

Concomitant Cerebellum Subdural Seeding and Spinal Epidural Metastasis of Pineal Germinoma

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Pineal region lesions in adults brings up various considerations. Central nervous system (CNS) germinoma should be considered. Cerebrospinal fluid (CSF) seeding is the main route of CNS germinoma dissemination. The spinal seeding rate has been reported to be 5-20%. Spinal epidural metastasis from CNS germinoma is very rare. Only one case report had been reported in the literature. Here, we report a 43 year-old man with pineal germinoma developing concomitant cerebellum subdural and spinal epidural metastasis 2 years after the right occipital transtentorial approach with total removal of the tumor and radiation. The literature reviews of the seeding mechanism and treatment modality will be discussed.

Key words: CNS germinoma, epidural metastasis, craniospinal irradiation , seeding.

INTRODUCTION

Germinoma in the central nervous system is uncommon in adults and mostly located in the pineal and suprasellar region. Central nervous system (CNS) germinoma has a tendency to “drop metastasis” in the subarachnoid space. We report an extremely rare case with concomitant cerebellum subdural seeding and spinal epidural metastasis and discuss the treatment modality.

CASE REPORT

A 43 year-old man had pineal germinoma initially presenting with headache and blurred vision (Fig. 1). Open surgery with the right occipital transtentorial approach was performed to resolve hydrocephalus and for tissue proof. The gross tumor was totally removed without the development of new neurological deficits. The cerebrospinal fluid (CSF) cytology, sampled after operation through lumbar puncture, was negative for germ cell tumor. Radiotherapy with 30 Gy was prescribed.

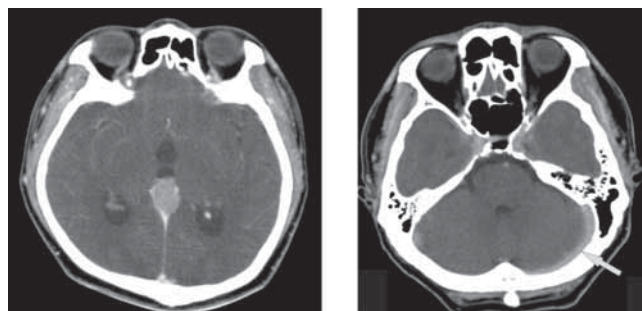


Fig. 1 Left: CT with contrast enhance axial view showed homogenous strong enhancement lesion in pineal lesion. Right: Lower cut level of brain CT with contrast. There was no concomitant lesion found in cerebellum. The left transverse sinus is labeled (grey arrow).

After a 6-month follow-up interval, the central nerve axis was shown to be radiologically and clinically stable. However, 24 months after the first operation, magnetic resonance image(MRI) showed a fusiform subdural mass 3.5×3.0 cm with homogenous Gadolinium enhancement in the left posterior fossa (Fig. 2 left and middle). Meningioma was first impressed by the radiologist because of the dura-tail pattern. The pineal region was free from tumor recurrence (Fig. 2 right). Left suboccipital craniotomy with total removal of the subdural lesion was performed. Macroscopically, the tumor was well-defined, and extra-axial with high vascularity. The pathology report revealed germinoma. Post-operation CSF cytology did not show any germ cell tumors but did show a few leukocytes.

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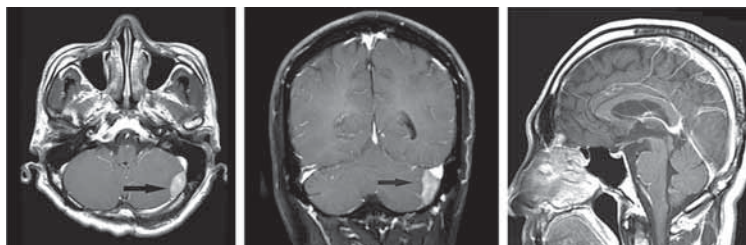


Fig. 2 MRI with Gd enhancement. *Left*: T1WI +Gd axial view showed a homogenous fusiform high signal lesion (black arrow) with dura adherence. *Middle*: T1WI + Gd coronal view showed the lesion(black arrow) located nearby the transverse sinus. *Right*: T1WI+ Gd sagittal view showed no evidence of tumor recurrence over pineal region.

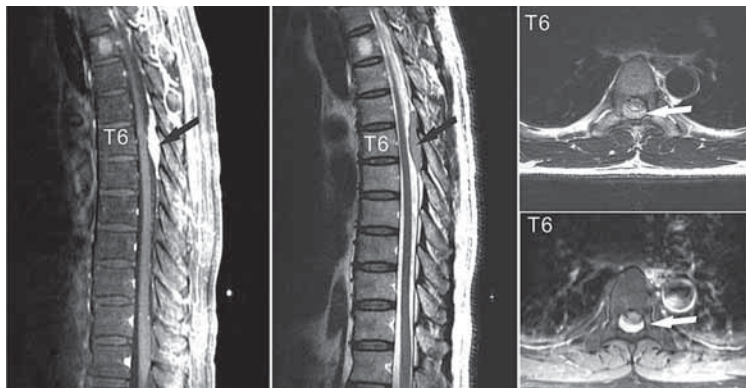


Fig. 3 MRI of spine. *Left*: T1WI + Gd enhancement sagittal view: a strong enhancement fusiform lesion over T5-6 level. *Middle*: T2WI sagittal view. Black arrow indicates the extradural lesion. *Right upper*: T2 axial view. *Right lower*: T1+Gd axial view. White arrow indicates the lesion.

Two weeks after the 2nd operation, he developed bilateral leg weakness with muscle power grade 4, and a clear cut sensory level below T6-7 paresthesia. Emergent spinal MRI showed an epidural mass over the T5-6 level with cord compression (Fig. 3). Due to progression of the neurological symptoms, total laminectomies of T4-6 with total removal of the epidural mass were performed for decompression and tissue proof. The frozen section and permanent pathology report suggested germinoma, consistent with previous pathology findings (Fig.4). The intradural space was explored but no intradural seeding was found. Radiation therapy with 30 Gy in 15 fractions was prescribed for the cerebellum and T5-6 region. At 10 months after spinal metastasis, the patient remained clinically and radiologically stable, and was free from V-P shunt implantation.

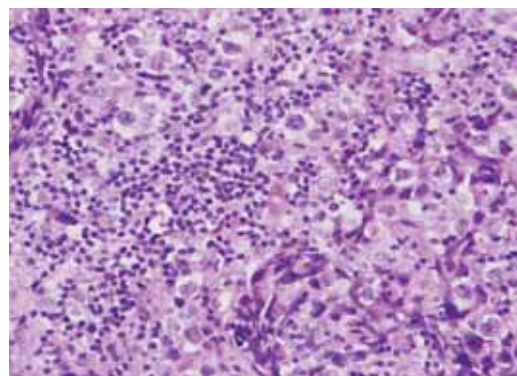


Fig. 4 Histology of spine epidural metastatic lesion. The tumor composed typical large germ cell with prominent nuclei, clear cytoplasm. Lymphocyte infiltration is also characteristic. (Hematoxylin-Eosin stain, X400)

DISCUSSION

The pathology diagnosis of pineal region tumor shows wide variety in the adult group.^{1,2} Regis et al found benign radiosensitive germinoma comprised only 27% of a 373 tumor group with an age range from 3-73 years-old.¹ In the adult group, a more pathological diagnosis should be considered, like astrocytoma, pineocytoma, pineoblastoma, lymphoma, metastasis, and meningioma. Both open surgery, endoscopic or stereotactic biopsy offer high diagnosis yield (100%, 94%, and 70% respectively), with acceptable morbidity.^{1,3} The reasons for inconclusive biopsy are insufficient tissue and loss of structure architecture. We performed open surgery to resolve hydrocephalus by tumor removal, and for adequate tissue to achieve histology diagnosis. We resolve obstructive hydrocephalus by total tumor removal, instead of V-P shunt implantation, which had been reported with tumor dissemination.⁴ There is disagreement that surgery intervention facilitates tumor seeding.⁵⁻⁷ However, an inadequate number of series have been reported and there may be selection bias. We think more data are needed to reach a definite conclusion.

Intracranial germinoma has been reported with a 5-20% spinal seeding rate.⁸⁻⁹ Primary spinal germinoma has been reported, but they were all intradural intramedullary or extramedullary tumors.¹⁰ Epidural metastasis from CNS germinoma had been found in only one case report.¹¹ The author suspected the dissemination route to

be CSF seeding since part of the tumor was located intradurally, although the CSF cytology revealed a negative finding.¹¹ There is a report of testicular and extragonadal germ cell tumor metastasizing to the vertebral body through the blood and forming an epidural mass.¹² Our patient, a prior subdural CSF seeding lesion in the left posterior fossa near the transverse sinus, may have also been vulnerable to blood dissemination.

The proper radiation dose and cover range to CNS germinoma varies, with considerations of tumor size, CSF cytology, and combination chemotherapy.^{9,13-14} Shibamoto et al also found no correlation between dose and tumor relapse.¹⁵ We believe surgery achieves immediate tumor debulking and may help in reducing the radiation dose, which is closely related to the post-radiation neurological deficit. There is no data on the relationship between residual tumor burden and proper radiation dose. Prophylactic spinal irradiation should be preserved only for the high-risk group using a limited dose.^{8,13} In literature reviews, negative CSF cytology showed little correlation to the spinal seeding tendency.¹¹ However, positive CSF cytology truly directs more attention to spinal seeding.^{9,13} Brada et al also found true pineal or suprasellar germinoma itself had a higher CSF seeding rate than other tumors, but the actual risk is not high.⁸ We think periodic clinical and radiological follow-up is mandatory.

CONCLUSION

Diagnosis and treatment policy toward pineal lesion should be tailored, according to age, pathology, tumor radiosensitivity, combined neurologic deficit, and concomitant disease. The prognosis and therapy mostly depend on the tumor type. With CNS germinoma, constant periodic follow-up for the central nervous system is mandatory. For this patient, the prognosis remained uncertain.

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