Intrathecal and intraperitoneal germinomas occurring 20 years after total removal of a pineal teratoma

Case report

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In 1980, a 10-year-old boy was admitted to the authors’ hospital with consciousness disturbance and vomiting. Computerized tomography (CT) scans revealed a pineal mass lesion and hydrocephalus. The tumor was totally removed and a ventriculoperitoneal (VP) shunt was inserted. Histological investigation of a surgical specimen revealed that it was a teratoma. Five years later, the patient was readmitted to the same institution with polyuria. Magnetic resonance (MR) imaging revealed a mass lesion in the suprasellar region. The patient received systemic chemotherapy, and the tumor disappeared; however, 2 years after the chemotherapy, MR imaging demonstrated a right lateral ventricular mass. The tumor was totally resected, and histopathological investigation revealed a teratocarcinoma. Three years after the chemotherapy, CT scanning revealed suprasellar and right lateral ventricular tumor recurrences, for which the patient received irradiation and chemotherapy. The tumors disappeared and the patient achieved complete remission that lasted longer than 10 years. On January 25, 2000, however, he noticed hip pain. Lumbar MR imaging demonstrated a spinal tumor below L-4 and also an abdominal tumor. The abdominal tumor was totally removed, and the histological findings identified it as a germinoma. The patient received systemic chemotherapy and the tumor disappeared completely. The authors believe that the suprasellar tumor was a metachronous germinoma and that it had metastasized through the intrathecal route and the VP shunt.

KEY WORDS • teratoma • germinoma • germ cell tumor • pineal gland • ventriculoperitoneal shunt

Abbreviations used in this paper: AFP = alpha-fetoprotein; CT = computerized tomography; HCG = human chorionic gonadotropin; MR = magnetic resonance.
tumor was resected totally via an occipital transcortical approach at another institution. Histopathological investigation revealed a teratocarcinoma, but the specimen was not preserved. On March 19, 1988, a CT scan demonstrated that both the suprasellar and right lateral ventricular tumors had recurred (Fig. 5). The patient received chemotherapy consisting of cisplatin, and radiation therapy for treatment of the suprasellar and ventricular tumors. For 12 years after the last chemotherapy course, MR imaging demonstrated no evidence of a recurrence. The patient has been followed up at our institution as an outpatient and has received anticonvulsant medications and desmopressin.

Examination. On January 25, 2000, the patient noticed pain in his hip and presented at our hospital. Lumbar MR imaging and abdominal CT scanning revealed an intrathecal mass below the L-5 level (Fig. 6) and three intraperitoneal masses (Fig. 7). Laboratory examinations revealed a slightly raised serum HCG level (17 mIU/ml) and a normal AFP level.

Operation and Pathological Findings. An abdominal operation was performed and the intraperitoneal tumors were totally removed. Histological investigation of specimens obtained at surgery revealed a germinoma (Fig. 8), although a slightly raised HCG level indicated that the tumor was instead a germinoma with syncytiotrophoblastic giant cells. Cytological findings in the cerebrospinal fluid were negative.

Postoperative Course and Further Treatment. One month after the surgery, a CT scan of the abdomen demonstrated tumor recurrence. The patient received three cycles of combination chemotherapy consisting of cisplatin, etoposide, and bleomycin. After this treatment, both intraperitoneal and intrathecal tumors disappeared. The patient was discharged with diabetes insipidus, diplopia, and hypesthesia of the lower limbs.

Discussion

Multiple intracranial germ cell tumors account for approximately 10% of all intracranial tumors of this type. The majority of these lesions occur synchronously along the midline and are thought to be multicentric in origin.
On the other hand, metachronous germ cell tumors, multiple de novo lesions arising at different sites with a long interval between their occurrences, are rare in the central nervous system. To our knowledge, only two case reports clearly demonstrate metachronous germ cell tumors. The authors of these reports described suprasellar germinoma occurring 15 years and 8 years after total removal of pineal mature teratoma, respectively. In our case, the primary pineal tumor was an immature teratoma and the right lateral ventricular tumor was a teratocarcinoma. These tumor sites were distinct from each other.

These results indicate that the intraventricular lesion was either a metachronous germ cell tumor or a metastatic tumor of the pineal teratoma with malignant change. The lateral ventricular tumor might be a metachronous germ cell tumor because these lesions are often located in the lateral ventricle. The primary pineal tumor did not recur, and recurrence of germ cell tumors usually takes place within 1 year after surgery. Although the suprasellar tumor was not diagnosed histologically, this lesion and the primary pineal tumor also originated at distinct sites. Because the intraperitoneal tumors were germinomas that seemed to be disseminated through a VP shunt, and because the serum HCG level was slightly elevated when these tumors appeared, the suprasellar tumor is thought to be a germinoma (or a germinoma with syncytiotrophoblastic giant cells). The suprasellar tumor might also be a metastatic lesion that was a different histological type from the pineal teratoma.

There are some case reports with descriptions of recurrent germ cell tumors that are a different histological type from the primary tumor. In these reports the following were demonstrated: 1) focal recurrence of mixed germ cell tumor; 2) mixed germ cell tumor including a malignant component that might have metastasized; or a
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radiosensitive component that might have been eliminated by radiation therapy, and 3) cases of metachronous germinoma. Our patient received no adjuvant therapy between the first and second operations. The primary tumor might have included a germinomatous component, despite thorough histological investigation, and the pineal tumor did not recur. The suprasellar tumor occurred 5 years after resection of the pineal tumor. Germinoma commonly originates from the suprasellar region. Accordingly, the suprasellar tumor was thought to be a metachronous germinoma. After the second operation, the patient received chemotherapy and radiation therapy. Because germinoma is the most radio- and chemosensitive lesion among germ cell tumors, the suprasellar germinoma had probably disseminated to the abdomen and spinal cord before the adjuvant therapy.

The newly detected intraperitoneal tumors were germinomas. Primary germinoma does not usually originate from the intraperitoneal or intrathecal space. Some reports have demonstrated intraperitoneal dissemination through a VP shunt, and ours was considered to be another such case. The intrathecal tumor was not investigated histologically but was considered to be an intrathecal dissemination because chemotherapy had the same effect on this one as on the intraperitoneal tumor, and both lesions were detected simultaneously. Namely, we speculate that a pineal teratoma, a lateral ventricular teratocarcinoma, and a suprasellar germinoma occurred metachronously, and that the suprasellar germinoma was disseminated into intraperitoneal and intrathecal spaces before chemotherapy and radiation therapy were administered.

In this case, the patient received cranial irradiation and systemic chemotherapy after the second operation, and brain MR imaging demonstrated no tumor recurrence for longer than 10 years. The growth rate of the newly detected abdominal and spinal tumors seems to be low. Long-term follow up is necessary in a patient with a germ cell tumor.

Fig. 8. Photomicrograph of the surgical specimen showing a germinoma. H & E, original magnification × 200.