Pinealoblastoma with Shunt Metastasis: A Case Report

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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 metastasis 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-enhancing, 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 complete 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 hemiparesis, 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 therapy, 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-
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-
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).

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 ventriculostemtic 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 develop 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 extraaxial 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 does occur, a di-
agnosis 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 in-
filtration 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 ve-
hicle of extracranial spread. Both intracranial and ab-
dominal 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 demon-
strated propensity for metastasis along this route.

REFERENCES

1. Berger MS, Baumeister B, Geyer JR, Milstein, Kanev PM, Le-

Roux PD. The risks of metastases from shunting in children with
primary central nervous system tumors. J Neurosurg 1991 ; 74 :
872-877
2. Pollack IF, Hurt M, Pang D, Albright L. Dissemination of low
grade intracranial astrocytomas in children. Cancer 1994 ; 73 :
2669-2678
3. Campbell AN, Chan HSL, Becker Le, Daneman A, Park TS, Ho-
ffman HJ. Extracranial metastasis in childhood primary intra-
cranial tumors : a report of 21 cases and review of the literature. 
Cancer 1984 ; 53 : 974-981
4. Jamjoon ZAS, Jamjoon AB, Sulaiman AH, Rahman NU, Al-
Rabiaa A. Systemic metastasis of medulloblastoma through
ventriculoperitoneal shunt : report of a case and critical analysis
5. Newton HB, Rosenbaum MK, Walker RW. Extraneural meta-
stases of infratentorial glioblastoma multiforme to the peritoneal
6. Paine JT, Handa H, Yamasaki T, Yamasita J. Suprasellar ge-
minoma with shun metastasis : report of a case with an immu-
nohistochemical characterization of the lymphocyte subpopu-
7. Wolf A, Cowen D, Stewart WB. Glioblastoma with extraneural met-
astasis by way of a ventriculoperitoneal anastomosis. Trans Am
Neurol Assoc 1954 ; 79 : 140-142