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## Case Report

# Anaplastic ependymoma metastases through a ventriculoperitoneal shunt

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## ABSTRACT

Ependymomas are rare glial tumors that comprise 10% of intracranial pediatric malignancies. Primary central nervous system malignancies can rarely metastasize extracranially. When metastases occur, it usually does so in the setting of surgical manipulation of the central nervous system and can spread through the blood, lymph, or artificial means, for example, a ventriculoperitoneal shunt. We describe the presentation and progression of an 18-month-old boy diagnosed with an ependymoma. Initially managed with surgery, radiation, and ventriculoperitoneal shunt placement for symptomatic hydrocephalus, the tumor later recurred with drop metastasis to the thoracic spinal cord. The patient subsequently developed extensive metastases within the abdominal cavity, which were seeded through a ventriculoperitoneal shunt. We present a case of a rare complication of intra-abdominal tumor seeding and carcinomatosis from an intracranial ependymoma through a ventriculoperitoneal shunt. This is a rare presentation of a possible complication, which requires awareness of both surgeons and radiologists.

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## Introduction

Ependymomas are rare central nervous system (CNS) neoplasms. Even though they are the third most common CNS tumor in children, they only comprise around 10% of pediatric intracranial tumors [1,2]. Tumors typically arise from the roof of the fourth ventricle; ependymomas within the spinal cord are less common. The World Health Organization (WHO) classifies ependymomas into four groups based on histologic

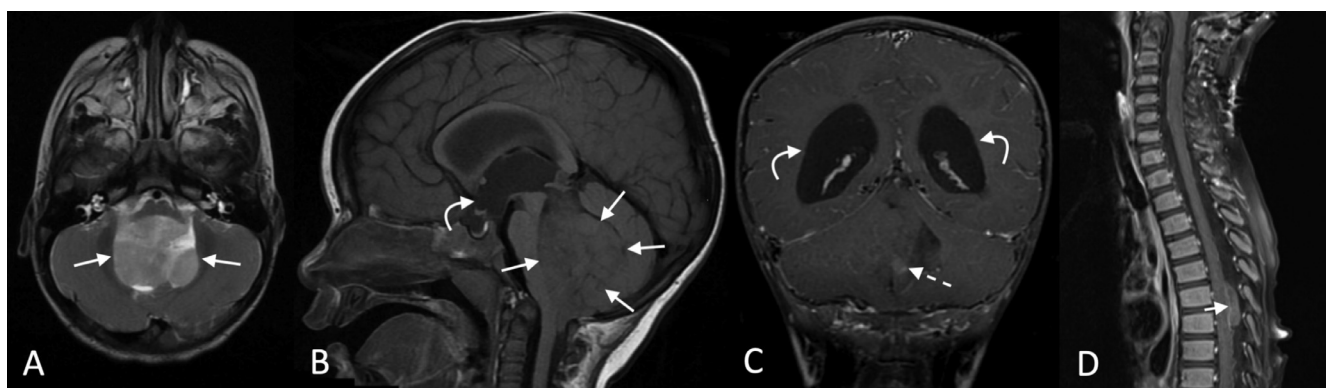
appearance: subependymoma (WHO grade I); myxopapillary ependymoma (WHO grade I); ependymoma with cellular, papillary, and clear cell variants (WHO grade II); and anaplastic ependymoma (WHO grade III) [3,4]. Regardless of the subtype of ependymoma, surgery is the mainstay treatment [2,3]. Extracranial metastases from CNS neoplasms are rare, and usually occur in the setting of surgery at the site of the neoplasm. If given access to systemic circulation, glial tumors are capable of seeding other organs in the body such as the lung, liver, and lymph nodes [5–7]. Tumor seeding via a ventriculoperi-

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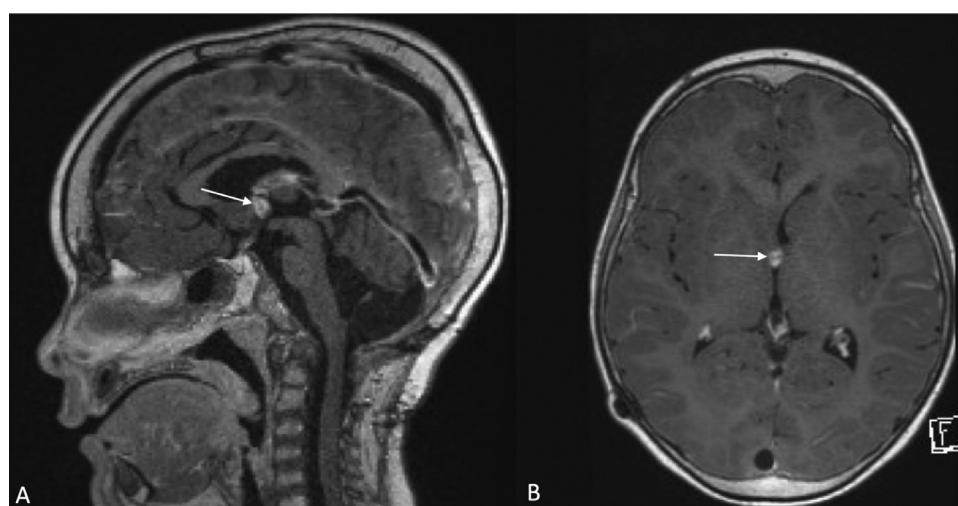
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**Fig. 1** – Axial T2 weighted (A), sagittal T1 (B), and contrast-enhanced coronal T1 (C) images show a large heterogeneous mass filling and expanding the fourth ventricle (arrows). There is associated hydrocephalus (curved arrows, B and C). Faint intralésional enhancement is present (dashed arrow, C). A subsequent examination of the spine demonstrated a drop metastasis dorsal to the cord in the upper thoracic spine (arrow, D).



**Fig. 2** – Sagittal (A) and axial (B) contrast-enhanced images demonstrate a new focus of enhancement near the foramen of Monro (arrows). The ventricles are slightly asymmetric, left larger than right, but remain decompressed due to the presence of a ventriculoperitoneal shunt (intracranial component not shown). Postoperative changes are seen in the posterior fossa from the primary surgical resection.

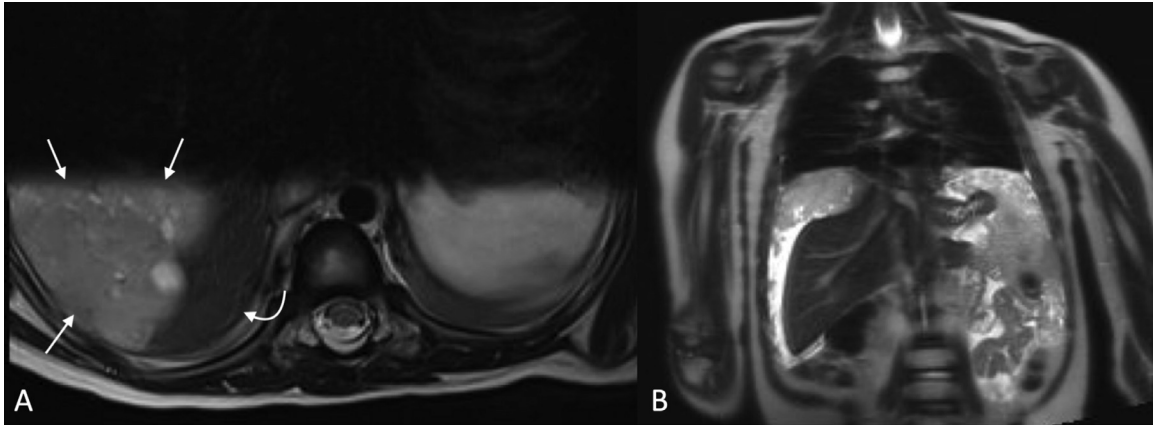
toneal shunt is a very rare complication. We searched using the medical subject heading terms: ependymoma AND ventriculoperitoneal AND metastases OR metastatic. This search produced 17 articles in PubMed [8-24].

### Case report

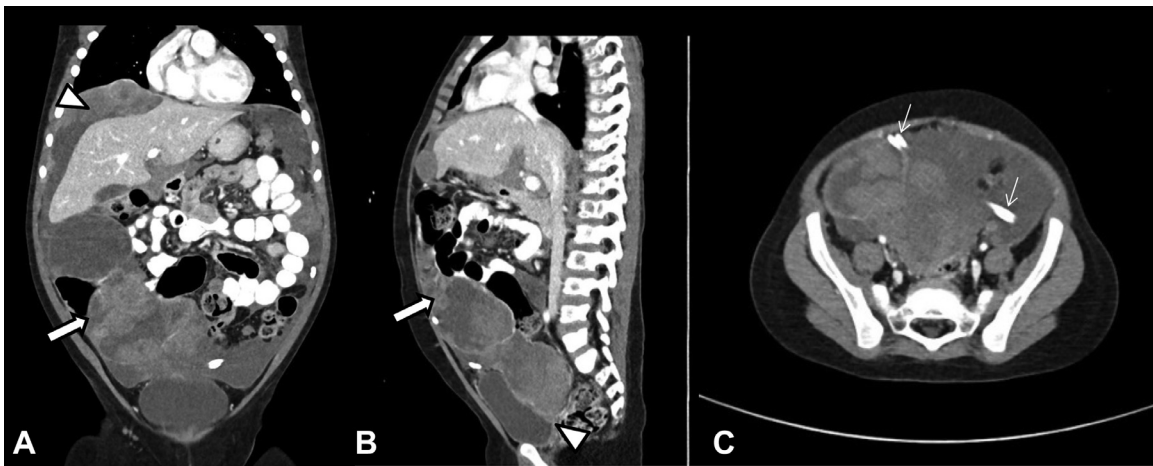
The patient presented at 18 months of age with a history of unsteadiness noted for approximately a month with fussiness and emesis, gradually worsening over time. He was initially evaluated at an outside institution, where he was found to have a large posterior fossa tumor (Fig. 1). He was then transferred to our center for further care in December 2015. The patient underwent primary resection and a course of radiation

therapy to the brain. Pathological workup revealed an anaplastic ependymoma (WHO grade III). Postoperative magnetic resonance imaging (MRI) showed no evidence of residual disease. In April 2016, the patient developed hydrocephalus for which a ventriculoperitoneal shunt was placed.

Repeat brain and spine MRI in May 2017 showed recurrent brain disease and a thoracic cord drop metastasis (Fig. 2). The patient underwent surgical resection of the spinal metastasis the same month, with pathology confirming anaplastic ependymoma (WHO grade III). Cerebrospinal fluid studies also returned positive for malignant cells. He was readmitted the following week with vomiting, generalized malaise, pain, lethargy, and listlessness. Computed tomography (CT) imaging showed new slit-like ventricles, and he underwent a shunt revision for presumed over-shunting, a complication of ventriculoperitoneal (VP) shunts in which excessive cerebrospinal



**Fig. 3 – Axial T2 (A) image of the thoracic spine demonstrated a soft tissue mass in the right upper quadrant (arrows), compressing the adjacent liver (curved arrow). Moderate ascites was noted throughout the abdomen on the localizer image (B).**



**Fig. 4 – (A and B) Coronal and sagittal contrast-enhanced CT of the abdomen and pelvis shows diffuse carcinomatosis with heterogeneous low attenuation masses (thick arrows) throughout the peritoneal cavity causing mass effect on the liver and bladder (arrowheads). (C) Axial contrast-enhance CT of the pelvis shows ascites along with peritoneal implants (arrowheads). Note the patient's ventriculoperitoneal shunt (thin arrows).**

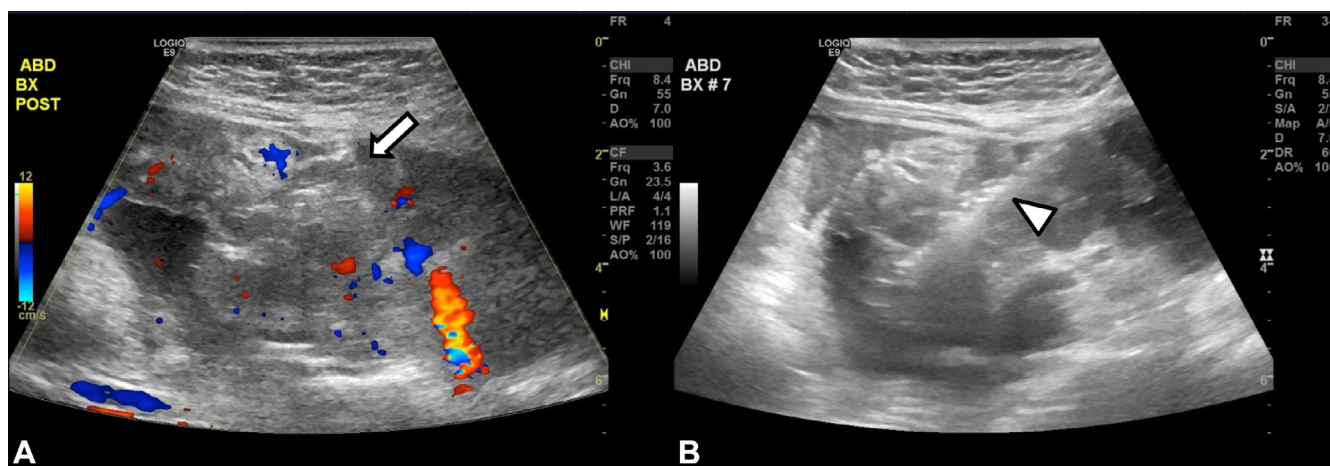
fluid drainage results in intracranial hypotension and ventricular collapse, in June 2017. The patient then completed another round of radiation to the spine and brain in August 2017, and remained stable until October 2019. He then began having episodes of emesis that were not accompanied by abdominal pain. Repeat brain MRI demonstrated a new lesion in the foramen of Monroe causing slight ventricular asymmetry (Fig. 3). He underwent resection of the mass; cerebrospinal fluid studies at that time were negative for malignancy.

In December 2019, the patient presented with abdominal enlargement. During his workup, a spinal MRI incidentally found an enlarging lobular right upper quadrant mass and ascites (Fig. 4). Subsequent CT of the abdomen and pelvis with intravenous contrast material showed diffuse peritoneal carcinomatosis with small-moderate volume ascites and thoracic lymphadenopathy (Fig. 5). An ultrasound-guided biopsy was performed and was positive for anaplastic ependy-

moma (Fig. 6). In January 2020, the patient underwent intra-abdominal tumor debulking surgery with bilateral diaphragm stripping in preparation for further chemotherapy.

## Discussion

Extracranial metastasis of CNS tumors can occur through several proposed mechanisms: hematogenous spread by means of primary tumor vessels or after tumor invasion of the dural veins, hematogenous, and/or lymphatic spread after invasion of the skull and extracranial soft tissues, and one of the more controversial mechanisms of spread via ventriculoatrial and ventriculoperitoneal shunts [3,25]. Surgical manipulation seems to have an important role as a risk factor for tumor metastasis [3,25]. It is thought that surgery dis-



**Fig. 5 – (A) Transverse ultrasound image of the right lower abdominal quadrant with color Doppler shows the heterogeneous mass (arrow) along the peritoneal lining as seen on CT. (B) Grayscale ultrasound image shows the peritoneal mass with the biopsy needle device within the mass (arrowhead).**

rupts the normal protective anatomy of the CNS, allowing malignant cells to access the blood and lymphatic vessels [3,26]. It is important to keep this in mind with malignancies such as ependymomas, in which surgical resection is the standard treatment modality, and post-operative management often requires additional surgeries and VP shunt placement [27].

Extracranial metastases of CNS tumor have been reported to occur in approximately 0.5% to 1% of cases, with ependymomas comprising 3.7% of such metastases in the pediatric population [3,27]. Around 27% of primary CNS malignancy extracranial metastases were directly related to placement of ventriculoperitoneal shunts [27]. The most common sites of extracranial metastasis of ependymomas are the lungs and lymph nodes. This suggests spread via hematogenous and lymphogenous mechanism and supports the idea that surgical manipulation and damage to structures in the CNS increases the risk for metastasis [27]. Our patient presented with a complex history of multiple surgeries and VP shunt placement with revision, both of which served as risk factors for metastatic disease. However, because the vast majority of extracranial metastases were within the omentum and diffusely throughout the abdomen and pelvis, the patient's ventriculoperitoneal shunt was presumed to have served as the pathway for metastatic spread. The possibility of spread, although rare, should be considered in patients with known history of CNS malignancy, particularly in those with shunted hydrocephalus and abdominal complaints.

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