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Case Report

Peritoneal metastasis of a cerebellar medulloblastoma through a ventriculoperitoneal shunt: A case report ☆

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ABSTRACT

Medulloblastoma is a frequent and aggressive pediatric tumor. It causes intracranial hypertension, necessitating ventriculoperitoneal shunting with surgical resection. Intraperitoneal metastases are rare and result from the migration of neoplastic cells through the shunt and into the peritoneal cavity. This metastatic form involving the ventriculoperitoneal shunt has a poor prognosis, making therapeutic management even more difficult. We report the case of a 14-year-old boy with a history of medulloblastoma of the cerebellum who was initially treated with complete resection of the tumor with placement of a ventriculoperitoneal shunt, followed by radiotherapy and chemotherapy, with good progression until he presented to the emergency department with acute abdominal symptoms. Imaging revealed multiple peritoneal masses with intra- and retroperitoneal lymphadenopathies. An ultrasound-guided biopsy revealed a metastatic medulloblastoma in the peritoneal cavity, and the patient underwent chemotherapy. The placement of the ventriculoperitoneal shunt in the tumor dissemination is therefore to blame.

Introduction

Medulloblastoma is a frequent tumor entity in children: it is a primitive neuroectodermal tumor involving the cerebellum and the fourth ventricle [1]. The mode of discovery is most often intracranial hypertension requiring imaging, metastatic localization in the peritoneal cavity is rare, even exceptional, the classic therapeutic approach associates a first surgical removal with a ventriculo-peritoneal shunt, followed by radiotherapy of the brain [1].

The ventriculo-peritoneal shunt is incriminating in the diffusion of tumors to the peritoneal cavity, which creates great difficulties of treatment and offers a poor prognosis [2].

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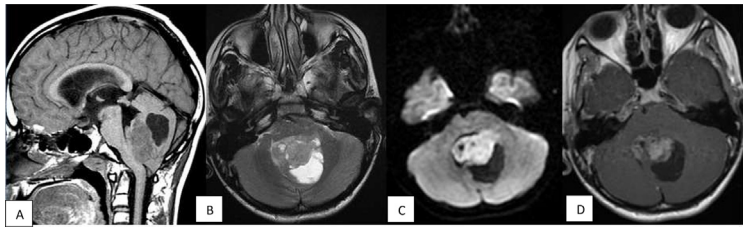

Fig. 1 – Encephalic MRI: Sagittal T1 (A), Axial T2 (B) showed a vermian tumor, hypoisignal on T1 hypersignal on T2 and on diffusion (C) enhancing after the injection of gadolinium (D).

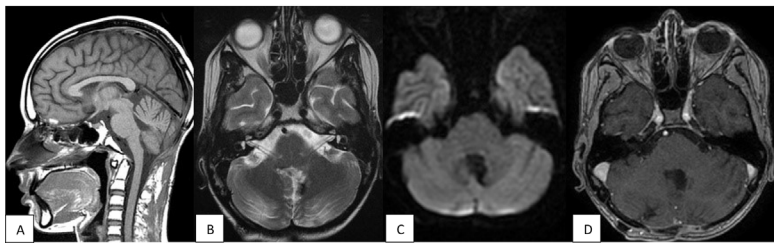

Fig. 2 – Encephalic MRI: Sagittal T1 (A), Axial T2 (B) diffusion (C) injection of gadolinium (D) at the end of the treatment showing no evident residual tumor.

Case presentation

We report the case of a 14-year-old boy with no previous pathological history who presented to the emergency department with intracranial hypertension, consisting of headaches and decreased visual acuity. Investigation by cerebral CT and cerebral and spinal MRI (Fig. 1) revealed a vermian tumoral process with upstream hydrocephalus and transependymal resorption of cerebrospinal fluid.

Imaging results suggested a medulloblastoma, which was confirmed by biopsy. The patient underwent surgery with complete resection of the tumor and placement of a ventriculoperitoneal shunt. Postoperative adjuvant chemotherapy and radiotherapy were then performed with good progression. MRI of the brain was performed 8 weeks after the end of treatment (Fig. 2) and revealed no tumor residue. Four months later, the patient presented to the emergency department with abdominal pain; clinical examination revealed an altered child with polypnoea and discolored conjunctiva; abdominal examination showed marked abdominal tenderness in the right iliac fossa; the biological evaluation was disturbed, with C-reactive protein at 100 mg/L and white blood cells at 13,000/yl. Abdominal ultrasound showed multiple peritoneal masses and intra- and retroperitoneal lymphadenopathies.

A neck, thorax, abdomen-pelvis (NCAP) CT scan was then performed, showing multiple lymphadenopathies on both sides of the diaphragm, pleural effusion with thickening, and multiple peritoneal masses, as well as ascites.

We then suspected either a peritoneal metastatic recurrence of his medulloblastoma, or a lymphoma. An ultrasound-guided peritoneal biopsy was performed, and the anatomopathological study showed a secondary localization of the medulloblastoma, so the patient began chemotherapy.

Discussion

Medulloblastoma is a primitive neuroectodermal tumor located in the cerebellum and fourth ventricle. It is the most common pediatric brain tumor. It accounts for 40% of cerebellar tumors, 15%-20% of all brain tumors, and is the leading cause of cancer-related death. It's the leading cause of malignant brain tumors in children [1], whereas it accounts for less than 1% of brain tumors in adults [2].

Most cases are sporadic, unless there are specific syndromes that predispose to tumor development, such as Gorlin's syndrome [3] or mutations in the SUFU gene [4]. Extracerebral metastases are exceptional.

The most frequent site of intraperitoneal localization was the liver (13%), the pancreas came second (4%, followed by the kidneys (2%), ureter (1%), and ovaries (1%) [5]. It should be noted that treatment is mainly surgical, combined with radiochemotherapy.

On the other hand, the placement of a ventriculoperitoneal shunt (VPS), which is an artificial conduit allowing communication between the central nervous system and the abdominal cavity in order to avoid an increase in intracranial pressure in the event of hydrocephalus, also represents a potential route for abdominal dissemination [6] . Migration of tumor glial cells through the CSF explains tumor implantation on the peritoneal surface, even in the absence of intracranial recurrence [7].

Between 18% and 27% of medulloblastomas present secondary localizations via the shunt route [8].

However, as not all patients with ventriculoperitoneal shunts present with peritoneal metastases, the hypothesis of immunological deficiency as possible cause of tumor implantation in the peritoneal cavity has been put forward, which explains the low number of published cases [8].

The therapeutic management of cerebellar medulloblastoma is based on complete surgical resection followed by radiotherapy, with doses of 35-36 Gy, and intensification on the surgical bed at 55 Gy [9] adjuvant chemotherapy delays the occurrence of tumor recurrence [9]. In our patient, the fact that the secondary localization occurred in the absence of any intracerebral recurrence suggests that dissemination may have occurred immediately after shunt placement, even before radiotherapy was performed [6].

Conclusion

Medulloblastoma is a very aggressive tumor of the posterior fossa, it can cause endocranial hypertension, which requires placement of a ventriculoperitoneal shunt after surgical resection. However, it is possible that neoplastic cells found in the cerebrospinal fluid may migrate through the shunt and implant in the peritoneal cavity, producing multiple metastases.

Patient consent

Written informed consent for the publication of this case report was obtained from the legal tutor of the patient.

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