Child's Nervous System
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CASE REPORT

Intramedullary metastasis in medulloblastoma: a case report and literature review

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Abstract

Background Cerebellar medulloblastomas are the most common malignant tumors of the posterior fossa in childhood that frequently metastasize. Leptomeningeal dissemination and distant metastasis have been associated with medulloblastomas; however, intramedullary metastases are rare with very few case reports in the literature available.

Methods We present a case of a 3-year-old girl with a medulloblastoma who underwent surgical resection of spinal intramedullary metastases. Histopathology revealed the tumor to be an anaplastic medulloblastoma similar to the intracranial lesions. The patient subsequently underwent postoperative chemotherapy followed by radiotherapy.

Results Following the surgery and subsequent follow-up, the patient showed a good recovery without any new neurological dysfunction.

Conclusions Intramedullary metastasis of medulloblastoma remains a rare disease. Surgical resection could play a possible role in the management in addition to radiation and chemotherapy.

Keywords Medulloblastoma · Metastasis · Chemotherapy

Introduction

Medulloblastoma is a highly malignant central nervous system tumor commonly seen in children, accounting for approximately 20% of all primary intracranial tumors [1]. It frequently spreads along the neuroaxis by leptomeningeal seeding through the cerebrospinal fluid (CSF) pathways. However, intramedullary spinal medulloblastoma metastasis throughout the central canal of the spinal cord is particularly rare; only few case reports are available in the literature.

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In this report, we present a case of a 3-year-old girl who developed intramedullary metastasis from a cerebellar medulloblastoma, and review the existing literature.

Case

History and examination

A 30-month-old girl with a history of nausea, vomiting, and ataxia underwent a ventriculoperitoneal shunt and resection of the posterior cranial fossa tumor in May 2019 owing to severe obstructive hydrocephalus and cerebellar medulloblastoma located in the fourth ventricle (Figs. 1, 2). Whole-spine magnetic resonance imaging (MRI) and cytological analysis of the CSF revealed no dissemination. The tumor had been removed completely, and the pathological examination established the diagnosis of a medulloblastoma. The postoperative period was uneventful.

Over the following 5 months, the patient received nine courses of regular chemotherapy with vincristine and carboplatin at an institution outside. Since the patient was too young, no radiation therapy was prescribed. Despite adjuvant therapy, the patient developed a slowly progressive

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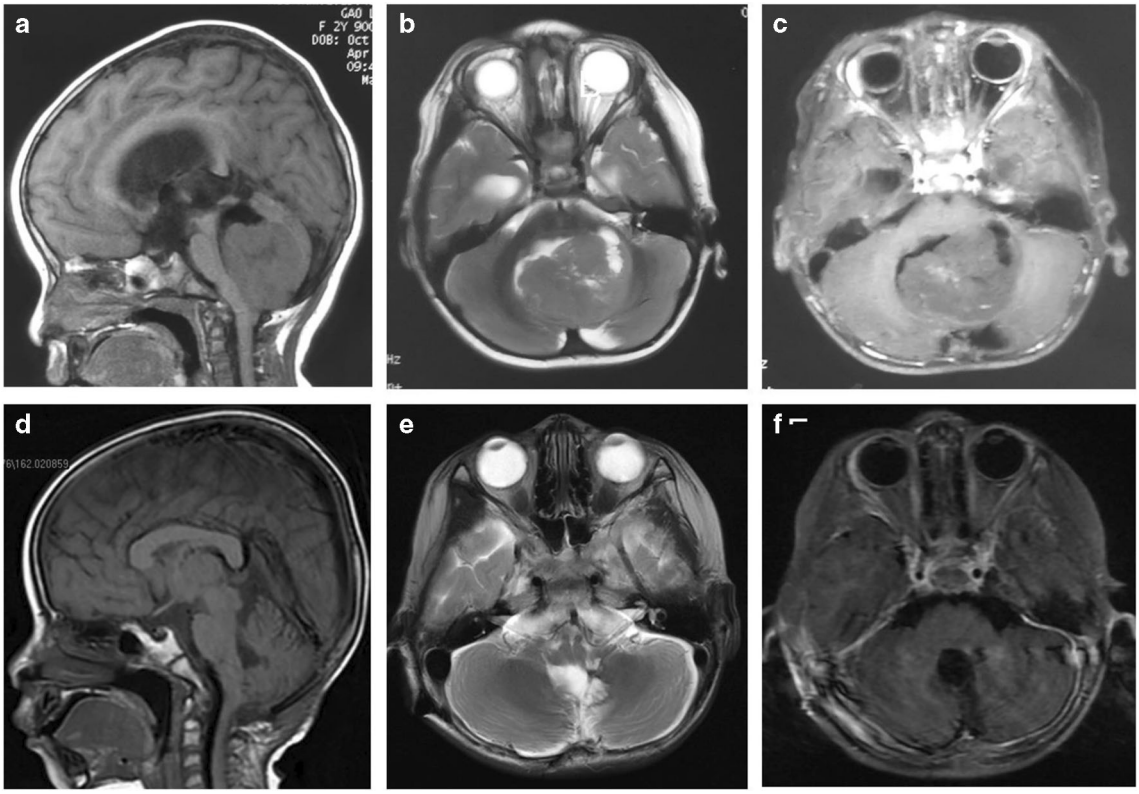

Fig. 1 Pre- and postoperative magnetic resonance imaging (MRI) scans of this case with cerebellar medulloblastoma. MRI showed long T1-weighted and short T2-weighted appearance located in the fourth ventricle (**a**, **b**). Sagittal contrast-enhanced MRI scan showed slightly uneven enhancement (**c**). On admission (5 months of follow-up), MRI scans showing total resection of the lesion without recurrence (**d**-**f**)

weakness in her left limb. Nausea, headache, and ataxia were absent; however, she complained of posterior neck pain with hyperextension. No bowel or bladder dysfunction was observed. She was admitted to our hospital for evaluation of these symptoms. Her neurologic exam revealed diminished strength in the left deltoid 4-, biceps 4-, and triceps 4-. The MRI revealed an obviously thickened and enlarged cervical spinal cord from C7 to T2. The spinal cord lesion measured 18 mm in height, 10 mm in the antero-posterior dimension, and 10 mm in width. The upper and lower central canals of the lesion were significantly enlarged, showing central cyst-like cavity changes. Following the injection of the gadolinium-

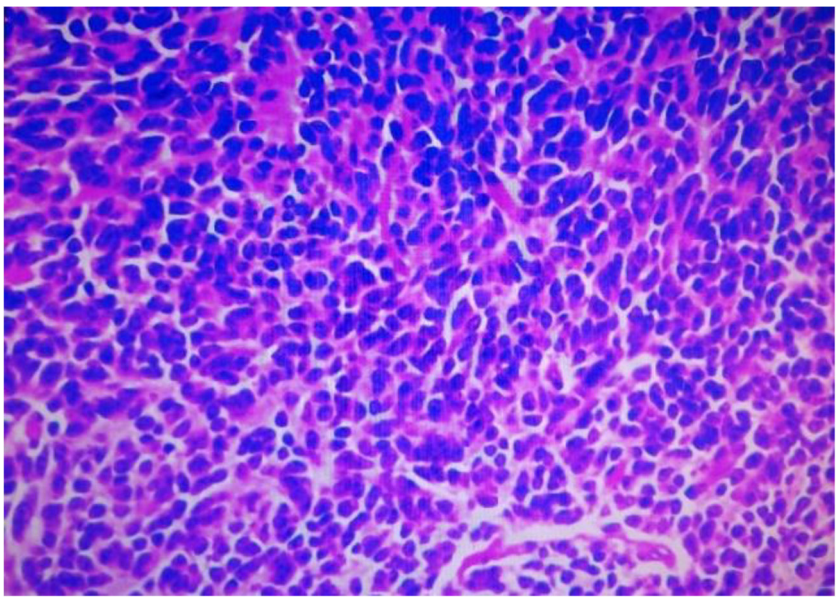

Fig. 2 Pathological examination of the fourth ventricular tumor (hematoxylin–eosin stain, original magnification ×400). It revealed a classic medulloblastoma, WHO grade IV. The cells had round to oval hyperchromatic nuclei and undefined cytoplasm. Immunohistochemical staining revealed a Ki-67 proliferation index of 10–30%

diethylenetriamine penta-acetic acid contrast agent, the lesion appeared unevenly enhanced; however, an obvious enhancement area was observed (Fig. 3) in the distribution area of the L3 horizontal cauda equina nerve.

Surgery

A total laminectomy of C7/T1/T2 was performed on November 4, 2019. A midline myelotomy was performed for the resection of the intramedullary mass with electrophysiological monitoring. The solid reddish-purple and gray tissue was encountered within the spinal cord substance, without a capsule. The boundary with the surrounding spinal cord tissue was unclear (Fig. 4a). Best efforts were made to resect the tumor, and intraoperative frozen pathological examination revealed the lesion to be a high-grade malignant tumor. A postoperative pathological diagnosis of desmoplastic/nodular medulloblastoma, WHO IV was formulated (Fig. 4b).

Postoperative course

A temporary decrease in the muscle strength of both lower limbs was observed following surgery. Over the following 40 days, the patient received radiation treatment. The total administered radiation dose was 3600 cGy to the craniospinal axis (total 20 times), a 1440 cGy boost to the fourth ventricle site (total 8 times), a 900 cGy boost to the C6-T4 area (total 8 times), and a 900 cGy boost to the L2-4 area (total 8 times). After a short break of less than a month, she received additional nine regular chemotherapy with vincristine and carboplatin. No change in her neurological status was observed during the follow-up.

Discussion

Medulloblastoma is one of the most common malignant tumors in children. It often spreads throughout the subarachnoid space. Intramedullary metastasis with medulloblastomas is very rare.

Based on the review, only seven cases (two adults and five pediatric) had been published in the English literature to date [2–5]. Zumpano et al. were the first authors to report a case of intramedullary medulloblastoma metastasis [3]. Since then, sporadic reports have emerged in the literature. In the reported cases, chemoradiation or biopsy followed by chemotherapy is the recommended management strategy.

In this patient, intramedullary metastasis occurred late in the course of the disease. At first, she received only regular chemotherapy. No radiation therapy was initiated since the patient was less than 3 years old. Her condition was very stable at the beginning, and evidence of intramedullary metastasis was observed after half a year.

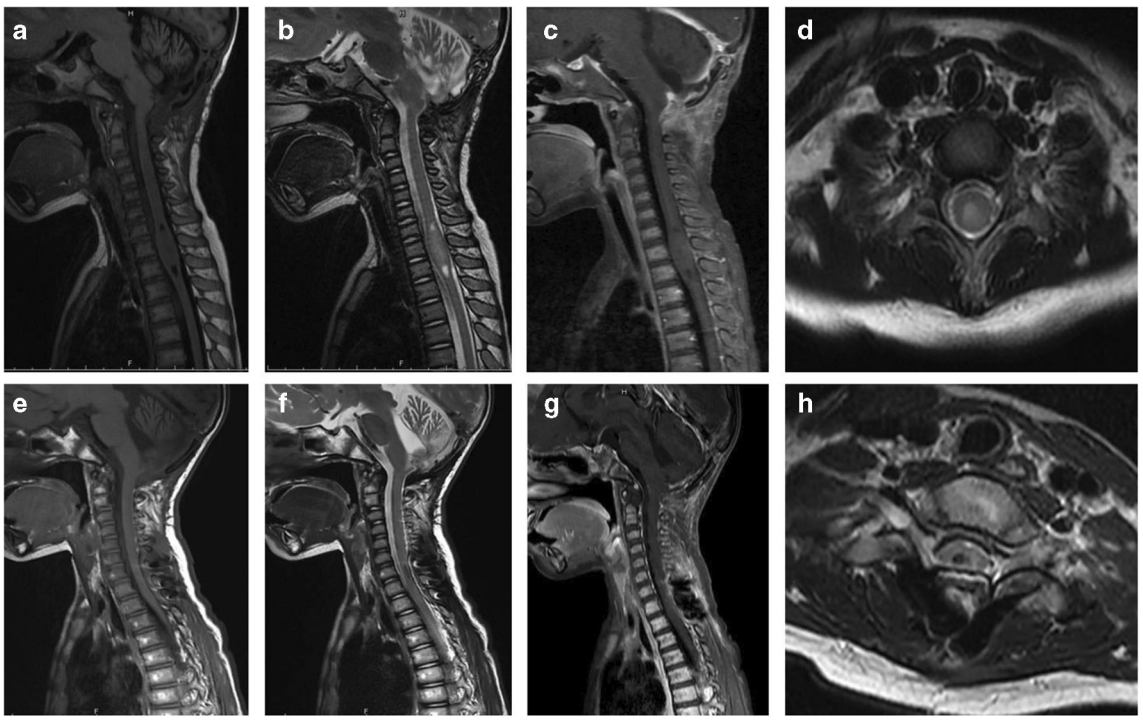

Fig. 3 Pre- and postoperative magnetic resonance imaging (MRI) scans of this case with intramedullary metastasis. Sagittal images revealed long T1-weighted and short T2-weighted appearance located in the level of cervical-7 to thoracic-2 of the spine (**a**, **b**). Sagittal contrast-enhanced MRI scan showed uneven enhancement (**c**). Postoperative MRI scans showing total resection of the lesion without recurrence after 12 months of follow-up (**e**-**h**)

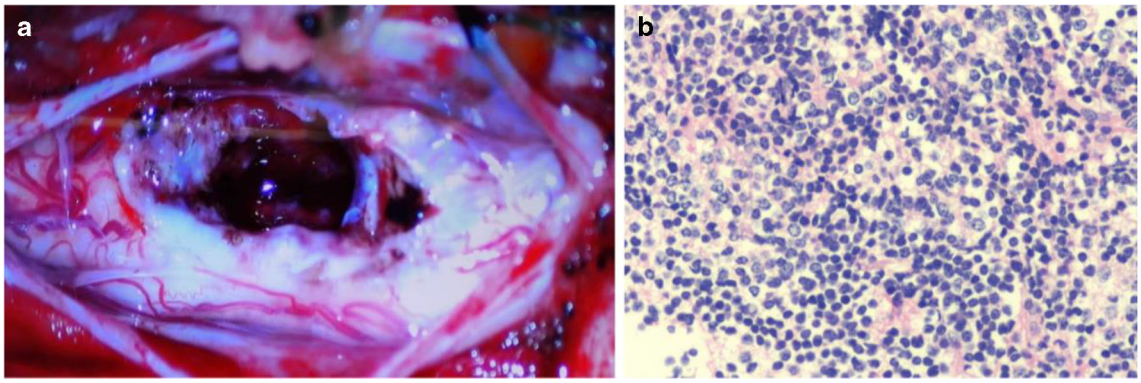

Fig. 4 Intramedullary tumors and postoperative pathology. Intraoperative picture (**a**) showed a residual cavity following tumor resection. The tumor was in the spinal cord. The boundary with the surrounding spinal cord tissue was unclear. Postoperative pathological examination from the intramedullary tumor (**b**) (hematoxylin–eosin stain, original magnification ×400). It revealed a classic medulloblastoma, WHO grade IV. It had the same pathological properties as tumors in the cerebellum

In general, the mode of metastatic spread appears due to local invasion and seeding of the CSF pathways. Leptomeningeal dissemination is common in medulloblastoma [4]. In this case, we also observed the lesion located in the conus-cauda region (Fig. 5). However, intramedullary metastases are very rare, and the mechanism of intramedullary spread of medulloblastoma remains a matter of speculation. In our patient, preoperative MRI clearly revealed that the lesion was seen within the substance of the spinal cord, and the central canal was significantly enlarged (Fig. 1). Postoperative pathological analysis confirmed its nature. Thus, it is likely to infer that the tumor had spread from the cerebellum through the central canal of the spinal cord.

Given the paucity of literature, the management of intramedullary metastases remains controversial. The case reports available thus far only suggest biopsy and chemotherapy with or without radiation [4, 6–8].

Based on the management strategy employed in this case, we observed that it is very important to surgically remove the intramedullary metastases as much as possible with the aid of electrophysiological monitoring. Surgery can provide a low recurrence rate and a reasonably low complication rate.

Pediatric patients are more sensitive to irradiation. For patients older than 3 years, radiotherapy should be considered in time following surgery [9]. In addition, chemotherapy is also necessary. Taylor et al. noted that postoperative chemotherapy combined with radiotherapy improved the prognosis compared with radiotherapy alone [10]. Chemotherapy was corroborated in our case, thus, reducing the radiation dose with tolerable side effects, which contributed to a good prognosis.

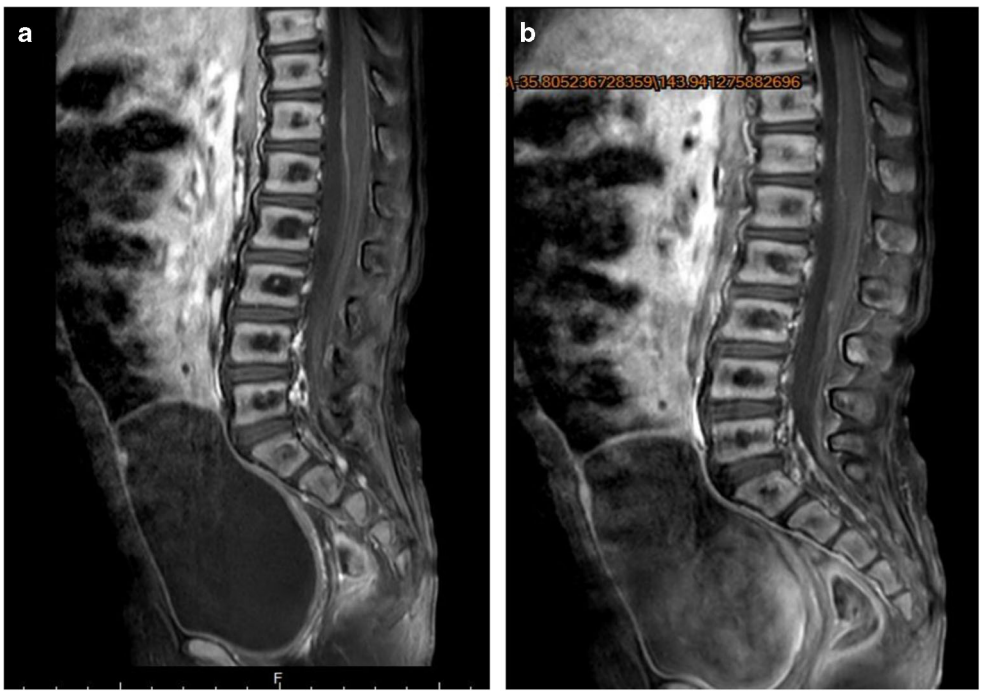

Fig. 5 Visible tumor coverage on the surface of the cauda equina plexus (**a**), postoperative magnetic resonance imaging (MRI) scans showing radiotherapy and chemotherapy of the lesion without recurrence after 12 months of follow-up (**b**)

Conclusion

A case of intramedullary spinal metastasis of a cerebellar medulloblastoma was presented. The spinal metastasis was mainly diagnosed based on contrast-enhanced MRI scans and clinical presentation. Surgical resection should be considered in order to promptly relieve spinal cord compression. In addition, regular radiotherapy combined with chemotherapy may contribute to a better prognosis for intramedullary metastasis in medulloblastoma. Further clinical research is warranted to confirm this possibility.

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Declarations

Conflict of interest The authors certify that they have no affiliations with or involvement in any organization or entity with any financial interest (such as honoraria; educational grants; participation in speakers' bureaus; membership, employment, consultancies, stock ownership, or equity interest; and expert testimony or patent-licensing arrangements), or non-financial interest (such as personal or professional relationships, affiliations, knowledge, or beliefs) in the subject matter or materials discussed in this manuscript.

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