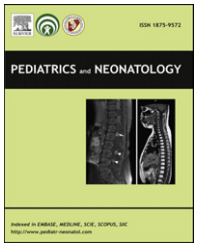
Pediatrics and Neonatology (2012) 53, 315–319

Available online at www.sciencedirect.com

SciVerse ScienceDirect

journal homepage: http://www.pediatr-neonatol.com



CASE REPORT

Effective Radiotherapy Cured Cauda Equina Syndrome Caused by Remitted Intracranial Germinoma Depositing

Ying-Chun Lu [a,d,e], Hueng-Chuen Fan [a,d,e], Hong-Wei Gao [b,d,e], Chang-Ming Chen [c,d,e], Yee-Min Jen [c,d,e], Shin-Nan Cheng [a,d,e], Shyi-Jou Chen [a,d,e,*]

[a] *Department of Pediatrics, Tri-Service General Hospital, Taipei, Taiwan*
[b] *Department of Pathology, Tri-Service General Hospital, Taipei, Taiwan*
[c] *Department of Radiation Oncology, Tri-Service General Hospital, Taipei, Taiwan*
[d] *Tri-Service General Hospital, Taipei, Taiwan*
[e] *National Defense Medical Center, Taipei, Taiwan*

Received May 9, 2011; received in revised form Aug 8, 2011; accepted Aug 14, 2011

Key Words
cauda equina syndrome;
craniospinal irradiation;
germinoma;
metastasis

Cauda equina syndrome (CES) in children is very rare and can permanently disable. A remitted intracranial germinoma depositing on the spinal cord, leading to CES, has never been reported. We discuss the case of a 10-year-old girl who presented with sudden ataxia, low back pain, sensory deficits of the left lower extremity, and difficulty urinating and defecating 7 months after totally remitted intracranial germinoma postintracranial surgery and cranial irradiation. Magnetic resonance imaging (MRI) of the brain and spine showed multiple intradural extramedullary homogeneous masses from the cervical to lumbar levels, compressing the conus medullaris and cauda equina. After emergent craniospinal irradiation, the patient's neurologic symptoms dramatically subsided. A remitted intracranial germinoma depositing on her spinal cord could be the cause of CES. Early identification and a proper craniospinal irradiation may halt the progression of symptoms.

Abbreviations: CES, cauda equina syndrome; MRI, magnetic resonance imaging.

* Corresponding author. Department of Pediatrics, Tri-Service General Hospital, National Defense Medical Center, No. 325, Cheng-Kung Road, Section 2, Neihu, Taipei 114, Taiwan.
E-mail address: chensjou@yahoo.com.tw (S.-J. Chen).

http://dx.doi.org/10.1016/j.pedneo.2012.07.007

1. Introduction

Germ cell tumors are common in children and mostly occur in patients younger than 20 years. Although rare in the West, they are common in Asia, accounting for approximately 3.0% of all primary intracranial tumors.[1] The incidence of germinomas is higher than that of other germ cell tumors in the central nervous system.[2] However, disseminating germinomas leading to neurologic deficits are rare.[3] Cauda equina syndrome (CES) in children is unusual and can permanently disable.[4] Symptoms vary and include low back pain, motor and/or sensory disorders of the lower extremities, and sphincter and bowel dysfunction.[5] The diagnosis of CES is difficult because there is no consensus on diagnostic criteria. Detailed history-taking, careful physical examination, and complete radiologic investigation may shed light on this syndrome. In addition, neurologic deficits such as urination or bowel dysfunction are signs of poor outcome and difficult recovery.[4,6] A variety of underlying pathogens have been identified, but neoplasms such as metastatic germinomas are scarcely noted. We herein present a case of a girl with previously remitted intracranial germinoma depositing on her spinal cord and presenting as CES. She regained normal neurologic function after craniospinal irradiation.

2. Case Report

A 10-year-old girl presented with low back pain for 1 week and sudden limping gait. She was admitted to a medical center for further treatment of ataxia. There was no history of recent trauma, infection, or vaccinations, and she had one suprasellar germinoma (4 × 2 cm in size) that had been partially resected via a cranioorbital zygomatic approach 7 months earlier (Figures 1 and 2). At that time, magnetic resonance imaging (MRI) of the spine and cerebrospinal fluid study revealed no metastatic evidence (Figure 3). The tumor had completely remitted after 22 fractions of whole-ventricle irradiation (24 Gray) and suprasellar boost (12 Gray). No residual or recurrent tumor had been found in the past 7 months' regular follow-up.

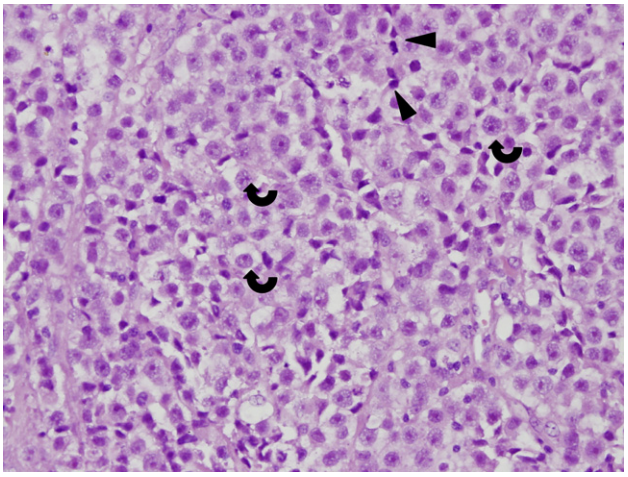


Figure 1 Photomicrograph shows a proliferation of germinoma cells, which are characterized by large tumor cells (curved arrows) with well-defined cell membrane border, clear cytoplasm, central nucleus, and prominent nucleoli as well as small round lymphocytes (arrowheads) infiltrating among the tumor cells (original magnification, ×200; hematoxylin-eosin stain).

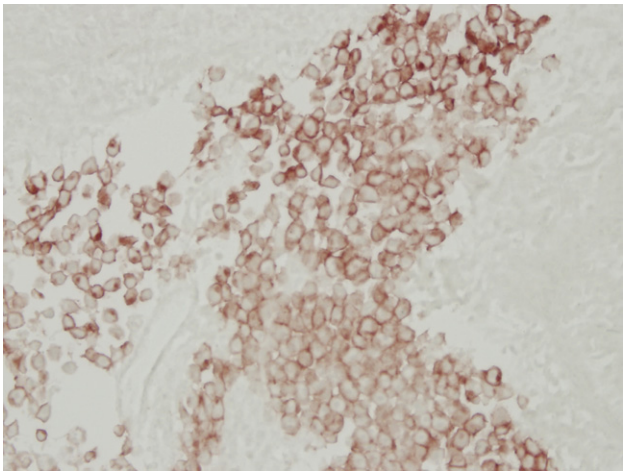


Figure 2 Positive C-kit immunohistochemical reaction for tumor cells (original magnification, ×200; C-kit).

On examination, this patient was alert with clear mentality. Palpation revealed local tenderness and numbness from the patient's left foot to the left side of her waist, conditions that were not present before admission. Severe tenderness and poor sensitivity to pin prick and light touch with a sensory level at T11-S2 on the left were found. The patient had difficult micturition and impaired bowel movement. The muscle power grading of her right lower limb was 2+, left lower limb 3+, and the upper limbs 5+. Deep tendon reflexes of the lower limbs had increased, but those of the upper limbs remained normal. Digital examination revealed a weak anal tone. Laboratory tests showed that β-human chorionic gonadotropin and α-fetal protein levels were normal. Taken together, the patient's clinical presentation and her laboratory investigations were not consistent. To rule out the recurrence of germinoma, we arranged a contrast-enhanced MRI of the brain and spine that showed multiple intradural extramedullary homogeneous masses from the cervical to lumbar levels, compressing the conus medullaris, cauda equina, and cervical and thoracic cords, especially at C7, T9, and T11 (Figure 4), and her brain displayed no abnormalities. Her clinical manifestations and imaging findings supported the diagnosis of CES, and recurrent germinomas depositing on the spine were highly suspected. However, her family rejected pathologic proof of CES. Therefore, empiric therapeutic plans focusing on germinomas were launched, including intravenous dexamethasone 10 mg three times per day and radiation therapy with 12 fractions covering the entire brain (18 Gray) and 15 fractions covering the entire spine (40 Gray). The patient made a full recovery after these treatments. Her neurologic impairments, including ataxia, sensory deficits, muscle weakness, and urination and bowel dysfunction were returned to normal status on the 35th day. She could walk freely without any assistance, and a series of follow-up MRIs of the spine showed no residual tumor after 22 months (Figure 5).

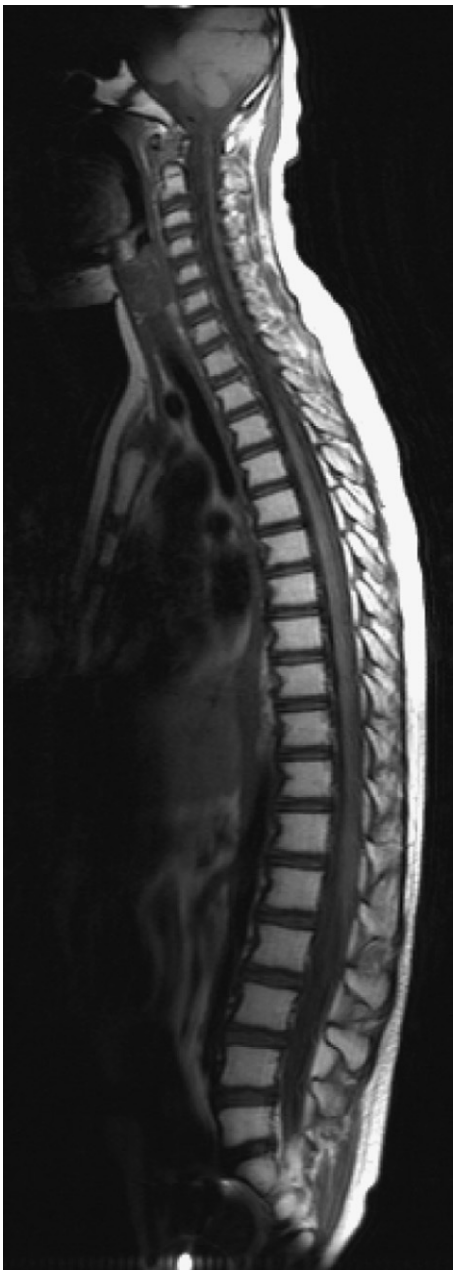


Figure 3 Sagittal gadolinium-enhanced T1-weighted MRI reveals no spinal lesions before the cranial irradiation.

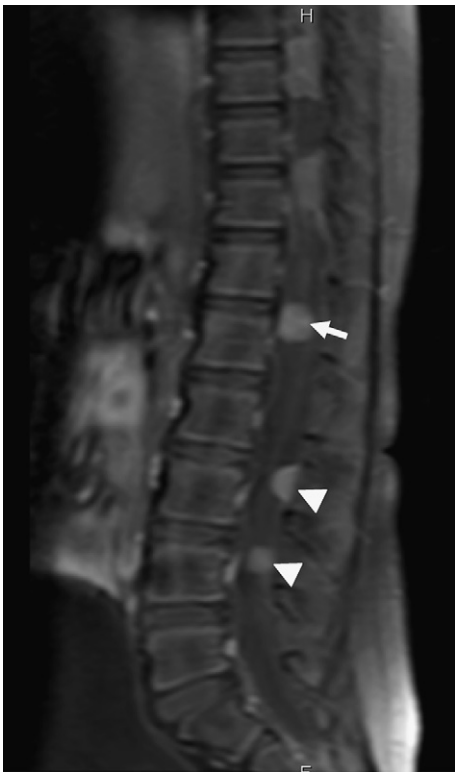


Figure 4 Multiple intradural extramedullary homogeneous spinal masses from cervical to lumbar levels, which compressed conus medullaris (arrow), cauda equina (arrowheads), and cervical and thoracic cord, especially at the levels of C7, T9, and T11.

3. Discussion

CES is very unusual and can permanently disable.[4] CES has a variety of causes, and neoplasms are seldom reported as an underlying etiology of CES. Germinomas are the most common intracranial germ cell tumors in children and constitute 7.6% of all intracranial tumors in Taiwan.[7] So far, the recurrence rate for a remitted intracranial germinoma is unknown. Literature review showed that a remitted intracranial germinoma had been reported to cause thoracic spine metastasis with leg paralysis.[3] In our case, the patient's situation was far more serious, including ataxia, sensory deficits, muscle weakness, and impaired urination and bowel movement. Although the mechanisms of how these tumors metastasize to the spine are not clear, accumulative evidence shows intracranial germinomas may invade the spinal cord with single or multiple loci through the spinal subarachnoid region or ventricular system with or without local intracranial recurrences.[3] Neurosurgery, on one hand, is used to remove tumors. On the other hand, a surgical approach may result in patients having a risk of tumor recurrence or cause tumors to drop and deposit somewhere, including the spine.[3] Therefore, the spinal tumors in this patient might be seeded from her previous remitted intracranial germinoma. Or it could be possible that her cranial surgical approach led to tumor invasion or deposition on her spine.

Pathologic proof is necessary for confirming spinal tumors. Despite unavailability of such proof in this case, the diagnosis of intracranial germinoma with spinal seeding could simply be based on clinical and radiologic features.[3] In this case, there were three reasons in support of the patient's spinal tumors being germinomas. First, this patient had a history of a previously remitted intracranial germinoma after a surgical resection and postoperative radiotherapy with pathologic proof. Second, spinal germinomas may show diffuse well-demarcated and homogeneous appearance and marked contrast enhancement[8]; in addition, on the T1-weighted image, the tumors were hypointense, whereas on the T2-weighted image, they were hyperintense,[9,10] which is exactly the same as the

Figure 5 Totally remitted spinal tumors after craniospinal irradiation.

imaging findings in this case, hence, the findings of this girl's spinal MRI were supportive of the diagnosis. Third, her good response to radiation strongly indicated that the tumors in her spine were germinomas.

In general, cranial irradiation is sufficient to eradicate an intracranial germinoma without spinal dissemination,[11] and cure rates of 70-100% can be achieved with radiation alone.[12–15] However, because the suspicion that the patient's spinal tumors could be stemming from her previously remitted intracranial germinoma depositing, the therapeutic plans for this condition were much more complicated and could cause tumor expansion and metastasis if the radiotherapy was ineffective and not appropriate. A case report showed that a patient with a previously remitted intracranial germinoma presented with sensory and motor abnormalities without urinary or bowel abnormalities.[3] His spine showed a new germinoma, which did not respond well to 10 fractions of local spinal irradiation (30 Gray).[3] Therapeutically, radiotherapy targeting spinal germ cell tumors can include craniospinal irradiation, total spinal irradiation, and local irradiation.[16] Evidence suggests that craniospinal irradiation is most favorable because it may decrease the possibilities of another seeding or recurrence.[17] The irradiation dosage in our case was 40 Gray for covering the entire spine and 18 Gray for covering the entire brain. Our treatment was proper and effective because no recurrence has been noted after the patient's discharge. Although chemotherapy is effective and adjunctive in spinal germinoma,[16] emergent and immediate radiotherapy could provide rapid and effective treatment to this case. To weigh the benefits and risks, radiotherapy instead of chemotherapy was favored because her CES was critical. Surgery was not indicated because the tumors in her spinal cord were diffuse and multiple. A radical operation might have led to a serious destruction in this girl.

In conclusion, CES is unusual and can permanently disable. A previously remitted intracranial germinoma with spinal depositing can be a cause of CES. Early identification and a proper craniospinal irradiation may stop the progression of symptoms.

References

1. The Committee of Brain Tumor Registry of Japan. Frequency of primary brain tumors: report of Brain Tumor Registry of Japan (1969–1993). *Neurol Med Chir (Tokyo)* 2003;**43** Suppl i-vii: 1–111.
2. Fujimaki T. Central nervous system germ cell tumors: classification, clinical features, and treatment with a historical overview. *J Child Neurol* 2009;**24**:1439–45.
3. Kahler RJ, Coyne T. Treatment of an intracranial germinoma spinal metastasis. *J Clin Neurosci* 1999;**6**:171–4.
4. McCarthy MJ, Aylott CE, Grevitt MP, et al. Cauda equina syndrome: factors affecting long-term functional and sphincteric outcome. *Spine* 2007;**32**:207–16.
5. Ma B, Wu H, Jia LS, et al. Cauda equine syndrome: a review of clinical progress. *Chin Med J (Engl)* 2009;**122**:1214–22.
6. Qureshi A, Sell P. Cauda equina syndrome treated by surgical decompression: the influence of timing on surgical outcome. *Eur Spine J* 2007;**16**:2143–51.
7. Wong TT, Ho DM, Chang KP, et al. Primary pediatric brain tumors. *Cancer* 2005;**104**:2156–67.
8. Kanagaki M, Miki Y, Takahashi JA, et al. MRI and CT findings of neurohypophyseal germinoma. *Eur J Radiol* 2004;**49**:204–11.
9. Fujimaki T, Matsutani M, Funada N, et al. CT and MRI features of intracranial germ cell tumors. *J Neurooncol* 1994;**19**: 217–26.
10. Chang T, Teng MM, Guo WY, Sheng WC. CT of pineal tumors and intracranial germ-cell tumors. *Am J Roentgenol* 1989;**153**: 1269–74.
11. Yen SH, Chen YW, Huang PI, et al. Optimal treatment for intracranial germinoma: can we lower radiation dose without chemotherapy? *Int J Radiat Oncol Biol Phys* 2010;**77**:980–7.
12. Bamberg M, Kortmann RD, Calaminus G, et al. Radiation therapy for intracranial germinoma: results of the German cooperative prospective trials MAKEI 83/86/89. *J Clin Oncol* 1999;**17**:2585–92.
13. Haddock MG, Schild SE, Scheithauer BW, Schomberg PJ. Radiation therapy for histologically confirmed primary central nervous system germinoma. *Int J Radiat Oncol Biol Phys* 1997; **38**:915–23.

14. Shirato H, Nishio M, Sawamura Y, et al. Analysis of long term treatment of intracranial germinoma. *Int J Radiat Oncol Biol Physics* 1997;**37**:511—5.
15. Wolden SL, Wara WM, Larson DA, Prados MD, Edwards MS, Sneed PK. Radiation therapy for primary intracranial germ cell tumors. *Int J Radiat Oncol Biol Phys* 1995;**32**:943—9.
16. Biswas A, Puri T, Goyal S, et al. Spinal intradural primary germ cell tumour: review of literature and case report. *Acta Neurochir* 2009;**151**:277—84.
17. Jack MS. Pediatric intracranial germ cell tumors. *UpToDate* 2011. Available at: http://www.uptodate.com/contents/pediatric-intracranial-germ-cell-tumors?view=print. Accessed June 8, 2012.