

Images

Primary intracranial meningeal melanoma mimicking chronic meningitis: A case report



Po-Cheng Hung^{a,*}, Yu-Ching Chang^a, Meng-Ying Hsieh^a,
Chieh-Tsai Wu^b

^a Division of Pediatric Neurology, Chang Gung Children's Hospital, Chang Gung Memorial Hospital, Chang Gung University, College of Medicine, Taoyuan, Taiwan

^b Division of Pediatric Neurosurgery, Chang Gung Children's Hospital, Chang Gung Memorial Hospital, Chang Gung University, College of Medicine, Taoyuan, Taiwan

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A 6-year -8-month-old girl was presented with headache, vomiting, and unstable gait. Neurologic examination revealed dysmetria, positive tandem gait, and negative Romberg test. Computed tomography of the brain showed mild ventriculomegaly (Fig. 1A). Cerebrospinal fluid (CSF) analysis revealed negative findings. However, there was a suspicion of cerebellitis. She was discharged with clinical improvement. However, the headache and vomiting recurred after 6 months. Magnetic resonance imaging (MRI) of the brain revealed marked dilatation of all ventricles with prominent leptomeningeal contrast enhancement. The CSF analysis showed elevated protein levels without pleocytosis. Based on these findings, the patient was diagnosed with meningitis complicated by hydrocephalus. She required insertion of a ventriculoperitoneal shunt. Brain biopsy from the third ventricular floor revealed gliosis. The clinical symptoms of the patient improved after the insertion of a ventriculoperitoneal shunt. Unfortunately, during the subsequent 10 months, she still had intermittent headache and

vomiting. Follow-up brain MRI (Fig. 1B) revealed persistent hydrocephalus with diffuse leptomeningeal enhancement despite several times of ventriculoperitoneal shunt revision. Serial CSF analyses demonstrated elevated protein levels without pleocytosis (Table 1). A tentative diagnosis of chronic meningitis was made after negative findings of serology; cultures of viral, bacterial, tuberculous, fungal, or parasitic infections; and CSF cytology for malignancy. At the age of 8 years, she suffered from headache, vomiting, unstable gait, and neck and back pain. Neurologic examination revealed lethargy, dysmetria, positive tandem gait, and negative Romberg test. Brain MRI showed the progression of hydrocephalus and irregular pial–arachnoid enhancement over the basal cistern, the peripontomesencephalic cistern, and the cerebellum (Fig. 1C). Spinal MRI showed extensive leptomeningeal lesions involving the entire spinal column (Fig. 1D). Brain biopsy (Supplementary Figure) from the cerebellar tonsil revealed epithelioid tumor cells containing melanin pigments that showed positive immunohistochemical staining for S-100, melanoma-associated antigen, and anti-melanoma antibody. Further appropriate evaluations showed the absence of cutaneous, mucosal, ocular, or visceral melanoma, based on which the patient was diagnosed with primary intracranial meningeal melanoma. She underwent whole brain

* Corresponding author. Division of Pediatric Neurology, Chang Gung Children's Hospital; Chang Gung University, College of Medicine, 5, Fu-Hsing St., Kwei-Shan, Taoyuan, 333 Taiwan. Tel.: +886 3 3281200 ext 8200; fax: +886 3 3288957.
E-mail address: h2918@cgmh.org.tw (P.-C. Hung).

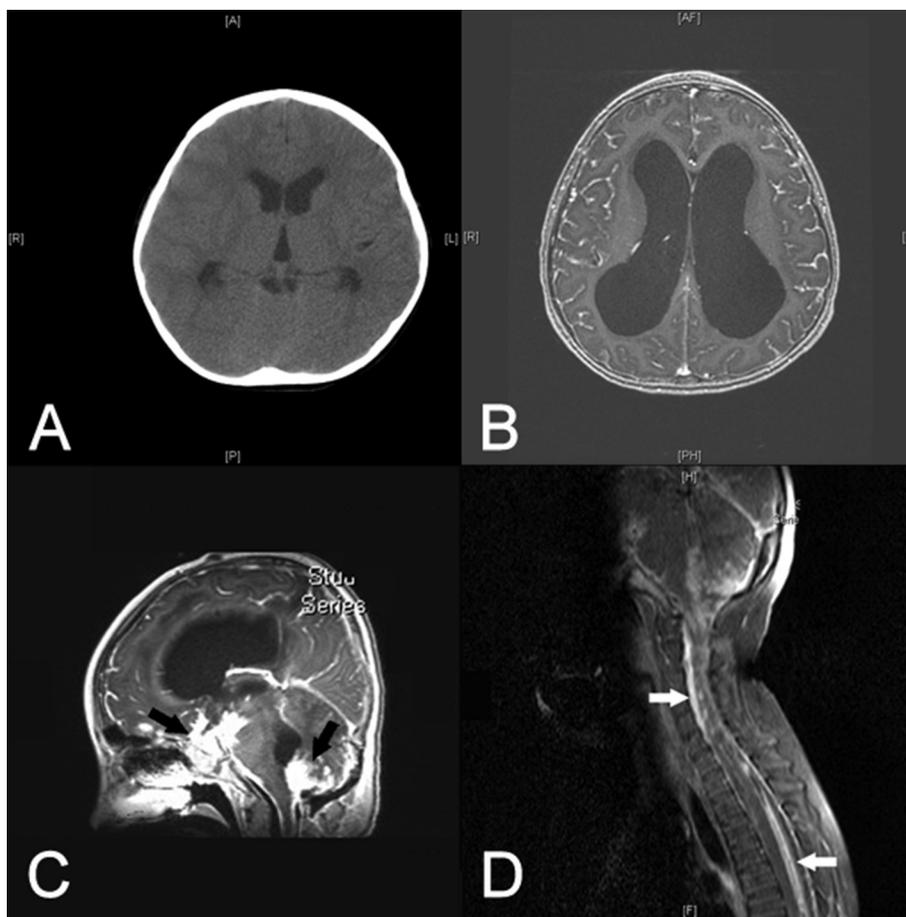


Figure 1 (A) Noncontrast brain computed tomography demonstrated mild ventriculomegaly. (B) Postcontrast T1-weighted axial brain MRI revealed dilated ventricles and diffuse leptomeningeal enhancement. (C) Postcontrast T1-weighted sagittal brain MRI showed dilated ventricles and diffuse pial–arachnoid enhancement in the basal cistern, the peripontomesencephalic cistern, and the cerebellum (arrow). (D) Postcontrast T1-weighted sagittal spinal MRI revealed diffuse intradural extramedullary lesions (arrow).

Table 1 Serial studies of cerebrospinal fluid.

Months after cerebellitis	0 month	6 months	11 months	15 months	16 months
RBC (cells/uL)	1	223	0	0	2
WBC (cells/uL)	3	3	5	2	0
Neutrophil (%)		10	0	10	
Lymphocyte (%)		60	40	10	
Monocyte (%)		30	5	80	
Atypical lymphocyte (%)			55		
Protein (mg/dL)	20.1	48	211.5	256.3	121.7
Sugar (mg/dL)	49	43	38	54	81
Lactate (mg/dL)		39.9	44.7	56.8	40.2

RBC: red blood cell; WBC: white blood cell.

radiotherapy with a total dose of 3000 cGy. However, the response to this treatment was poor, and she died 1.5 months after the diagnosis.

The incidence of primary intracranial meningeal melanoma is rare in children, accounting for <1% of all pediatric malignancies.¹ It has been classified pathologically into two types, one diffusely invades the pia mater and the subarachnoid space, and the other causes nodular

tumors.¹ Elderly children and adults tend to present with the symptoms of space-occupying lesions. In either form, primary intracranial meningeal melanoma is generally a highly malignant tumor. The diagnosis can be confirmed based on the findings of histopathological and immunohistochemical examination.² The management of primary intracranial meningeal melanoma has not yet been standardized, and surgery remains the preferred treatment.³

This report emphasizes that chronic meningitis should be considered as another potential manifestation of primary intracranial meningeal melanoma.

Conflict of interest

The authors declare no potential conflicts of interest with respect to research, authorship, and/or publication of this article.

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Appendix A. Supplementary data

Supplementary data to this article can be found online at <https://doi.org/10.1016/j.pedneo.2018.10.003>.