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Case report

Medulloblastoma with suprasellar solitary massive metastasis: Case report



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ARTICLE INFO

Article history:

Received 2 December 2015

Accepted 6 February 2016

Available online 18 February 2016

Keywords:

Medulloblastoma

Metastasis

Suprasellar region

ABSTRACT

It is extremely rare for metastasised medulloblastoma to form a large tumour in the suprasellar region. We present a case of medulloblastoma with large suprasellar metastasis at initial presentation. A 3-year and 5-month-old boy presented with a 1-month history of vomiting and loss of appetite, and body weight. Computed tomography and magnetic resonance imaging revealed a 20 mm × 20 mm mass in the suprasellar region and a 30 mm × 30 mm mass in the fourth cerebral ventricle. We performed endoscopic biopsy of the suprasellar tumour, and subsequently totally removed the vermian tumour through a suboccipital craniotomy. The histopathological findings revealed that both the suprasellar and vermian tumours were classic type and non SHH/WNT type medulloblastoma. The postoperative course was uneventful. The patient showed complete remission after chemotherapy. The tumour in the suprasellar region was most likely metastatic from the vermis. Endoscopic biopsy of the tumour in the suprasellar region and total removal of the tumour in the vermis in a one-stage operation followed by intensive chemotherapy with reduced dose radiotherapy may provide a satisfactory outcome.

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1. Introduction

Medulloblastoma is the most common paediatric brain tumour with invasive and metastatic potential. The presence of metastatic disease in patients newly diagnosed with

medulloblastoma remains one of the most important prognostic factors [1]. Although leptomeningeal metastasis of medulloblastoma is frequent, suprasellar metastasis is uncommon [2–6]. We report here an extremely rare case of medulloblastoma that formed tumours of similar size in the suprasellar region and vermis.

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<http://dx.doi.org/10.1016/j.pjnns.2016.02.001>

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2. Case report

2.1. History and examination

A 3-year and 5-month-old boy presented with a 1-month history of vomiting and loss of appetite and body weight. The patient was originally healthy and had no developmental abnormalities. His only medical history was of bronchial asthma. Although headache and vomiting were observed, no other neurological abnormalities were evident on admission. Laboratory studies showed negative serum tumour markers, and endocrinological examination detected incomplete hypopituitarism. The posterior pituitary function was preserved. Thyroid hormone was replaced preoperatively. Ophthalmological examination showed no optic atrophy or papilloedema.

2.2. Imaging findings

Two tumours in the suprasellar region and cerebellar vermis were evident on screening computed tomography (CT). Cranial

CT revealed a 20 mm × 20 mm mass in the suprasellar region and a 30 mm × 30 mm mass in the fourth cerebral ventricle with mild ventricular enlargement. Small punctate calcifications were noted in the tumours. Enhanced CT showed homogeneous enhancement of the tumours without subarachnoid or intraventricular metastatic lesions (Fig. 1). Cranial magnetic resonance imaging (MRI) showed isointense tumours on T1 weighted, T2 weighted, and FLAIR images in both the suprasellar region and fourth cerebral ventricle. The size of the suprasellar tumour was 20 mm × 20 mm × 32 mm and the size of vermian tumour was 30 mm × 30 mm × 35 mm (Fig. 2a). Because the patient experienced an asthma attack during contrast-enhanced CT, gadolinium-enhanced MRI was not performed. No clear dissemination was apparent on the FLAIR image of either cranial or spinal MRI.

2.3. Operation and postoperative course

Endoscopic biopsy was performed on the suprasellar tumour and an indwelling ventricular drain was placed. Intraoperative pathological diagnosis of this tumour was not germinoma but

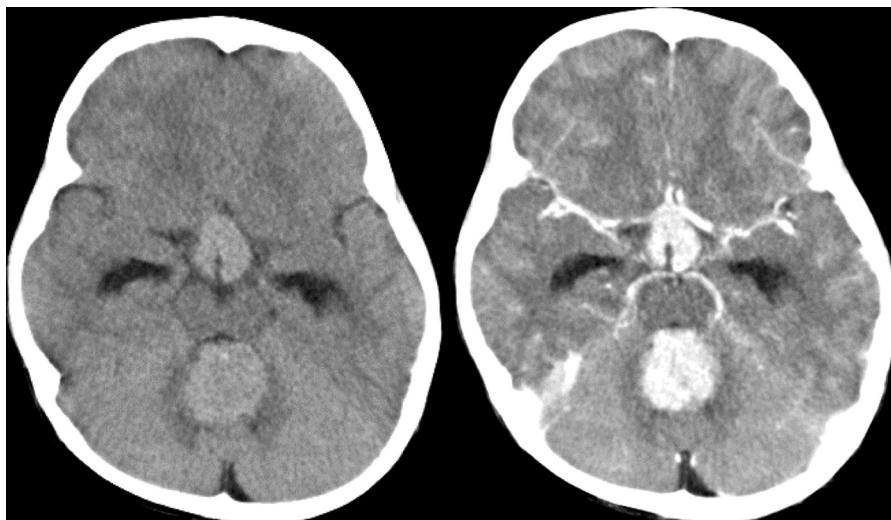


Fig. 1 – Preoperative images. Preoperative axial computed tomography scan showing tumours in the suprasellar mass and vermis, with mild hydrocephalus (Left). Homogeneous enhancement on contrast-enhanced images without evidence of dissemination (Right).

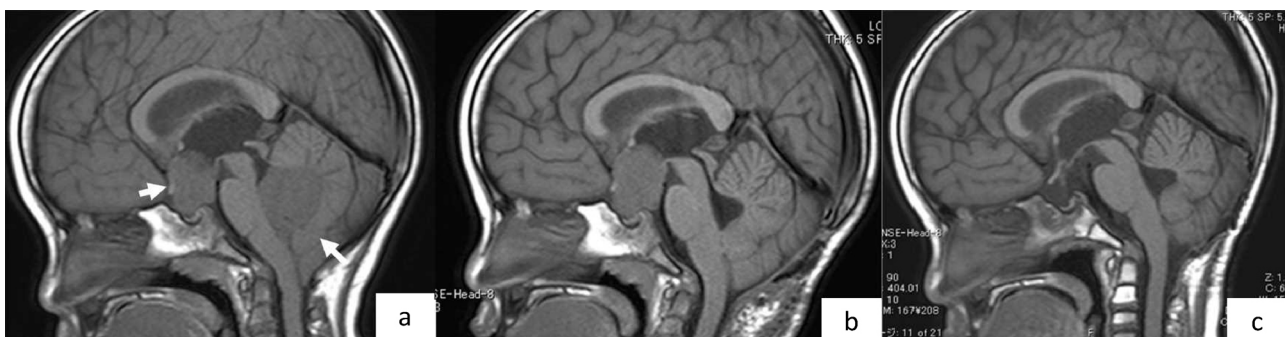


Fig. 2 – Sagittal magnetic resonance T1-weighted images obtained at the initial presentation. Arrows indicate homogeneous hypointensity in tumours in the suprasellar region and vermis (a). Image after removal of the tumour in the vermis (b) and following chemotherapy and radiation therapy, showing resolution of the tumour in the suprasellar region (c).

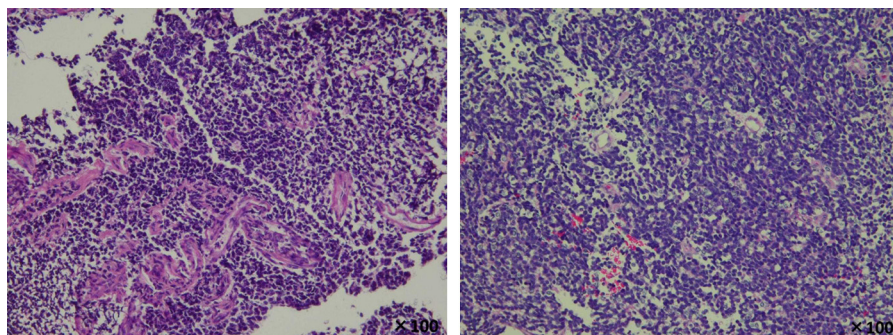


Fig. 3 – Photomicrographs of the tumours in the suprasellar region (Left) and vermis (Right). Similar histopathological features of classic medulloblastoma in each of highly dense oval shaped cells having a high N/C ratio, karyokinesis, and karyorrhexis. H&E, original magnify, both 100×.

embryonal tumour, such as medulloblastoma or primitive neuroectodermal tumour (PNET). The patient was then placed in the prone position and the vermian tumour was totally removed through a suboccipital craniotomy; the intraoperative pathological diagnosis was also medulloblastoma. Cerebrospinal fluid (CSF) cytology from the third ventricle was negative.

The postoperative course was uneventful and the extraventricular drain was removed on postoperative day 3. MRI showed no residual tumour in the vermis (Fig. 2b) and CSF cytology was negative for malignant cells.

2.4. Pathological findings

Histological findings confirmed a diagnosis of medulloblastoma, classic type (Fig. 3). A diffuse proliferation of oval blue cells with a high nuclear cytoplasmic (N/C) ratio, karyokinesis, and karyorrhexis were observed. Similar images were obtained for the suprasellar region and vermis. The immunostaining results for both tumours were BAF47 (+++), synaptophysin (+++), neurofilament (+++), GFAP (+++), S-100 (±), and a MIB1 index of 41.6%. The pathological diagnosis was embryonal tumour, WHO grade 4, classic medulloblastoma of the fourth ventricle or CNS PNET of the suprasellar region. The β -Catenin (–), Dickkopf-1 (DKK-1) (–), Yes-associated protein (YAP) (–), Secreted frizzled-related protein-1 (SFRP-1) (–), c-MYC (–). The finding suggest that the tumour was non sonic hedgehog (SHH)/wingless (WNT) type medulloblastoma.

2.5. Chemotherapy and radiation therapy

Chemotherapy was started at 21 postoperative days as stated in the protocol of the Japanese Pediatric Brain Tumor Consortium. Assuming this was a case of suprasellar metastasis from the primary vermian medulloblastoma, we categorised the case as high-risk based on the postoperative disease stage classification for medulloblastoma. After four chemotherapy courses (cyclophosphamide, cisplatin, vincristine, and etoposide), high-dose chemotherapy (thiotepa and melphalan) was performed together with autologous peripheral blood haematopoietic stem cell transplantation as salvage therapy. The patient received reduced-dose craniospinal irradiation (18 Gy) with a boost to the tumour bed (total dose, 50 Gy). The suprasellar tumour had regressed to achieve

complete remission during the first course of chemotherapy. Cranial and spinal MRI after completion of chemo-radiation therapy showed complete remission (Fig. 2c). No recurrence or neurological deficit has been evident as of 3 years after the operation.

3. Discussion

3.1. Clinical features

Medulloblastoma is the most common malignant brain tumour in childhood and has invasive and metastatic properties. These tumours account for 15–20% of all paediatric central nervous system neoplasms [7]. Medulloblastoma mostly occurs in boys aged 3–9 years and approximately 10–15% of cases are diagnosed in infancy [1]. The tumour occurs mainly in the cerebellar vermis and tends to invade the fourth ventricle and cerebellar hemispheres. The presence or absence of metastasis is important for postoperative treatment. Metastasis of medulloblastoma is common, and it is believed that seeding occurs mainly in the spinal subarachnoid space, basal cistern, Sylvian fissure, and in any subependymal lesions of the lateral ventricle or any subfrontal lesions [7]. Although several cases of medulloblastoma metastasising to the suprasellar region have been reported, the formation of a single large tumour at initial diagnosis has been rarely reported.

Metastasis to the suprasellar region has been documented in recurrent medulloblastoma, but bulky metastatic disease at presentation is rare [2–6]. Helton et al. reviewed 130 children with medulloblastoma and reported 6 cases (4.6%) of medulloblastoma metastatic to the anterior third ventricular recess at diagnosis [5]. The suprasellar metastases ranged in size from 4 to 11 mm. Fukusumi et al. reported 2 cases with a nodular metastatic focus in the suprasellar region [3]. Both cases were clinically occult and the tumours were smaller than in the present case. Shelton et al. presented a 32-year-old man with classic medulloblastoma that showed intraventricular spread via subarachnoid dissemination of the disease from the fourth ventricle to the suprachiasmatic recess of the third ventricle [6]. In addition, Lee et al. reported 3 cases of recurrent medulloblastoma within the fourth ventricle and a large

metastatic lesion in the suprasellar region (3 and 6 cm in diameter) with accompanying ependymal metastasis [8]. Gupta et al. reported an adult case of medulloblastoma with a large suprasellar mass in which the pathological diagnosis was classic medulloblastoma and CSF cytology was negative, as in our case [4]. To our knowledge, the present case is the first involving such a large metastatic medulloblastoma (approximately 20 mm) in the suprasellar region.

It is possible that our case represents vermian metastasis of a suprasellar PNET. PNETs typically occur in the cerebral hemisphere or lateral ventricle [9] and less typically in the suprasellar region. In addition, there have been no reports of vermian metastasis of suprasellar PNETs to date. Accordingly, in our case, metastasis of a suprasellar PNET to the vermis would be a less likely diagnosis. There is also the possibility that our case is a supratentorial PNET with a synchronous medulloblastoma. Multifocal medulloblastoma has previously been reported in 4 adult cases, all of which involved infratentorial lesions that were classic medulloblastoma histologically [10–13]. There have been no reports of cases in which medulloblastoma occurred in both the suprasellar region and vermis simultaneously. Therefore, in our case, simultaneous onset of PNET in the suprasellar region and medulloblastoma in the vermis would also be a less likely diagnosis. When cerebellar and suprasellar region tumours co-exist, even if there are no findings of metastasis on MRI or cerebrospinal fluid analysis, we should consider metastasis.

3.2. Treatment and outcome

Medulloblastoma is commonly treated with surgery, chemotherapy, and radiotherapy and the course decided depends on patient age, size of the residual tumour, and the presence or absence of dissemination [1,14]. There is no standard approach to treatment for tumours occurring simultaneously in the supratentorial and infratentorial regions [4–6]. In the present case, the tumours were considered to be malignant, so the operative approach was designed to enable chemotherapy to be started as early as possible. Endoscopic biopsy of the suprasellar tumour would provide a histological diagnosis without further hypothalamo-pituitary dysfunction caused by tumour removal; simultaneous endoscopic third ventriculostomy was impossible due to the presence of the massive third ventricle tumour; and total removal of the fourth ventricular tumour would provide a histological diagnosis and enable CSF circulation to be reestablished to avoid a shunt operation.

In this rare case of medulloblastoma with tumours of similar size in both the suprasellar region and vermis, the radiographic variation may be clinically relevant when considering the differential diagnosis from preoperative images. In similar cases, endoscopic biopsy of the tumour in the suprasellar region and total removal of the tumour in the vermis in a one-stage operation followed by intensive chemotherapy with reduced-dose radiotherapy may provide a satisfactory outcome.

Conflict of interest

None declared.

Acknowledgement and financial support

None declared.

Ethics

The work described in this article was conducted in accordance with the World Medical Association (Declaration of Helsinki) Ethical Principles for Medical Research Involving Human Subjects, and uniform requirements for manuscripts submitted to Biomedical Journals.

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