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Pinealoblastoma with Shunt Metastasis : A Case Report[1]

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An unusual case of pinealoblastoma metastasizing through a ventriculoperitoneal(VP) shunt to the peritoneal cavity in a 10-year-old girl is presented with a review of the literature.

Index Words : Pineal body, neoplasms
Pineal body, CT
Neoplasms, metastases

INTRODUCTION

The extracranial metastaisis is an uncommon complication of primary central nervous system(CNS) tumors of childhood. In contrast, leptomeningeal dissemination or multiple tumor recurrence is not a rare event. Metastatic spread of pediatric CNS tumors occurs most commonly with primitive neuroectodermal tumors(PNET) and malignant gliomas. In general, the incidence rates of extracranial metastases in children with PNET is very low, 0.4% to 6.9%, although some investigators report rates as high as 10% to 20%(1—4). Extracranial metastases to the peritoneal cavity are very rare in primary CNS tumors, despite the frequency of ventriculoperitoneal(VP) shunting. They have been recorded occasionally in medulloblastomas (infratentorial PNET)(5).

CASE REPORT

A 10-year-old girl had been well until the first admission, when she developed headache, dizziness, and diplopia for one week. Physical and neurologic examinations were within normal limit at that time, except for slight drowsy mentality. Computed tomography(CT) of the head revealed a posterior third ventricle tumor and severe hydrocephalus. Brain MR imaging disclosed a large, enhancing, isointense mass in the posterior third ventricular region and severe hydrocephalus with periventricular edema(Fig. 1a). A right VP shunt and a stereotaxic biopsy of the tumor was performed. Histological diagnosis of the tumor was a pinealoblastoma.

Follow-up CT, which was performed 9 days after the insertion of VP shunt revealed a new, non-enhnacing, isodense mass at the anterior body of the right lateral ventricle in addition to the posterior third ventricular tumor. The patient received a 42 Gy of radiation to the region of right lateral ventricle, pineal gland, and spinal cord over the course of 4 weeks. Brain CT scan, which was performed after the radiation therapy, showed almost compeate regression of the masses and no hydrocephalus(Fig. 1b). After treatment, the patient remained asymptomatic with free of tumor recurrence on the follow-up CT scans for about 1 year after which she began to develop general weakness, left heimparesis, and vomiting. Brain MR imaging(Fig. 2a) disclosed a large, enhancing, isointense mass with small central cystic portion occupying the right lateral ventricle. There was also severe hydrocephalus.

A second course of radiation therapy over a 6-week period was given, which included 50 Gy to the brain. Brain CT, 5 weeks after the second course of radiation theraphy, showed marked decrease in size of the tumor in the right lateral ventricle and restoration of the dilated ventricles to normal size. But slightly hypodense subdural collection developed along the right frontotemporal region(Fig. 2b). The tumor reduced in size progressively on the follow-up CT scans during 2 months thereafter.

The last admission, 3 years and 8 months after the VP shunt, was prompted by a large, palpable pelvic mass. CT of the abdomen(Fig. 3) demonstrated a large, multilobulated, thick-walled, mixed-density mass ar-

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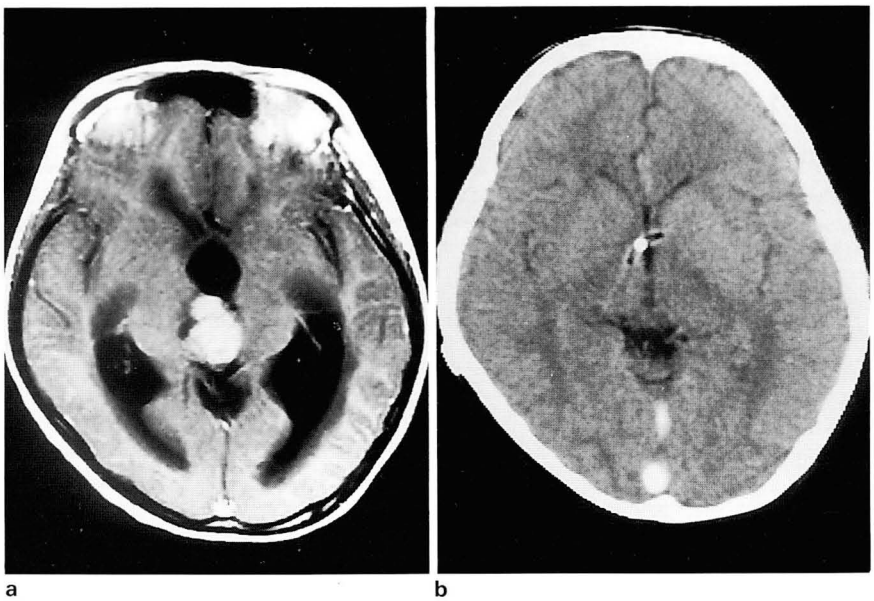


Fig. 1. Postcontrast MR imaging(a) shows an enhancing tumor in the posterior third ventricle, causing marked obstructive hydrocephalus with periventricular edema. Postcontrast CT scan(b) 4 1/2 months following radiotherapy reveals complete resolution of the tumor.

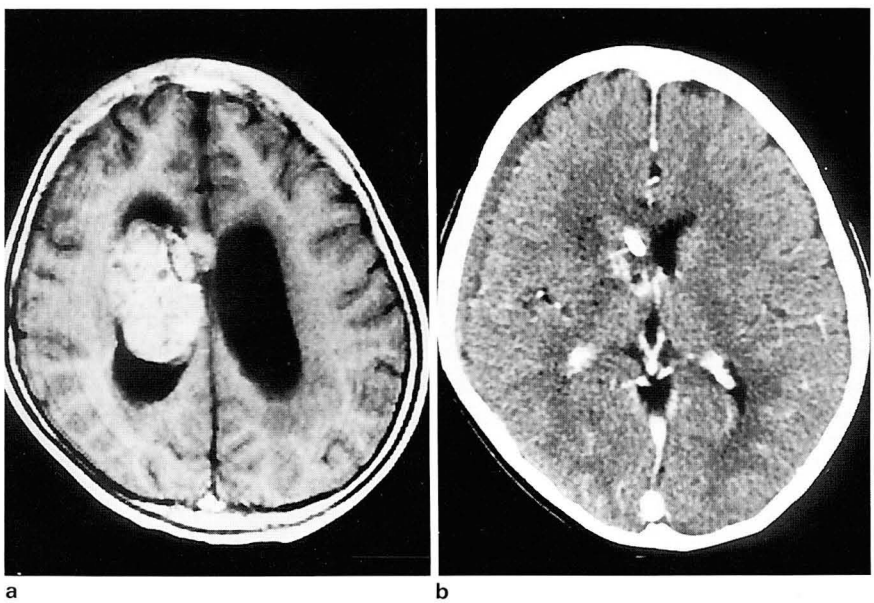


Fig. 2. Postcontrast MR imaging(a) 1 year following radiotherapy demonstrated a new, large enhancing tumor in the right lateral ventricle with an exophytic component and severe obstructive hydrocephalus. Postcontrast CT scan(b) 5 weeks following the second radiotherapy demonstrates marked decrease in size of the tumor and hydrocephalus. Subdural fluid collection along right frontotemporal region is also seen.

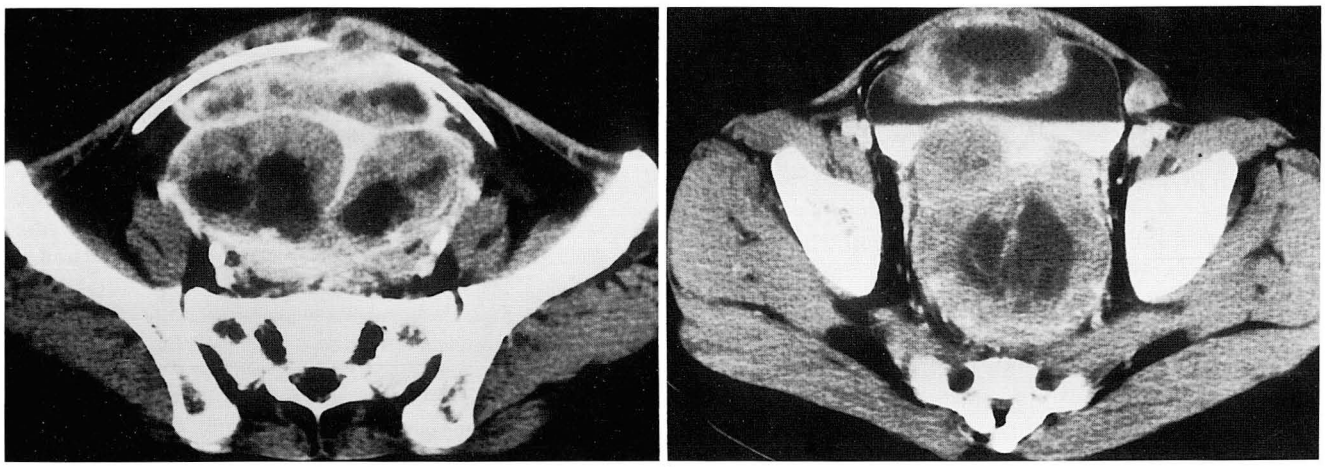
ound the distal portion of the VP shunt catheter in the pelvis filling the vesicorectal peritoneal space. The pelvic mass showed solid enhancing one with central low density area. The urinary bladder, rectum, and small bowel loops were compressed by the mass. On exploratory laparotomy, the mass noted on the CT scan was accompanied by diffuse peritoneal spread and invasion of the right colon, small bowel loops, aorta, right ovary, and urinary bladder. Histologic study demonstrated a primitive neuroectodermal tumor(PNET) with ependymal differentiation, which was identical in appearance to the initial stereotaxic biopsy specimen.

DISCUSSION

The metastatic dissemination via CSF shunting devices has been observed for a number of primary brain tumors and the causal relationship between systemic CSF shunts and extracranial metastases was first suggested by Wolf and coworkers in 1954(6, 7).

The clinical pattern of systemic metastases in pediatric brain tumors is extremely varied. The metastases may present soon after the initiation of therapy or may be detected several years after diagnosis. In some instances, the presence of metastases was not suspected until autopsy(3).

Several factors may be responsible for the develop-



a b

Fig. 3. Postcontrast CT scans at different levels of the pelvis 3 years and 8 months after the VP shunt. A large, heterogenously enhancing, mixed-density tumor surrounds distal portion of the VP shunt catheter(a). Extrinsic compression or invasion anteriorly and posteriorly on contrast-filled bladder is also demonstrated(b).

ment of extracranial metastases of the primary brain tumors. These include surgical intervention resulting in the access of neoplastic cells to the vascular and lymphatic system, and prolonged survival due to advances in radiotherapy and chemotheraphy, coupled with improvements in diagnostic imaging for the identification of extracrainal metastases. An additional factor implicates dissemination of tumor cells through CSF shunt device to body cavities outside the CNS. Among the factors, tumor type appears to be the most important in the previous reports, with medulloblastoma as the most common primary source. Thus, patients with medulloblastomas and CSF shunts most often developed extracranial metastases to bone, bone marrow, and lymph nodes. Yet very few cases with VP shunts demonstrated metastases which primarily involve intra-abdominal structures without extra-abdominal sites, or lungs and pleura with ventriculoatrial(VA) shunts. Other tumors, less frequently associated with dissemination outside the CNS are ependymomas, germ-cell tumors, malignant gliomas, supratentorial PNET's, meningeal sarcomas, and choroid plexus tumors. Pollack et al. (2) reported the first documented case of peritoneal seeding of a benign astrocytoma after placement of a VP shunt. The criteria for the assumption that systemic metastasis from mdulloblastoma has probably occurred through a VP shunt are not well defined, because this tumor can metastasize to almost all organ system in the absence of CSF shunts. Jamjoon et al. suggested four levels of confidence for the assumption that systemic metastasis from medulloblastoma occur through a VP shunt : (1) Unlikely includes all cases with bone, bone marrow, and lymph node metastasis with or without involvement of other abdominal viscera, and applies to both atrial and peritoneal shunts ; (2) Probable includes patients with VP shunts who developed predominantly lung and/or pleural metastases ; (3) Most probable includes patients with peritoneal seeding and/or ascites developing in the presence of a VP shunt ; and (4) Certain includes cases in which tumor deposits were detectable around the tubing of the shunt as in this presenting case. Using these four confidence levels, only 11 out of the 160(6.9%) cases of the systemic metastases of medulloblastoma could have occurred through or been facilitated by ventriculosystemic shunts in their analysis of the literature(4).

Berger et al. (1) reported a comparable incidence of extracranial metastases in 40 shunted and 37 unshunted patients with medulloblastoma. Extracranial metastasis was documented in three of 40 shunted patients, and five of 37 unshunted patients, so the difference between these two groups was not statistically significant($p=0.47$, Fisher's exact test). In their report, they concluded that CSF shunts do not predispose pediatric patients with brain tumors to develp extracranial metastasis and that a diagnosis of shunt-related metastases should be based on the development of intra-abdominal(VP shunt) or pulmonary(VA shunt) dissemination.

Dissemination from the primary tumor to the neuraxis is an important prerequisite before development of extracranial metastases. This is especially true in cases of intraperitoneal metastases, in which meningeal dissemination has been documented in virtually every patient. Once peritoneal metastases develop, they may or may not be associated with ascites(5). Berger et al. (1) reported that no children with extraneural metastases developed ascites in the absence of a VP shunt and that pineal germ-cell tumors had a slightly increased incidence of abdominal dissemination with a VP shunt in place. Paine et al. (6) found in 12 cases of germinomas with VP shunt metastasis that the principal site of VP shunt metastasis was related to the site of the distal aspect of the shunt tube, involving

the peritoneal, retroperitoneal, and pelvic cavities. In the literature, six patients with VP shunts presented with some degree of ascites in addition to evidence of intra-abdominal tumor dissemination, regardless of the primary tumor type. Overall, very few children with shunts in place develop abdominal tumor dissemination with VP shunts, however, when it dose occur, a diagnosis of shunt-related metastasis should be made.

In the presenting case, there is no doubt that the abdominal tumor around the distal portion of VP shunt catheter is a metastasis of the pinealoblastoma and infiltration of the colon, small bowel, retroperitoneum, aorta, ovary, and bladder implies direct access of tumor cells to these regions via a VP shunt as the vehicle of extracranial spread. Both intracranial and abdominal tumors showed identical histopathology, which ruled out a multineoplastic syndrome.

In conclusion, follow-up abdominal CT scans should be obtained, even in asymptomatic patients with VP shunts associated with CNS tumors with a demonstrated propensity for metastasis along this route.

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단락을 통한 송과체아세포종의 전이 : 1예 보고[1]

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김 종 덕

송과체아세포종 때문에 뇌실-복막 단락을 시행한 10세의 여아에서 이 단락을 통하여 복막강내로 전이된 1예를 문헌고찰과 함께 보고하는 바이다.