Journal of Clinical Neuroscience 18 (2011) 1303–1307

Contents lists available at ScienceDirect

Journal of Clinical Neuroscience

journal homepage: www.elsevier.com/locate/jocn

Clinical Study

Clinical features and microsurgical treatment of pediatric patients with cerebral cavernous malformation

Jianping Song †, Nidan Qiao †, Liqian Xie, Tianming Qiu, Peiliang Li, Jingsong Wu, Wei Zhu, Liang Chen, Kang Min He, Nan Zhang, Ying Mao, Yao Zhao *

Department of Neurosurgery, HuaShan Hospital, Shanghai Medical College, FuDan University, 12 Wulumuqi Zhong Road, Shanghai, 200040, China

ARTICLE INFO

Article history:
Received 18 September 2010
Accepted 28 January 2011

Keywords:
Brain
Cavernous malformation
Child
Microsurgery

ABSTRACT

The aim of the present study was to describe the clinical features and to evaluate the surgical treatment outcomes of pediatric patients with cerebral cavernous malformations (CCM). We investigated 85 children (53 boys and 32 girls), aged from 6 months to 17.9 years with CCM. Seizures and symptomatic hemorrhages, which were the most frequent symptoms, occurred in 81 patients. Nine patients had a positive family history of CCM. Eighty patients underwent microsurgical treatment after strict operative indications were met. Neuronavigation, combined with intraoperative ultrasonography or functional MRI, was used for precise localization of the lesions. The principles of minimally invasive techniques were followed during surgery. A total of 89 lesions were removed in 80 patients, and there were no deaths. During their hospital stay, only nine patients suffered from postoperative seizures, which were controlled with medication. Postoperative neurological deficits improved in 27 patients, were unchanged in nine, and worsened in two. With the help of advanced neuroimaging, a satisfactory surgical outcome was achieved for 10 lesions located in eloquent brain areas and four lesions in the brain stem. A follow-up study of 66 patients showed that all of these patients remained seizure-free, and nine patients with postoperative neurological deficits gradually recovered. Microsurgical treatment should be performed early for pediatric patients with CCM. Accurate localization of the lesions and the use of minimally invasive techniques and functional MRI monitoring were the key features of the surgical procedures.

© 2011 Elsevier Ltd. All rights reserved.

1. Introduction

The apoplectic hemorrhages of cerebral cavernous malformations (CCM) are being increasingly recognized as a cause of seizures and focal neurological deficits in young patients.[1,2] The incidence of pediatric CCM is estimated to be 0.37% to 0.53%, which represents 25% of all CCM.[3] Because children possess unique physiological features during development, the surgical management of pediatric CCM patients differs from that of adult patients with CCM. In this retrospective review of a consecutive series of 85 children with CCM treated in the Department of Neurosurgery of HuaShan Hospital we focus on the description of the clinical features of pediatric patients with CCM and the key aspects of the microsurgical treatment.

2. Patients and methods

2.1. Patients

Between December 1994 and April 2009, 85 pediatric (under the age of 18) patients with CCM were admitted to the Department of Neurosurgery of HuaShan Hospital, which is affiliated with FuDan University. Of this group, 53 patients were male and 32 were female, and their ages ranged from 6 months to 17.9 years (average, 13.1 years). The disease duration ranged from 2.7 hours to 8 years (average, 13.7 months). Nine patients with multiple lesions were found to have a positive family history based on brain MRI scanning or questionnaire and interviews.

2.2. Size and location of the lesions

The size of the CCM ranged from 0.5 cm to 6.0 cm (average, 2.7 cm), and a total of 99 lesions were detected before surgery. The distribution of the lesion locations is summarized in Table 1. One patient had a single lesion that was coupled with a tentorial dural arteriovenous fistula, and one patient with two lesions also

* Corresponding author. Tel.: +86 21 52887200; fax: +86 21 62492884.
E-mail address: zhaoyaohs@hotmail.com (Y. Zhao).
† These authors contributed equally to this work.

0967-5868/$ - see front matter © 2011 Elsevier Ltd. All rights reserved.
doi:10.1016/j.jocn.2011.01.033

Table 1
Number and location of cerebral cavernous malformation lesions in pediatric patients

	Single lesion		Multiple lesions
No. patients	76		9
No. lesions	76		23
Location per patient		Location per patient	
Cerebral hemisphere	45	Cerebral hemisphere	5
Brain stem	13	Cerebral and cerebellar hemispheres	3
Basal ganglia	8	Cerebral hemisphere and cervical cord	1
Cerebellar hemisphere	7		
Ventricle wall	3		

had a cutaneous hemangioma that was located on the dorsum of the hand.

2.3. Clinical presentation

The clinical presentation and the location of the lesions are summarized in Table 2. Seizure was the initial symptom in 43 patients (grand mal in 23 and partial seizures in 20), of whom 35 had seizures that were refractory to medication, six had medically controlled seizures and two were not treated with medication. Of these 43 patients, 41 had a single lesion, whereas the other two had multiple lesions. The patients suffered seizures over periods that ranged from 2 days to 8 years (average, 12.2 months). In 38 patients with intracranial hemorrhage (ICH), the initial presentation was with neurologic deficits caused by lesional hemorrhage: the symptoms are detailed in Table 2. Four lesions were discovered incidentally during routine head examination by MRI.

2.4. Morphological and functional characterization

2.4.1. CT scan

The CCM were characterized by isodense or small hyperdense spherical nodules with sharp, regular, non-enhancing margins.

2.4.2. MRI

T1-weighted and T2-weighted spin-echo sequences and fluid-attenuated inversion recovery (FLAIR) sequences were performed for all patients. The results showed the typical characteristic appearance of CCM with a mixed signal reticular core surrounded by a hypointense hemosiderin rim.

2.4.3. Electroencephalogram

A total of 32 patients with seizure symptoms received an electroencephalogram examination, and marked spikes that correlated with the CCM location were noted.

2.4.4. Family investigation and detection of genetic mutations

Family investigations, with the approval of the Human Subjects Review Committee of HuaShan hospital, were conducted for four patients with a family history of CCM. Each family member underwent brain MRI and a detailed clinical assessment with an emphasis on neurological, dermatological, and ophthalmological examinations. The diagnosis of CCM was based on characteristic radiographic findings. Then, genomic DNA was extracted from whole blood and inheritable mutations of the *CCM1* gene were examined by direct sequencing.[4,5]

2.5. Treatment methods

2.5.1. Patient selection

A total of 80 patients (including 42 with seizures and 38 with intracranial hemorrhage [ICH]) underwent microsurgical treatment, whereas the other five patients underwent conservative treatment; of these five patients, four had asymptomatic lesions, and one presented with epileptic CCM combined with systemic disease. No patient underwent radiotherapy. The operative indications were: (i) seizure onset; (ii) focal neurological deficits caused by hemorrhage; and (iii) in the case of multiple lesions, surgical treatment was aimed at symptomatic or bleeding lesions.

2.5.2. Microsurgical procedure

2.5.2.1. Localization. The lesions were localized according to skull landmarks and MRI in all surgical patients. A neuronavigation system (StealthStation Treon) (Medtronic, Minneapolis, MN, USA) was used in 35 patients to locate small or deep-seated lesions precisely. Intraoperative ultrasonography (US) was used in six patients to provide convenient, real-time localization of the lesion. Neuronavigation combined with US was used in four patients. A "microcatheter guided technique" was used during neuronavigation-assisted surgery for deep-seated or multiple lesions to correct for the brain shift due to cerebrospinal fluid (CSF) leakage caused by the opening of the dura.[6] Functional MRI (fMRI), including blood oxygen level-dependent imaging and diffusion tensor imaging (DTI), was specifically adopted to label the motor or speech area in the 10 patients with lesions in eloquent brain areas.

Table 2
Clinical presentation and location of pediatric cerebral cavernous malformations (CCM)

	No. patients	Presentations	No. patients	Frontal lobe	Temporal lobe	Parietal lobe	Occipital lobe	Cerebellum	Brain stem	Basal ganglia	Ventricle wall	Multiple lesions
Seizure	43	Grand mal	23	11 (9)	6 (5)	3 (3)				1		2 (2)
		Partial seizures	20	8 (6)	6 (6)	4 (3)	2 (1)					
ICH	38	Hemiparesis	11	1					6	4		
		Facial paresis	7					2	1	2		2
		Diplopia	6				1		5			
		Ataxia	4					3				1
		Acute intracranial hypertension	2					1			1	
		Behavioral abnormality	2	1								1
		Aphasia	1	1								
		Non-specific symptoms	5					1	1	1	1	1
Incidental	4				1						1	2

The numbers in parentheses represent the number of patients with seizures refractory to medication.
ICH = Intracranial hemorrhage.

Download English Version:

https://daneshyari.com/en/article/3060743

Download Persian Version:

https://daneshyari.com/article/3060743

Daneshyari.com