

Medulloblastoma: seeding of VP shunt tract and peritoneum

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Abstract

We report on a 5-year-old boy with seeding of the peritoneum and a ventriculoperitoneal shunt tract by anaplastic medulloblastoma. The role of ventriculoperitoneal shunting in the spread of primary central nervous system tumors has been controversial. In the case reported here, the unique distribution of tumor implants on ultrasound and multiplanar computed tomography gives further credence to the argument that ventriculoperitoneal shunting is a pathway for extraneural metastases of primary central nervous system tumors.

Introduction

Medulloblastoma (MB) is the most common pediatric central nervous system (CNS) malignancy.¹ Arising from cerebellar vermis, it commonly spreads along cerebrospinal fluid (CSF) pathways, with subarachnoid metastases present in 33% of patients at diagnosis.¹ Extraneural metastases (ENM) occur in only 7% of cases, most often by hematogenous dissemination to bone or lymphatic dissemination to lymph nodes.^{1,2} Only 17 cases of MB metastasis to the peritoneum have been reported and CSF shunts were present in all 17 cases.² Yet the role of CSF shunts in ENM of primary CNS tumors remains controversial.^{1,5} We report the first case of concurrent ventriculoperitoneal (VP) shunt tract seeding and peritoneal metastasis of MB. The unique distribution and timing of ENM development presented in this case provide support for the theory that VP shunts are a conduit for MB spread.

Case Report

A 5-year-old Hispanic male presented with one month of nausea, vomiting and headache. Physical examination was normal. Brain and spine magnetic resonance imaging (MRI) showed hydrocephalus and a solid fourth ven-

tricular mass but no leptomeningeal metastases (Figure 1). Based on MRI appearance, medulloblastoma was the primary diagnostic consideration. Signal homogeneity and diffusion restriction along with absence of calcification, hemorrhage, and foraminal tumor extension distinguished the mass from ependymoma. Diffusion restriction and absence of a macrocystic component with mural nodule differentiated it from juvenile pilocytic astrocytoma. Patient age, midline tumor location, and absence of a cystic component argued against atypical teratoid-rhabdoid tumor.

Before primary tumor resection, a right frontal external ventricular drain (EVD) was placed for hydrocephalus. Surgical approach for primary tumor resection included suboccipital craniotomy and C1 laminectomy with microdissection of the cerebellar vermis and medullary tissue. Exposure of the fourth ventricle revealed a vascular, firm, rubbery tumor that was removed piecemeal using electrocauterization and forceps. The resection was incomplete due to tumor involvement of the fourth ventricle floor. Bone was replaced and secured with an absorbable cranium fixation system. Pathology showed MB with severe anaplasia (WHO grade IV).

Four days after tumor resection, the right frontal EVD was exchanged for a right frontal VP shunt. VP shunt surgery started with removal of the EVD and passage of the new ventricular catheter though the preexisting burr hole into the right lateral ventricle. The new catheter was clamped after noting clear flow of CSF. A galeal pocket was created. An abdominal incision was carried down to the subcutaneous fatty tissue. A shunt passer was passed from the abdominal incision to the cranial incision and the peritoneal tubing was threaded through the tract. The ventricular catheter and tunneled body wall tubing were connected to an ultra-small medium pressure medical valve yielding spontaneous flow of clear CSF at the abdominal end of the tubing. Sharp dissection and a trocar were used to enter the peritoneum. The catheter, dripping spontaneously with CSF, was inserted into the peritoneum. The abdominal and scalp wounds were irrigated and closed.

In postoperative week 3, the patient returned to the hospital with vomiting, headache and abdominal pain. Brain and spine MRI revealed interval development of diffuse smooth and nodular leptomeningeal enhancement of the brain and spinal cord, later proven by biopsy to represent leptomeningeal carcinomatosis. A shunt revision was performed for suspected shunt malfunction. The subgaleal pocket was opened. The intracranial portion of the shunt was found to be occluded and was replaced. The distal tubing was found to be patent. During the revision, the abdomen was opened and the abdominal portion of the shunt

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Key words: medulloblastoma, extraneural metastases, VP shunt, US, CT.

Contributions: DP, authored primary and a few subsequent drafts; reviewed literature; KRS, reviewed literature, contributed substantive edits; JMP, reviewed multiple drafts; contributed substantive edits; assisted with technical aspects of manuscript preparation; KLH, authored final draft.

Conflicts of interests: the authors declare no potential conflicts of interests. DP, speakers bureau for Philips Medical.

Received for publication: 29 September 2011.

Revision received: 18 February 2012.

Accepted for publication: 18 February 2012.

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Clinics and Practice 2012; 2:e37

doi:10.4081/cp.2012.e37

was dissected free to facilitate repositioning of the valve and shunt in the subgaleal pocket. The patient began chemoradiotherapy with six weeks of intravenous Vincristine and total neuraxis radiation (55.8 Gy to posterior fossa; 50 Gy to thecal sac).

In postoperative week four, the patient developed worsening headache. Brain MRI showed enlarged lateral and third ventricles. A second shunt revision was performed in a manner similar to the first, with re-opening of scalp and abdominal wounds and replacement of the obstructed intracranial portion of the shunt. Third ventriculostomy was performed and a third ventriculostomy was attempted in an effort to prolong the life of the shunt. Abnormal nodular distortion of third ventricular anatomy was noted. Biopsy of a third ventricle nodule revealed MB tumor. Third ventriculostomy was aborted due to obscuration of normal anatomy.

Twelve weeks after primary tumor resection, the patient returned with fever, nausea, vomiting, and a firm 4 cm right lateral thoracoabdominal wall mass adjacent to the VP shunt scar. Multiple CSF samplings grew coagulase-negative *Staphylococcus aureus*. CSF cytology was positive for medulloblastoma. Shunt explantation and EVD placement were performed. Sonography of the right thoracoab-

dominal wall showed a row of five heterogeneously hypoechoic masses (Figure 2). The masses had internal and peripheral Doppler flow. On contrast-enhanced abdominopelvic computed tomography (CT), the thoracoabdominal wall masses had high-attenuation and were oriented in a craniocaudal direction along the prior VP shunt tract. Peritoneal nodules, thickened and heterogeneously attenuating omentum, and bone marrow hypodensities were also present, consistent with peritoneal and bone marrow metastases (Figure 3). Surgical biopsy with immunohistochemical staining of the largest abdominal wall mass revealed a malignant small blue cell tumor, identical to the primary anaplastic medulloblastoma. Iliac crest bone marrow biopsies were also positive for MB. Concurrent CSF infection resolved with intravenous vancomycin, and a third VP shunt was placed. The

patient was discharged on a new systemic maintenance chemotherapy regimen (intravenous vincristine, cisplatin, and cyclophosphamide). Due to progression of metastatic disease, he was referred to hospice 5 months after diagnosis and expired soon after.

Discussion

ENM of primary CNS tumors is uncommon, with incidence in autopsy series of less than 0.5%.² MB is the most likely pediatric CNS tumor to result in ENM,² with bone being the most common site, followed by lymph nodes, liver, lung and muscle.¹ There are reports of subcutaneous MB growth at surgical scars in the head and neck and of MB skin metastases in patients without prior surgery.² Peritoneal

spread of MB is rare with only 17 reported cases.² One case of MB metastases to peritoneum and a lumboperitoneal shunt tract has been reported.⁶ MB metastases to a shunt tract without documented peritoneal metastases have also been described.⁵ To our knowledge, we report the first case of concurrent metastases of MB to a VP shunt tract and peritoneum.

Causative and permissive factors of ENM of primary CNS tumors have been proposed, including tumor cell anaplasia, tumor friability and propensity to metastasize in CSF, and tumor ability to invade leptomeninges and blood vessels. Iatrogenic disruption of the blood brain barrier during CNS surgery is also implicated in ENM and is thought to provide tumor cells with direct vascular and lymphatic access.^{2,3} The role of VP shunts in ENM remains controversial, due in part to the rarity

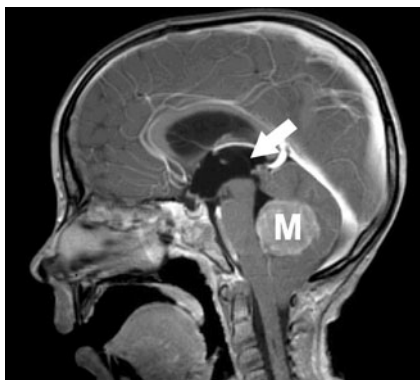


Figure 1. Sagittal post-gadolinium fat saturated T1-weighted brain magnetic resonance imaging at presentation shows an enhancing fourth ventricular mass (M) and an enlarged third ventricle (arrow). Lateral ventricles were also enlarged (not shown).

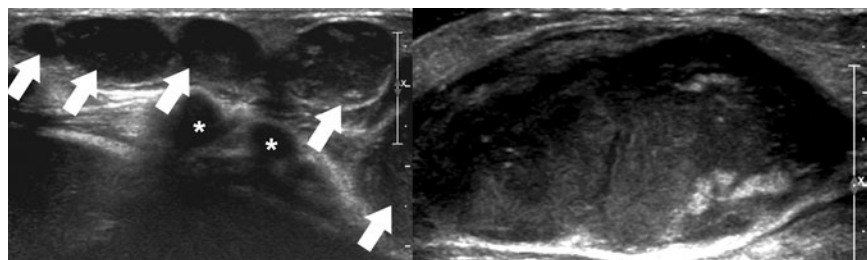


Figure 2. Contiguous sagittal sonographic images of a palpable mass in the right anterolateral thoracoabdominal wall 12 weeks after primary tumor resection show longitudinal arrangement of five heterogeneously hypoechoic medulloblastoma nodules (arrows; biopsy-confirmed) along a ventriculoperitoneal shunt tract. The shunt had been removed. Distribution of the nodules suggests shunt-related tumor implantation. *Rib

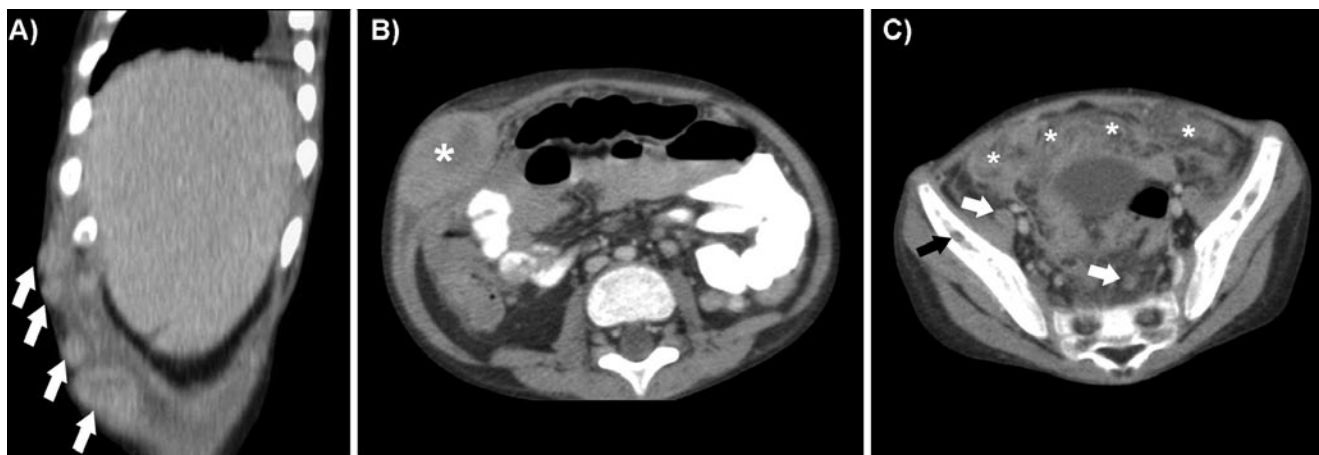


Figure 3. Contrast-enhanced abdominopelvic computed tomography 12 weeks after primary tumor resection. (A) Sagittal image shows linear arrangement of four medulloblastoma tumor implants in the thoracoabdominal wall (arrows), corresponding in location to prior ventriculoperitoneal shunt tract. (B) Axial image shows the largest abdominal wall metastasis (*) splaying right-sided anterior abdominal wall muscles. (C) Omental cake (*), nodular peritoneal tumor implants (white arrows), and bone marrow metastasis in the pelvis (black arrow; biopsy-confirmed) are present concurrently.

of ENM of primary CNS tumors and in part to inherent difficulties in establishing a causative relationship between VP shunts and ENM.¹⁻⁵ Most cases of ENM of primary CNS tumors occur in the absence of CSF shunts.^{2,3} When ENM occurs in the presence of VP shunts, tumor spread is attributed to the shunt in only one third of patients.⁴ Establishing a relationship between VP shunts and ENM is usually based on the pattern of metastatic disease in relation to shunt type and location.^{2,4} Development of shunt tract metastasis has been suggested as the most specific finding of direct tumor spread attributable to a shunt.⁴

Of the 17 reported cases of pediatric peritoneal MB metastases, all occurred in patients with VP (16) or lumboperitoneal (1) shunts.² To our knowledge, there is no report of peritoneal MB metastases in the absence of a peritoneal shunt, implying that shunts are a necessary permissive factor. ENM is also associated with concurrent leptomeningeal spread of MB.¹ MB metastases to lung are three times more common in patients with ventriculoatrial shunts than in patients without such shunts.⁵ Additionally, pediatric ENM develop earlier and are associated with poorer outcomes in shunted patients than in non-shunted patients.²

Linear arrangement of implants along the thoracoabdominal portion of a VP shunt tract and at no other cutaneous or subcutaneous location favors the argument that the shunt was not only a permissive factor but also a conduit for MB dissemination. Seeding of the tho-

racoadominal wall likely occurred by CSF spillage or cellular implantation during shunt placement or revision. In contrast, peritoneal tumor seeding could have occurred at any time during shunt utilization by direct cell migration through the shunt. Development of bone marrow metastases is less clearly linked to the shunt and rather suggests concurrent hematogenous tumor spread. Hematogenous spread requires disruption of the blood brain barrier and could have resulted from CNS surgery, CSF shunting or direct venous invasion.^{2,3} In previously reported cases of shunt-related peritoneal MB, over 40% had concurrent bone or lymph node metastases.²

Although metastases along VP shunt tracts are rare, radiologists, oncologists, and surgeons should be aware of their potential. They may occur in the absence of intracranial tumor recurrence.² Differential diagnosis includes inflammatory mass and abscess, which likely would be distinguished by fever, leukocytosis, and physical findings of inflammation. Our patient had both shunt infection and metastatic disease, but his thoracoabdominal wall masses lacked fat stranding on CT that is typical of an inflammatory process, and internal Doppler flow within his masses was inconsistent with abscess. CSF pseudocyst, seroma, and lymphocele could occur along a shunt tract but are differentiated by their cystic appearance and lack of internal flow. Adenopathy and other primary tumors are less likely considerations. As in our case, imaging might provide important clues to the etiology of disease. Most

importantly, a linear arrangement of masses in the neck or thoracoabdominal wall in proximity to a shunt or shunt tract is highly suggestive of shunt-related metastases. Tissue pathology is necessary for definitive diagnosis.

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