A systematic analysis by Lagman et al. [11] showed that patients with either mature or immature teratomas who received adjuvant therapy had increased survival rates compared to those who were managed with surgery alone. However, this could have been attributed to clustering chemotherapy, radiotherapy, and radiosurgery under the same categorical variable (adjuvant therapy). Therefore, further research is required to investigate whether adjuvant radiotherapy truly has a benefit in the management of mature teratomas.

Conclusion

This case illustrates the possibility of the late recurrence of a germinoma after the complete removal of a mature intracranial teratoma. It also raises the issue of whether mature teratomas should be treated with adjuvant therapy after surgical resection to prevent their recurrence as another germ cell tumor.

What is new?

This is the second study in the literature to report the recurrence of a dysgerminoma after resection of a mature pineal teratoma.

List of Abbreviations

ER	Emergency Room
CT	Computed Tomography
MRI	Magnetic Resonance Imaging
hCG	Beta-human chorionic gonadotropin
C-Kit	Tyrosine-protein kinase Kit

Conflict of interest

The authors declare that there is no conflict of interest regarding the publication of this article.

Funding

None.

Consent for publication

A written informed consent to publish this case was obtained from the patient.

Ethical approval

Ethical approval is not required at our institution to publish an anonymous case report.

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Summary of the case

1	**Patient (gender, age)**	Male, 6
2	**Final diagnosis**	Mature pineal teratoma, germinoma
3	**Symptoms**	Headache, diplopia
4	**Medications**	None
5	**Clinical procedure**	Surgical resection
6	**Specialty**	Pediatric oncology