

Case – Intra-abdominal metastases following ventriculoperitoneal shunt insertion for primary intracranial germ cell tumor

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Cite as: McAlpine K, Clark R, Jiang M, et al. Case – Intraabdominal metastases following ventriculoperitoneal shunt insertion for primary intracranial germ cell tumor. *Can Urol Assoc J* 2022 April 11; Epub ahead of print. <http://dx.doi.org/10.5489/cuaj.7895>

Published online April 11, 2022

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Case report

A 20-year-old male was seen at a Canadian academic hospital for recurrence of his previously treated germ cell tumor (GCT). His past medical history was significant for a diagnosis of intracranial non-germinomatous germ cell tumor (NGGCT) at the age of 17 years old. At the time of his initial diagnosis, he had symptoms of fatigue, polydipsia, dizziness, headaches and diplopia.

On magnetic resonance imaging (MRI) he was found to have three lesions including one in the pituitary fossa, one in the fourth ventricle and one in the pineal gland. The diagnosis of metastatic intracranial NGGCT was made based on characteristic MRI findings and elevated tumor markers including a human chorionic gonadotropin (hCG) of

Key Messages

- Ventriculoperitoneal shunts used to alleviate obstructive hydrocephalus can facilitate peritoneal seeding of intracranial germ cell tumors.
- Surveillance imaging for patients with intracranial germ cell tumors should include the path of the shunt used to alleviate obstructive hydrocephalus, especially in the setting of elevated germ cell tumor markers.
- A multi-disciplinary approach to treatment is necessary for patients with peritoneal seeding of primary intracranial germ cell tumors to attempt to render them disease-free.

3425 IU/L. No biopsy, surgical resection or imaging of the chest, abdomen, pelvis and scrotum was performed.

He required a ventriculoperitoneal (VP) shunt insertion to alleviate symptomatic obstructive hydrocephalus. The shunt functioned well while he received 6 cycles of chemotherapy (carboplatin and etoposide alternating with ifosfamide) and 54 Gray radiation to treat the intracranial GCT. Upon completing chemo-radiotherapy, he was followed by the pediatric neuro-oncology and radiation oncology teams. His follow-up imaging showed excellent response to therapy with no evidence of disease recurrence.

Unfortunately, 17 months after completing therapy, he was found to have an elevated hCG at 44 IU/L. He underwent MRIs of the head and spine, a lumbar puncture and a scrotal ultrasound. No evidence of recurrent GCT was detected. His hCG continued to rise and was 14,885 IU/L at his next appointment. His other tumor markers were also elevated with an alpha fetoprotein (AFP) of 15 ug/L and lactate dehydrogenase (LDH) of 3,906 U/L.

He reported an increasing abdominal girth and urgently underwent computed tomography (CT) scans of the chest, abdomen and pelvis. These found an extensive number of soft tissue masses throughout the abdominal cavity. The masses were in atypical locations for GCTs as they had a more peritoneal-based than nodal-based distribution. There were also multiple enlarged lymph nodes in the thorax. He was seen urgently by the adult medical oncology team who specialize in treatment of patients with testicular GCTs and started immediately on salvage chemotherapy with four cycles of paclitaxel, ifosfamide and cisplatin (TIP).

It was felt that he had a poor prognosis, given the extent of the disease and the level of tumor marker rise and chemotherapy was provided urgently with the understanding that surgical resection of any remaining lesions after chemotherapy would be necessary for a chance at cure. His tumor markers normalized rapidly on chemotherapy (hCG <1 IU/L, AFP 6.8 ug/L, LDH 159 U/L). Re-staging CT scans showed a dramatic decrease in the size and number of metastatic lesions. The lymph nodes in the thorax resolved and were all <1 cm after chemotherapy. The metastatic lesions in the abdomen also drastically decreased in size (Figure 1).

He was referred to the surgical oncology team (urology, general surgery and gynecological oncology for possible peritoneal stripping) to consider resecting the remaining lesions. The surgical teams recommended a diagnostic laparoscopy to assess the extent of the disease and to determine if complete resection with limited morbidity was possible. The patient consented and a diagnostic laparoscopy was performed. Images from the laparoscopy are shown in Figure 2.

The intra-operative opinion was that the extent of disease was resectable, however adjuvant procedures were potentially necessary including a bowel resection and/or partial cystectomy. The proposed operation was explained to the patient and informed consent was obtained. The patient returned to the operating room for a laparotomy 3 weeks later. The procedure involved a resection of all visible metastatic lesions, a retroperitoneal lymph node

dissection and a resection of the sigmoid colon which was tethered to one of the lesions. The surgical pathology of all 16 masses and 44 lymph nodes resected was negative for malignancy showing fibrosis/necrosis only. The patient had a prolonged post-operative recovery due to a bowel leak requiring a diverting colostomy and removal of his VP shunt leading to an intraventricular hemorrhage.

He was recently seen in clinic, 12 months post-operative, with no evidence of disease recurrence (negative CT chest/abdomen/pelvis, negative MRI head, normal tumor markers) and a good functional recovery.

Discussion

Primary intracranial germ cell tumors (GCT) are a rare and heterogeneous group of malignancies¹. Approximately 1% of pediatric primary brain tumors are GCTs with the median age at diagnosis of 16 years old¹. Intracranial GCTs are divided into germinomas and non-germinomas with non-germinomas including yolk sac, embryonal, teratoma and choriocarcinoma similar to non-seminomatous testicular GCTs. The diagnosis of intracranial GCTs can be challenging and is delayed in more than 50% of patients due to the non-specific symptoms and rare incidence of the disease^{2,3}. Upon diagnosis, more than half of patients require treatment for symptomatic obstructive hydrocephalus⁴. Where appropriate based on anatomy, endoscopic third ventriculostomy is recommended to alleviate obstructive hydrocephalus in order to limit the morbidity associated with external ventricular drainage or open surgery^{5,6}. When a ventriculostomy is not possible, external ventricular drainage is preferred over placement of a permanent ventricular shunt to avoid the risk of seeding to other cavities in the body⁷. Treatment of intracranial GCTs typically involves chemotherapy and radiation^{1,2}.

In the case presented in this report, the patient was initially treated for metastatic primary intracranial NGGCT. A VP shunt was placed to alleviate symptomatic obstructive hydrocephalus. The patient received chemotherapy and radiotherapy with curative intent and did well with normalized tumor markers and excellent radiographic response. The development of intraabdominal metastatic GCT lesions was unexpected.

Seeding of primary brain tumors is a known risk of shunts used to alleviate hydrocephalus and has been described in previous case reports and case series⁶. Unfortunately, intracranial GCTs are the most frequent primary pediatric intracranial malignancy associated with shunt-related metastases⁶. The average length of time from treatment of intracranial NGGCT and the development of shunt-related metastatic disease is 14 months which was in keeping with the clinical course of our patient⁶.

Unfortunately, the prognosis of patients with shunt-related metastases from intracranial NGGCT is poor⁶. Salvage chemotherapy in the setting of elevated tumor markers is appropriate given the presumed systemic disease. In our patient, the residual masses measuring >1 cm following an excellent response to chemotherapy were treated surgically, similar to residual tumors post-chemotherapy for non-seminomatous testicular cancer.

Peritoneal seeding of primary intracranial GCT is a known risk of VP shunt insertion for obstructive hydrocephalus. For these patients, a key point is that even though surveillance imaging of the primary tumor post-treatment would not include imaging the abdomen, it is important to monitoring the path of the VP shunt with imaging and physical examinations to detect metastatic disease, especially in the setting of rising tumor markers. It is possible that alternative means to alleviate hydrocephalus may help prevent shunt-related metastases for these patients who are often being assessed and treated by the neurosurgical and oncology teams. Treatment of residual tumor in the peritoneal cavity following normalization of tumor markers with salvage chemotherapy may be a reasonable treatment option similar to the management of non-seminomatous testicular cancer, however limited data is available to inform this practice. A thorough, and multi-disciplinary approach is necessary to ensure these patients receive the best care possible.

Conclusions

Peritoneal metastases following insertion of a VP shunt to alleviate cancer-related obstructive hydrocephalus is a known risk and follow-up imaging should be altered to include the shunt drainage path. Patients who are found to have extra-neural spread of primary brain cancers require a multi-disciplinary approach to attempt to render them disease free. Urologists and surgical oncologists should be aware of this group of patients and the possible role for surgical resection following salvage chemotherapy, however long-term outcomes have not been reported.

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Figures and Tables

Figure 1. Coronal computed tomography (CT) scan images of peritoneal metastases along course of ventriculoperitoneal shunt (A) before and (B) after salvage chemotherapy. Arrows indicate metastatic lesions.

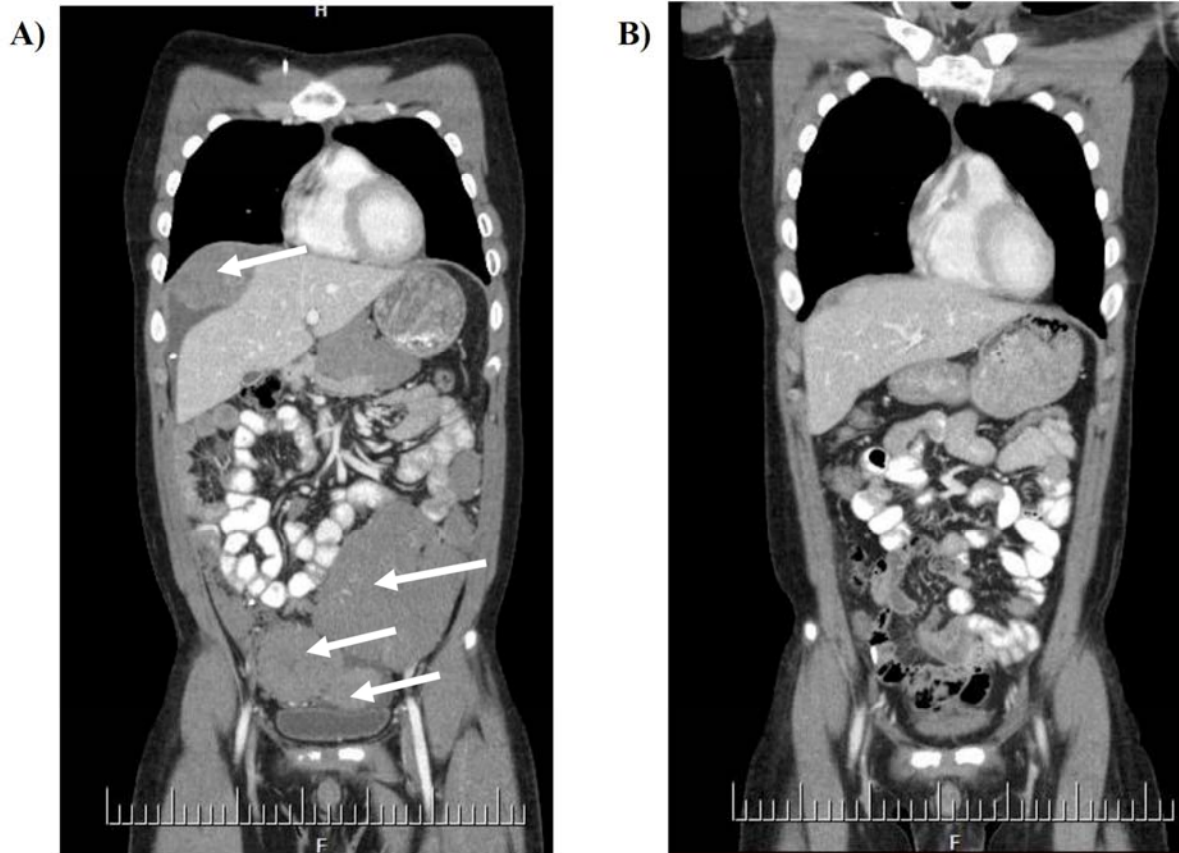


Figure 2. Images from diagnostic laparoscopy showing multiple metastatic lesions in the peritoneal cavity including (A) one in close proximity to the sigmoid colon, (B) several along the length of the ventriculoperitoneal shunt (B), (C) including in the pelvis. Metastatic lesions indicated by arrows.

