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Pineal Germinoma with Extracranial Metastases:
Case Report[1]

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Pineal germinoma is the commonest pineal region tumor of childhood and adolescence. Metastatic germinoma most commonly occurs via the cerebrospinal fluid (CSF), and it is usually limited to the cerebrospinal axis. Extacranial hematogenous metastasis is known to be very rare.

We report here on a case of pineal germinoma with gradual extracranial metastases that occurred both through the CSF pathway and by hematogenous spread. The patient had multifocal CSF seeding after his surgery for pineal germinoma, and the left iliac metastasis and lung metastasis then occurred.

Index words : Bone neoplasms, metastases
Lung neoplasms
Pineal body, cysts
Spinal cord

Pineal germinoma is the commonest pineal region tumor of patients in the 3rd decade of life. It is a metastatic disease that usually occurs via the cerebrospinal fluid (CSF), and on rare occasions, via a ventriculoperitoneal shunt (VP shunt). Extracranial hematogenous metastasis is, however, very rare and it usually follows multiple surgical interventions (1).

We describe here a case of pineal germinoma with extracranial metastasis that occurred through the CSF pathway, and the disease progressed onward to hematogenous metastasis.

Case Report

A 25-year-old man presented with a recent, 2-week onset of headache, intermittent episodes of nausea and vomiting, and limitation of his upward gaze. Cranial CT and MRI scan demonstrated a large calcified contrast-enhanced pineal mass with mild obstructive hydrocephalus (Figs. 1A, B). A near-total tumor removal and internal shunt was performed. The histological diagnosis was proven to be pineal germinoma (Fig. 1C).

Eight months later, he experienced recurrent symptoms of headache, nausea and vomiting. Cranial CT disclosed a recurred mass around the right cerebellopontine angle and cerebellum with ventriculomegaly. At this time, a ventriculoperitoneal shunt operation with filter was done.

Three years later from the time of the first neat total removal of pineal germinoma, he complained of the motor weakness of his left lower extremity. The spinal MRI scan revealed an intramedullary mass at the level of T12 (Fig. 1D). Subtotal tumor removal and adjuvant radiation therapy was performed. The microscopic findings of the tumor specimen were identical to those of the original pineal germonima.

Five years later from the first surgery, he was again

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hospitalized because of persistent left pelvic and lower extremity pain, and an abnormal chest X-ray finding. The chest CT scan showed a large mediastinal lymphadenopathy and a cavitary nodule in the right lower lobe (Fig. 1E). Bone scintigraphy showed an increased uptake in the left iliac bone (Fig. 1F), which strongly sug-

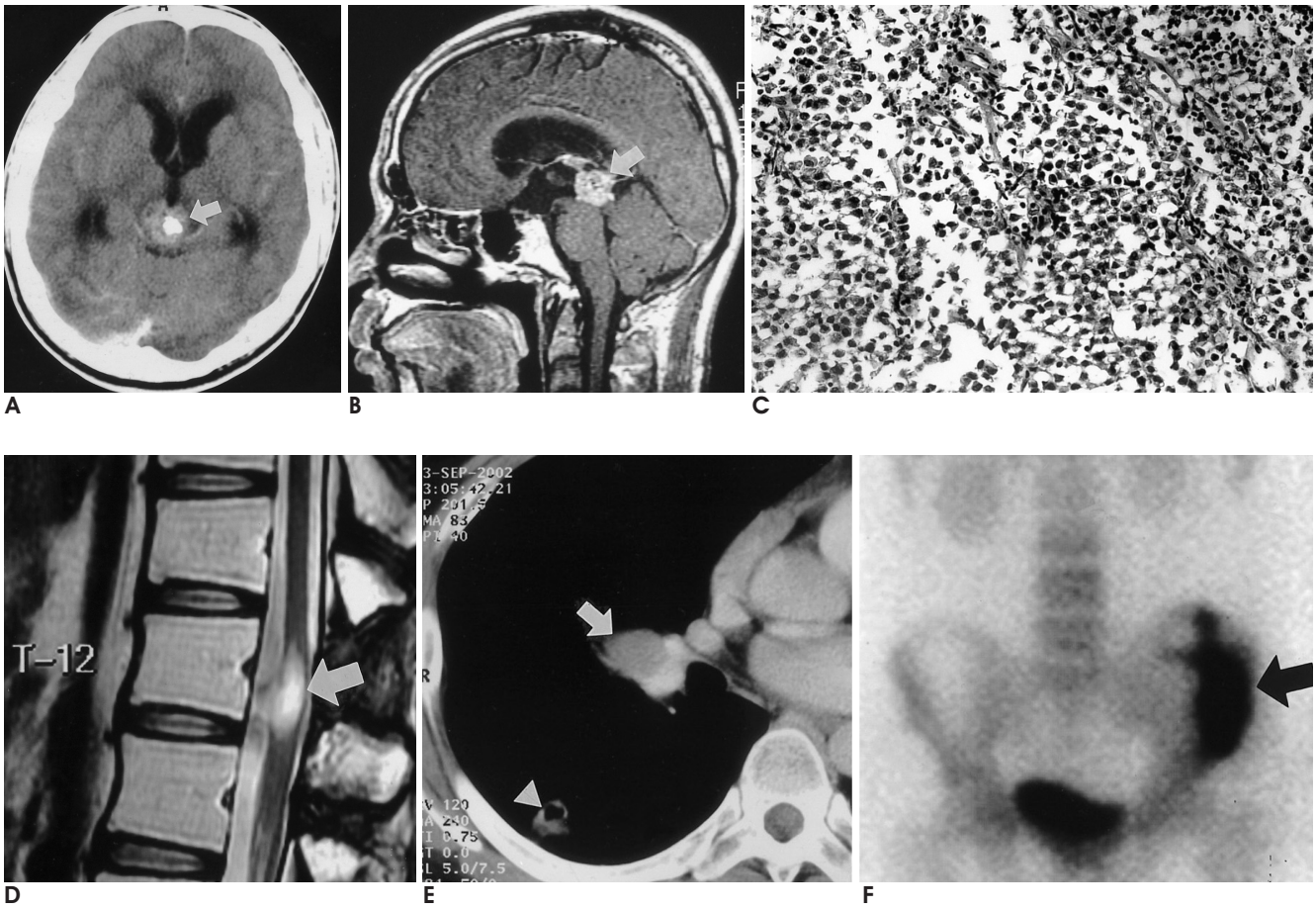

Fig. 1A-C. A 25-year-old man with surgically confirmed pineal germinoma who initially complained of headache, nausea, vomiting and limitation of his upward gaze.

A. Contrast-enhanced CT scan of the brain shows a well-enhanced mass (arrow) with a calcification in the area of pineal gland causing mild obstructive hydrocephalus.

B. Contrast-enhanced sagittal T1-weighted MR scan of the brain demonstrates a well-enhanced mass (arrow) with small poorly enhanced portion, and this was probably due to calcification in the quadrigeminal cistern.

C. Photomicrogram of a specimen from the pineal mass shows that the mass consists of large tumor cells arranged in sheets or lobules, and small lymphocytes along the fibrovascular septae (Hematoxylineosin stained, ×200).

D. Contrast-enhanced T1-weighted sagittal MR scan of the spine three years after the surgery shows a well-enhanced intramedullary mass (arrow) at the level of the twelfth thoracic vertebra, which was confirmed as a metastatic germinoma by the surgical biopsy.

E-G. CT scan and bone scintigraph and photomicrogram of a specimen from the lung mass five years after the first surgery.

E. Contrast-enhanced CT scan of the chest shows a large mediastinal lymphadenopathy (arrow) and a cavitary lung nodule (arrowhead) in the superior segment of the right lower lobe of the lung.

F. Tc-99m bone scintigraph demonstrates an increased uptake (arrow) in the left iliac bone that was strongly suggestive of metastasis.

G. Photomicrogram of a specimen from the lung mass obtained by fine needle aspiration biopsy shows large tumor cells (arrows) with abundant clear cytoplasm, round nuclei and prominent nucleoli that were very similar to the cells from the pineal mass. (Hematoxylin-eosin stained, ×400).

gested a diagnosis of metastasis. Fine needle aspiration biopsy of the lung mass revealed the tumor as a metastatic germonima (Fig. 1G). The metastatic thoracic and pelvic lesions were controlled with radiation therapy.

Discussion

Germinomas are uncommon tumors of the central nervous system comprising only 0.4 to 2.0% of all intracranial neoplasms. They usually arise within the pineal gland, and they constitute more than one-half of all tumors of the pineal gland and suprasellar region (2).

These tumors respond well to radiotherapy and chemotherapy and are known to have a favorable prognosis, despite of a propensity to spread locally (2). The prognosis is not significantly affected by the development of extracranial metastasis, although these metastases are only infrequently noted.

Since 1977, metastasis of pineal region tumors to the abdominal and pelvic cavities via the patient 's VP shunt has been described (3, 4). The germinoma invades locally, eroding into the ventricular system, and it leaves implanted cells throughout the subarachnoid space, even as far as the cauda equina (5). Therefore, peritoneal metastasis is thought to develope probably secondary to a VP shunt (6). However, the metastatic dissemination to lung and bone could not be explained by peritoneal seeding alone. These secondary tumors would have to be explained as a hematogenous spread of neoplastic cells. The surgical manipulation of the pineal lesion, anatomic discontinuity with subarachnoid spaces and root sleeves suggest the possibility of a blood-borne spread from the original tumor (7).

Two cases of hematogenous pulmonary metastasis from the primary pineal germonima were reported by Borden et al. (5); these were confirmed by an open lung biopsy. Galassi et al. (8) also described a case of multiple lung and bone metastases from a presumed primary pineal germinoma 15 months after the initial diagnosis.

In our case, there might have been extracranial metastases via the CSF pathway through the central canal of the spinal cord and the hematogenous spread to the lung and bone. The extracranial metastasis gradually recurred, although he was thoroughly treated with near-total tumor removal and craniospinal irradiation. Even so, the use of a filter in this patient 's shunt may have prevented his peritoneal metastasis, and none of serial examinations for the abdomen revealed peritoneal seeding. As far as we know, all the previous reports have described either hematogenous metastasis or metastasis through the CSF pathway (1. 3 - 5. 9. 10). However, our case had a very rare combination of hematogenous metastasis and metastasis through the CSF pathway with gradual recurrence.

In conclusion, primary pineal germinoma with secondary extracranial metastasis through the CSF pathway and hematogenous metastasis is very rare. The development of the metastasis and the long time intervals between each of the metastatic events in this case serve to emphasize that continued follow-up is necessary despite the numerous reported instances of long term survival following radiation therapy and chemotherapy (2). This follow-up should include the investigation for local recurrence and distant tumor spread with the use of regular radiologic evaluations.

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송과선 배아종의 다발성 두개외 전이: 증례 보고[1]

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송과선 배아종은 유년기와 청소년기에서 가장 많은 송과선 암종이며 전이는 대개 뇌척수액을 통해 이루어져 뇌척수지주에 한하며 혈행성 두개외 전이는 매우 드문 것으로 알려져 있다.

이에 저자들은 송과선 배아종에 대한 수술 후 뇌척수액과 혈행성으로 점차적인 전이가 발생한 증례를 경험한 바 이를 보고하기로 한다. 그는 송과선 배아종 수술 후 다발적인 뇌척수액 전이를 보였고, 그 후에도 좌측 장골과 폐 전이가 일어났다.