Chinese Journal of Cancer

Case Research

Malignant meningioma of the cerebellopontine angle in a 2-year-old girl: a case report and literature review

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Abstract

Meningioma is a common intracranial tumor in adults. Pediatric cases account for approximately 1.5% of all intracranial meningiomas, and very few cases show malignant histological features. Primary pediatric malignant meningioma in the cerebellopontine angle is extremely uncommon. Herein, we report a 2-year-old girl with malignant meningioma in the cerebellopontine angle. The clinical features, diagnosis, and treatment protocol are discussed.

Key words Malignant meningioma, the cerebellopontine angle, pediatric, craniotomy

Meningiomas account for less than 3% of all primary intracranial tumors in children[1,2], are predominate in teenagers, and are scarce in infants[3,4]. Pediatric cases account for approximately 1.5% of all intracranial meningiomas, and very few cases show aggressive histological features[5-8]. Primary pediatric malignant meningioma in the cerebellopontine angle is extremely uncommon, and the clinical course and prognosis of this tumor in children is uncertain[4]. Herein, we report a 2-year-old girl with malignant meningioma in the cerebellopontine angle.

Case Presentation

A 2-year-old girl with no significant medical history presented at our department in July 2006, describing a 4-month clinical history of droop of the right corner of the mouth and an inability to close the right eye normally and a 2-month history of aggravated headache. A physical examination at admission was abnormal: the right side of her face presented a distorted commissure and hypophasis. Computed tomography (CT) scan of the brain revealed an inordinate globate and defined slightly high-density lesion in the right cerebellopontine angle (Figure 1A). Magnetic resonance imaging (MRI) showed that the lesion had a slightly short T1 signal (Figure 1B) and a long T2 signal (Figure 1C). Contrast-enhanced MRI revealed that the lesion had a conspicuous heterogeneous enhancement, with patchy unenrichment in the lesion. The dural tail sign was visible and measured approximately 38.7 mm × 31.2 mm × 34.7 mm (Figure 1D). A meningioma was diagnosed before the histopathologic examination.

Resection with a right suboccipital retrosigmoid approach was performed. During the operation, the lesion was observed to arise from the meninges of the right cerebellopontine angle and have obscure boundaries. The lesion was grayish-red and tenacious, with copious blood supply. The tumor was completely removed and subjected to pathologic examination immediately.

The tumor tissue was processed and stained with hematoxylin & eosin (HE). The histopathologic examination demonstrated that the tumor was composed of cells with nuclear pleomorphism, prominent nucleoli, a high mitotic index, and a high nucleus/cytoplasm ratio. The mitoses occurred at a rate of approximately 15–20/10 hpf (highest possible frequency) (Figure 2A). Immunohistochemically, the tumor cells were positive for vimentin (Figure 2B), epithelial membrane antigen (EMA) (Figure 2C), and S-100 (Figure 2D) and negative for Syn (Figure 2E) and glial fibrillary acidic protein (GFAP), leading to a final diagnosis of malignant meningioma (WHO grade III).

The patient had no complications after the operation. After surgery, the patient was sent to the neurosurgery intensive care unit (NICU) for vital sign surveillance and neurologic observation. Head CT was performed and showed no residual lesion. Routine follow-up was performed and was scheduled at 1, 6, and 12 months and annually thereafter. The clinical and radiological findings were normal during a 6-year follow-up. No radiotherapy or chemotherapy was administered after surgery.

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doi: 10.5732/cjc.012.10211

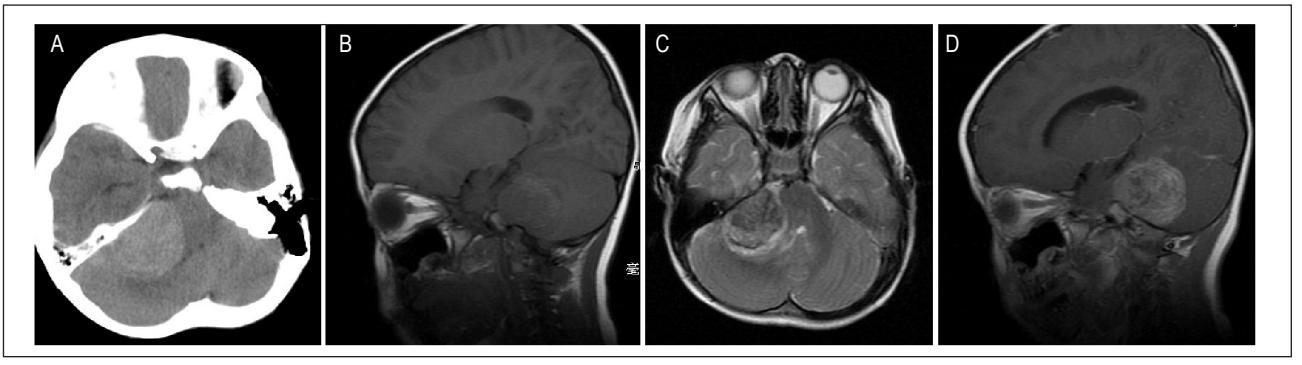

Figure 1. **Computed tomography (CT) and magnetic resonance imaging (MRI) of malignant meningioma in the cerebellopontine angle in a 2-year-old girl.** A, CT scan of the brain reveals an inordinate globate and defined, slightly high-density lesion in the right cerebellopontine angle. B, T1-weighted MRI shows slightly short signals in the lesion. C, T2-weighted MRI shows long signals in the lesion. D, contrast-enhanced MRI reveals conspicuous heterogeneous enhancement, with patchy un-enrichment. The dural tail sign was visible and measured approximately 38.7 mm × 31.2 mm × 34.7 mm.

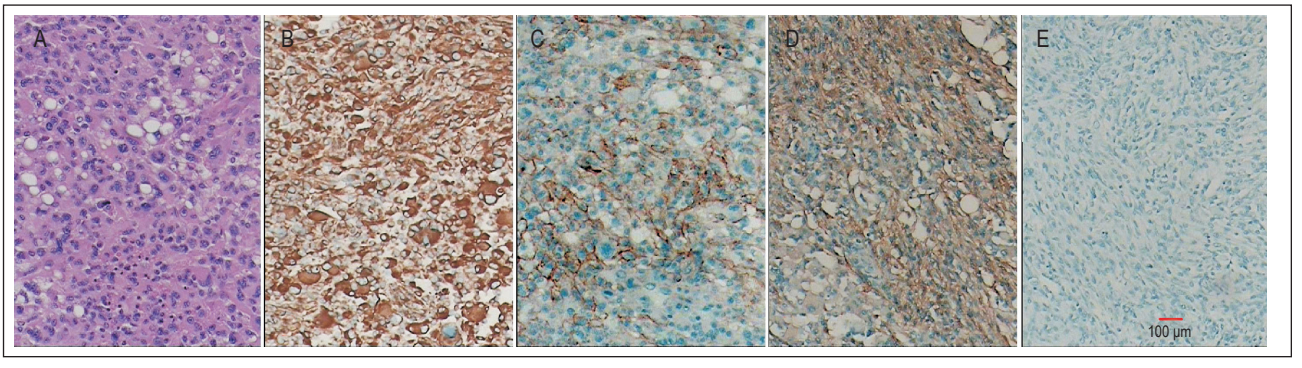

Figure 2. **The histopathologic examination of the malignant meningioma in the presented case.** A, the histopathologic examination shows that the tumor is composed of cells with nuclear pleomorphism, prominent nucleoli, a high mitotic index, and a high nucleus/cytoplasm ratio. Approximately 15–20 mitoses/10 hpf (highest possible frequeney) are visible. Immunohistochemically, most tumor cells are positive for vimentin (B), epithelial membrane antigen (EMA) (C), and S-100 (D) and negative for Syn (E).

Discussion

Meningioma is the second most common tumor of the cerebellopontine angle, accounting for 10% to 15% of tumors in this location, and malignant meningioma is rare[9]. Meningiomas in the first two decades of life have a predilection for occurring in uncommon sites, including intraventricular and infratentorial sites, and display a variety of histological patterns[6,7,10]. The pathogenesis of primary pediatric malignant meningioma is still ambiguous. The ratio of male to female cases is 1:2 for all intracranial meningiomas, but there is a clear male predominance in the pediatric meningioma group, with a boy to girl ratio of approximately 2–3:1[1,3,11]. This may suggest that the pathogenesis differs between the two age groups. The possible etiologic factors in children may include neurofibromatosis, ionizing radiation and congenital causes[10]. In our case, the etiologic factor was unconfirmed, but congenital factors may have played the primary role.

The symptoms of pediatric malignant meningioma are nonspecific and vary according to its location and the extent of involvement. Headache, epilepsy, cranial nerve dysfunction, and circumscribed functional neurologic disorder are typical features of pediatric malignant meningioma. In our case, the lesion was in the cerebellopontine angle, and the facial nerve was affected, causing distortion of the commissure. Headache, a sign of increased intracranial pressure, is a common symptom. Chronic increased intracranial pressure can also lead to an increased head circumference and late fontanel closure in younger children[12].

The radiologic imaging features of our case were consistent with previous reports. The radiologic features of pediatric malignant meningiomas include an enormous size, irregular circumscription, heterogeneous enhancement, necrosis, and hemorrhage[5]. In our case, MRI showed that the lesion had a slightly short T1 signal and a long T2 signal in addition to a conspicuous heterogeneous enhancement with contrast administration on T1-weighted images, and measured approximately 38.7 mm × 31.2 mm × 34.7 mm. The CT images of this girl showed a hyperdense mass in the right cerebellopontine angle, similar to previous reports. The extremely large size may be related to the strong compensation ability of children[11].

The WHO classification divides meningiomas into three grades:

I, benign; II, atypical; and III, anaplastic/malignant[1]. The histology of meningioma differs between children and adults. In children, 7% to 16% of meningiomas are malignant, which is significantly higher than in adults[11,13]. Histopathologic and immunohistochemical examinations, even electron microscopy, are unique methods for making a definite diagnosis of malignant meningioma. The histopathologic features of malignant meningiomas include hypercellularity, nuclear pleomorphism, prominent nucleoli, a high mitotic index, a high nucleus/cytoplasm ratio, loss of architecture, focal necrosis, and brain infiltration or metastasis[11]. In this case, the tumor showed aggressive histological features coincident with the WHO criteria.

The mainstay of therapy for malignant meningioma is complete surgical resection of the lesion. However, resection does not guarantee a good outcome. Postoperative focal or systematic radiotherapy remains controversial in younger patients for several reasons, including uncertainty in prognosis, little supporting evidence, and the potential adverse long-term effects on brain function[7,8,11]. Additionally, ionizing radiation is also an important pathogenic factor of these lesions. However, if a tumor is malignant or recurrent and cannot be removed completely, radiotherapy should be considered. The histological type and the extent of resection are the most important factors influencing recurrence. Complete lesion resection was performed for our case, and radiotherapy was therefore not performed after surgery. The recurrence and mortality of children with malignant meningioma are higher than those of adults[4]. However, only a few malignant meningiomas have been reported in younger children, confirming the poor prognosis.

Received: 2012-08-22; revised: 2012-12-05;
accepted: 2012-12-06.

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