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Case Report

Metachronous Intraabdominal and Liver Metastasis in a Treated Case of Suprasellar Germinoma Due to Ventriculoperitoneal Shunt

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Abstract

A rare, treated case of suprasellar Germinoma with a ventriculoperitoneal shunt *in-situ* with metachronous presentation of extensive intraabdominal and hepatic metastasis successfully treated with combination chemotherapy. We report a case of a 14-year-old boy, previously treated for suprasellar Germinoma with a ventriculoperitoneal shunt *in-situ*, who subsequently presented 3 years later with acute severe abdominal pain and anuria. He developed extensive extraneural metastasis, intraabdominal and liver metastasis, acute renal failure and hyperuricemia. The patient was successfully treated with chemotherapy and continues to be disease free after 35 months. Despite its rare occurrence, we emphasize a low threshold for abdominal imaging to be an included investigation at follow up visits of such patients to ensure timely detection.

Keywords: Germinoma; Intra-Abdominal; Ventriculoperitoneal Shunt; Liver Metastasis

Introduction

The incidence of Extraneural Metastasis (ENM), that is any involvement of the primary Central Nervous System (CNS) tumours beyond usual sites of brain, meninges or spinal cord is very rare, ranging from 0.1-3.7%. [1]. The common causes are Medulloblastoma and Germinoma in paediatric population [2]. While germinomas have favourable outcomes, the risk of obstructive hydrocephalus is high, especially in pineal GCT, due to interventricular foramen compression. This requires insertion of a Ventriculoperitoneal [VP] shunt to alleviate the high intracranial pressure. Though germinomas have the propensity to spread via cerebrospinal fluid during diagnosis, migration of malignant tumour cells via VP shunt resulting in ENM is very rare with only few case reports and case series reported. [3]. We discuss a rare case of an acute presentation of VP shunt related extensive intrabdominal metastatic disease with liver metastasis. We also highlight the challenges in delivering palliative chemotherapy in a patient who has already received prior anti-cancer therapy further complicated by additional acute renal failure.

Case Report

14-year-old boy was diagnosed as suprasellar CNS Germinoma and underwent a trans ventricular biopsy with placement of Right frontal Medtronic inline medium-pressure VP shunt for hydrocephalus. Cerebrospinal Fluid (CSF) malignant cytology was clear. Serum and CSF Alpha Feto Protein (AFP) and Beta Human Chorionic Gonadotrophin (beta HCG) levels were normal. He received 4 cycles of combination chemotherapy with Etoposide and cisplatin followed by radiation (40Gy, 21#). Regular follow up Magnetic Resonance Imaging (MRI) Brain with perfusion did not show any disease.

27-months later he presented with 5-day history of acute severe abdominal pain, anuria, acute renal failure (Creatinine 1.49 mg/dl {0.7-1.20}) and hyperuricemia (11.1 mg/dl {3.4-7.0}). Contrast Enhanced Computed Tomography (CECT) abdomen showed a 12.73 x 9.25 cm heterogenous midlines mass in the pelvic cavity distal with compression of the distal 1/3rd of the ureters bilaterally causing bilateral hydroureteronephrosis and subcapsular 6.34* 6.0 9cm right segment VII liver lesion. Pelvic mass biopsy was positive for SALL4, Pan CK, CD99, NKX2.2, SATB2 and Desmin negative, Ki67- 30%, suggestive of germ cell tumor. MRI Brain and spine were clear. Serum AFP level was 4.62ng/ml, beta HCG 209.3 mIU/ml and LDH 290 U/L. Positron Emission Tomography Computed Tomography (PET CT) whole body scan revealed a large coalescent pelvic mass alone with multiple discrete and coalescent necrotic omental, peritoneal, serosal and liver surface deposits, upper abdominal and anterior diaphragmatic lymph nodes. He was initially planned to be given palliative 3 weekly combination regimen of ifosfamide, paclitaxel and cisplatin. However, in view of acute renal failure, cisplatin was avoided and instead he received carboplatin (dose calculated based on glomerular filtration rate and area under the curve; modified TIP based chemotherapy). Patient underwent bilateral DJ stenting for obstructive uropathy. We faced multiple challenges while delivering chemotherapy, due to co-existing acute renal failure and obstructive uropathy (due to the tumor mass), resulting in increased chemotherapy related toxicities and cycle delays. He was thus initiated on modified weekly regimen (three doses of weekly chemotherapy equivalent to 1 cycle). He developed recurrent episodes of complicated neutropenia despite receiving the weekly chemotherapy regimen which precluded us from switching to once in 3- weekly regimens even after resolution of renal failure. He received a total of 4 complete cycles of chemotherapy. End of treatment PET CT scan demonstrated a complete metabolic response and patient continues to remain event free after 35 months (Fig. 1).

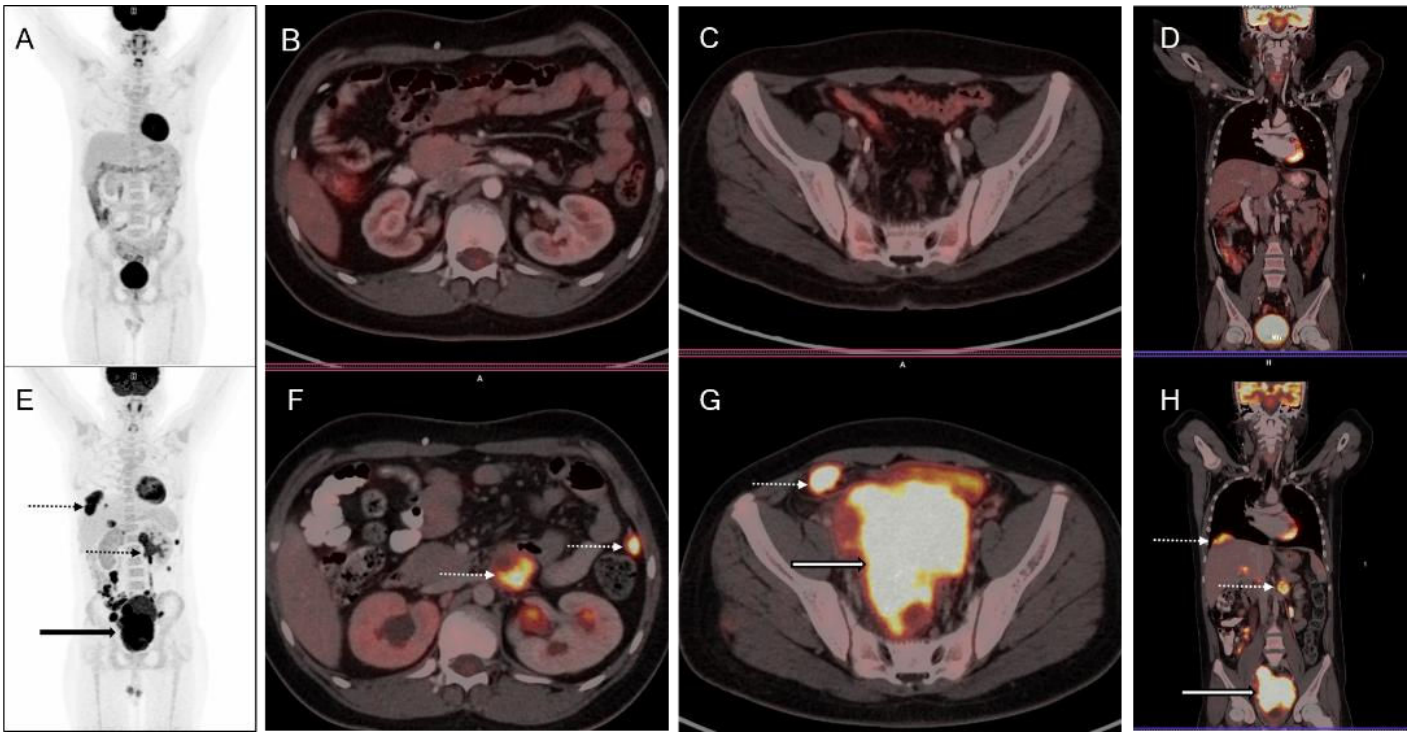

Figure 1: Baseline whole-body FDG PET-CT scan (E-H) in a known case of CNS Germinoma demonstrates a large coalescent mass (arrow) in the pelvis compressing rectum and urinary bladder with focal loss of fat planes, along with multiple FDG avid discrete and coalescent necrotic omental, peritoneal, serosal and liver surface deposits (dotted arrow). Follow-up whole body PET-CT scan (A-D) done after 08 weeks demonstrates interval resolution of these lesions suggestive of complete metabolic response to treatment.

Discussion

Due to the high sensitivity to chemotherapy and radiotherapy, the 10- year survival rates in CNS Germinomas are over 90 percent [4]. Tumour dissemination to the extrameningeal tissue can occur due to craniotomy or during VP shunt placement. VP shunts are placed very commonly to help relieve hydrocephalus and location of shunt tip is a potential site of dissemination [2]. The exact mechanism of spread is still unknown. A literature review by Xu, et al., reported 25 cases of germinoma out of 106 cases of primary intracranial tumors causing ENM, with a 26 months median time to metastasis [5]. Most cases (92 out of 106 cases)

reportedly had either residual or recurrent disease at the time of shunt placement. Only 1 such case of shunt related ENM in a completed resected patient has been reported [5]. It remains unclear which patients with a shunt will ultimately go on to develop shunt related ENM. The common sites of metastasis reported were 17% neural, 4% each of bone and lymph nodes and visceral in all 100% (25/25 cases). The average reported overall survival is 20 months from shunt placement (range from 17 to 84 months) and 56% of the population were alive, underscored its better outcomes compared to other primary CNS tumors, despite being the commonest [2,5]. The previously used Ventriculo-Atrial (VA) shunts were notorious for their short mean time to ENM and short survivals due to its direct connection to systemic circulation, are rarely used today. In our case the CSF cytology at the time of shunt was negative for malignant cells.

A possible alternative to VP shunt is Endoscopic Third Ventriculostomy (ETV) for patients with obstructive hydrocephalus. [6]. There is increasing evidence supporting ETV as a superior option with lower morbidity, shunt failure and chances of ENM [7-9].

Our patient presented with a large heterogenous coalescent pelvic mass along with extensive abdominal and liver metastasis, 27 months after receiving standard of care treatment with chemotherapy and radiotherapy for an initial diagnosis of primary CNS germinoma. There is very limited literature on imaging findings of peritoneal dissemination and only 2 case reports suggest presentation as multiple heterogeneously enhancing large intraabdominal masses as this case [4,9].

Conclusion

This case represents a rare presentation of a combination of acute renal failure due to obstructive uropathy and large peritoneal and liver metastasis in a previously completely treated case of CNS Germinoma. In previously pretreated patients, it can be challenging to readminister such high doses of chemotherapy requiring dose frequent modifications. We emphasize, while it is rare to have intrabdominal metastasis via VP shunt, awareness of this complication allows having a low threshold for abdominal imaging, especially when a patient presents with subtle symptoms. It may be worthwhile performing an abdominal ultrasound during regular follow up MRI Brain imaging in patients with a VP Shunt.

Conflict of Interests

The authors have no conflict of interest to declare related to this article.

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Informed Consent

Written consent was taken from the patient and his guardians.

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