Spontaneous intratumoural and intraventricular haemorrhage associated with a pilomyxoid astrocytoma in the hypothalamic/chiasmatic region

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ABSTRACT

Pilomyxoid astrocytoma (PMA) is a rare, low-grade glioma that is recognised as a variant of pilocytic astrocytoma. There have been few reports on this pathologic entity presenting with spontaneous haemorrhage. In this study, we report a rare case of PMA in the hypothalamic/chiasmatic region presenting with intratumoural and intraventricular haemorrhage. An external ventricular drain was urgently inserted. A ventriculo-peritoneal shunt (VP) was undergone 4 weeks thereafter. The patient received fractionated Gamma Knife radiosurgery in another hospital 3 weeks after the VP shunt. Three months later, subtotal resection of the tumour was performed in our hospital via a pterional approach. The pathological diagnosis was PMA. Postoperatively, no adjuvant therapy was given, and the neurologic deficits were improved. However, the presentation of endocrine deficits remained. Notably, PMAs in the hypothalamic/chiasmatic region presenting with massive intratumoural and intraventricular haemorrhage may result in a severe condition and long-term impairment of endocrine function. Long-term follow-up is required to monitor the recurrence of the tumour and endocrinopathy.

1. Introduction

Pilomyxoid astrocytoma (PMA) is a rare, low-grade glioma that is recognised as a variant of pilocytic astrocytoma (PA). Tihan et al. described a series of patients with PMA, which showed histologically and clinically different features from PA, and cited the difficulty of gross total resection because the tumours are commonly located in the hypothalamic/chiasmatic regions [1]. The number of reports on PMA has recently increased, but there have been few studies on this pathologic entity presenting with spontaneous haemorrhage [2–4].

We report a rare case of PMA in the hypothalamic/chiasmatic region presenting with massive intratumoural and intraventricular haemorrhage and discuss the clinical features and treatment of this tumour with regard to the literature.

2. Case report

2.1. History and examination

A 13-year-old boy with sudden headache and vomiting presented to our hospital. He had severe visual impairment and became comatose immediately. Moreover, he presented with severe evidence of hypothalamic and pituitary dysfunction, including hypernatremia and diabetes insipidus. CT scan of the head showed massive intratumoural and intraventricular haemorrhage with marked hydrocephalus (Fig. 1A, B). MRI revealed a large lesion in the suprasellar region and enlarged ventricles with massive acute haemorrhage (Fig. 1C). An angiogram was obtained, which was negative for cerebrovascular abnormalities.

2.2. Treatment course

An external ventricular drain was urgently inserted. After this procedure, the general condition of the patient improved. Ventriculo-peritoneal (VP) shunt was performed 4 weeks thereafter. The patient's family refused the surgical resection of the lesion. The patient was transferred to another hospital, where he received fractionated gamma knife radiosurgery (GKRS) two sessions 3 weeks after the VP shunt; the marginal dose administered per session was 6 Gy. Therefore, the cumulative dose of 12 Gy was prescribed at the tumour margin. The prescription isodose was 45%. The patient's neurologic status deteriorated 3 months after the fractionated GKRS. An MRI study of the neuraxis revealed growth of the tumour size (Fig. 2A). There was no evidence of leptomeningeal and spinal dissemination. The patient was transferred back to our hospital for further surgical debulking of the tumour. Craniotomy tumour removal was performed via a pterional approach. The tissue extended into the third ventricle and involved the hypothalamus. Subtotal resection was performed to avoid damage to the hypothalamus.

2.3. Pathologic findings

A microscopic examination of the tissue showed a glial neoplasm composed of monomorphous, bipolar cells with a myxoid background. No Rosenthal fibres or eosinophilic granular bodies were detected. Immunocytochemistry showed an astroglial immunophenotype positive for glial fibrillary acidic protein (GFAP). The pathological diagnosis was PMA (Fig. 3A, B).

2.4. Postoperative course

Some neurologic deficits were obviously improved postoperatively. However, the endocrinopathy with hypopituitarism and diabetes insipidus remained. The patient required pituitary hormone replacement and treatment for diabetes insipidus. Follow-up MRI studies of the neuraxis obtained at 3 and 10 months after the resection indicated no evidence of tumour recurrence (Fig. 2B, C). There was also no evidence of leptomeningeal and spinal dissemination. No adjuvant therapy was given after the sur-

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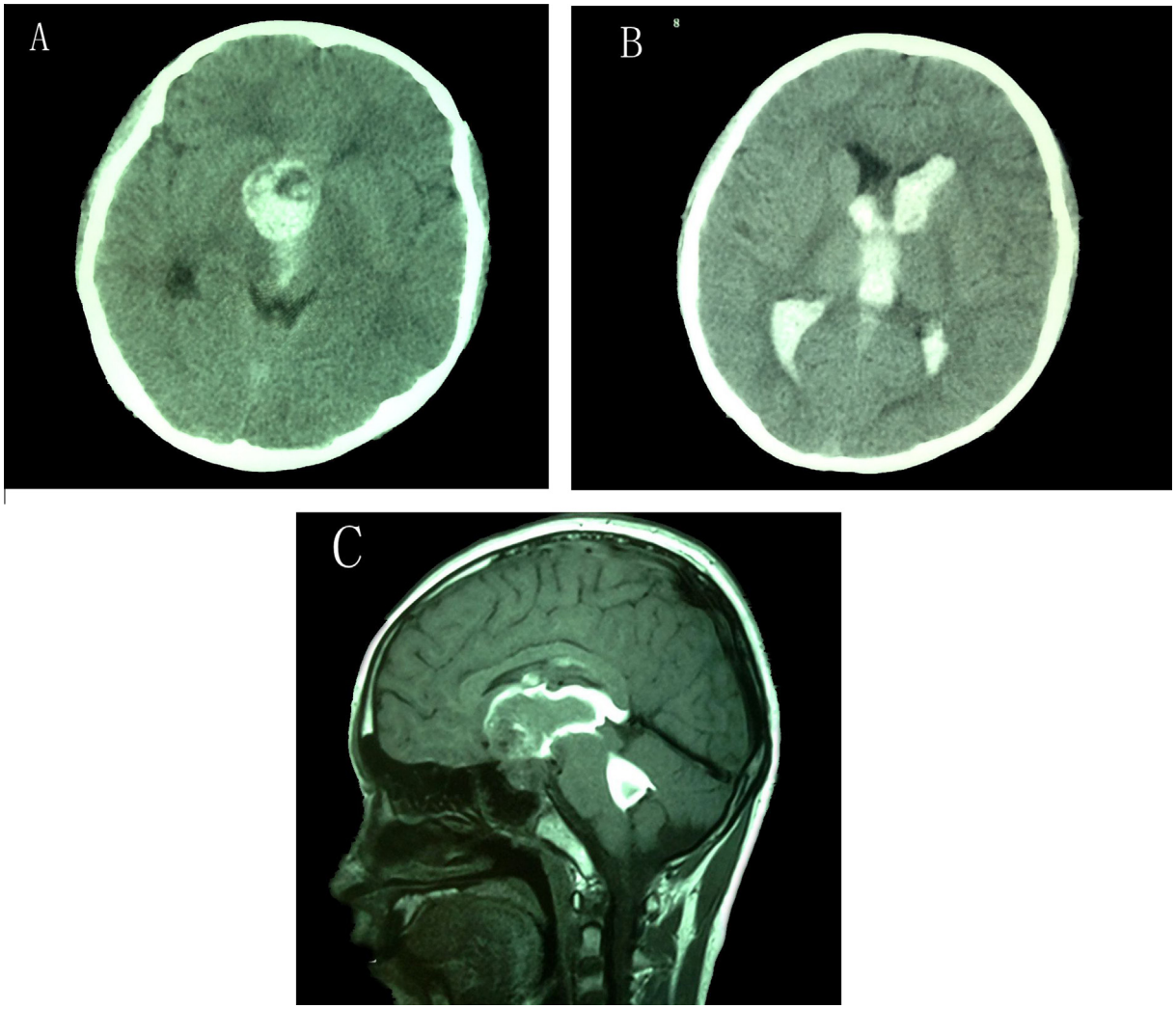

Fig. 1. CT scan of the head showing a mass lesion in the suprasellar region with (A) haemorrhage and (B) intraventricular haemorrhage with marked hydrocephalus. (C) Sagittal T1-weighted MRI revealing a lesion in the suprasellar region with haemorrhage and hydrocephalus.

gery. At the time of this writing, the neurologic evaluation of the patient showed stable results.

3. Discussion

Patients with PMAs rarely present with spontaneous haemorrhage [5]. The mechanism that results in haemorrhages in this type of low-grade astrocytoma is not well understood [6,7]. Gottfried et al. reported the first case of a PMA of the temporal lobe presenting with intratumoural haemorrhage. They proposed the possibility of blood obscuring the histologic changes that contribute to the haemorrhage [6]. Hamada et al. described a patient with a PMA in the suprasellar region presenting with intratumoural haemorrhage; the occurrence of intratumoural and intraventricular haemorrhage during adjuvant chemotherapy 4 months after the operation was fatal. They presumed that the second haemorrhage was possibly associated with the neurotoxicities of the chemotherapeutic agents and the partial resection of the tumour [8].

In the present case, the patient initially presented with massive intratumoural and intraventricular haemorrhage. Such a case is very rarely described in the neurosurgical literature. The mechanisms underlying the haemorrhage associated with this type of tumour remain unclear. It was possible that intratumoural bleeding broke into the third ventricle adjacent to the tumour and spread to the whole ventricular system.

PMAs may occur anywhere along the neuraxis; however, they exhibit predilection for the hypothalamic/chiasmatic region and tend to affect very young children. The management of PMAs in the hypothalamic/chiasmatic region among children is challenging and remains controversial [9]. Any intervention should be carefully performed considering the potential risk of treatment-related morbidity. Goodden et al. suggest that surgical debulking has a clear role in diagnosis, tumour control and relief of mass effect. Partial resection can be primarily used as a safe and effective treatment for PMA [10]. However, Massimi et al. regard surgery not as the initial treatment but as an intervention of last resort with a potentially high risk [11]. Tsugu et al. point that chemotherapy should be used as the main treatment for PMA in the hypothalamic/chiasmatic region. The particular advantages of chemotherapy are that it can be given to younger children with relatively low rates of associated complications and low risks of long-term effects [12]. Radiotherapy is considered to be effective in controlling the growth of PMA; however, the late side effects of radiotherapy have led to its decreased use. Avoidance of radiotherapy is recommended for patients less than 5 years old [10,11,13]. Stereotactic radiosurgery (SRS) allows for the delivery of high-dose radiation