Clinical Neurology and Neurosurgery 115 (2013) 1407–1414

Contents lists available at SciVerse ScienceDirect

Clinical Neurology and Neurosurgery

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

Diagnosis and surgical treatment of sporadic meningioangiomatosis

Rui Feng[a,1], Jie Hu[a,1], Xiaoming Che[a,*], Li Pan[a], Zhiqiu Wang[a], Mingguang Zhang[a], Fengping Huang[a], Bin Xu[a], Renling Mao[a], An Sun[a], Weimin Bao[a], Ping Zhong[a], Yin Wang[b]

[a] Department of Neurosurgery, Huashan Hospital, Fudan University, Shanghai 200040, China
[b] Department of Neuropathology, Huashan Hospital, Fudan University, Shanghai 200040, China

ARTICLE INFO

Article history:
Received 19 April 2012
Received in revised form
24 December 2012
Accepted 20 January 2013
Available online 26 February 2013

Keywords:
Sporadic meningioangiomatosis (MA)
Intractable seizure
Neurofibromatosis type 2 (NF2)
Radiological features
Surgical treatment

ABSTRACT

Objective: To discuss the clinical characteristics, radiological features, surgical treatment and prognosis of sporadic meningioangiomatosis (MA).
Methods: We retrospectively analyzed the medical records of ten histopathologically confirmed MA patients who were treated in the Department of Neurosurgery of Huashan hospital from 2002 to 2011. All of the patients presented with symptomatic seizure attacks before craniotomy surgeries. Magnetic resonance imaging (MRI) and/or computed tomography (CT) were the main radiological examination for preoperative diagnosis of all cases.
Results: All patients underwent craniotomy surgeries with gross total resections (GTRs) of the MA lesions. Postoperative follow-ups range from 8 to 108 months, in average 42.7 months, median 40.5 months. No radiological recurrence can be found in any case. Eight patients (80.0%) have achieved total symptomatic remission after surgeries (one of them underwent delayed remission), while two (20.0%) are still suffering from seizure attacks infrequently under several antiepileptic drugs (AEDs).
Conclusion: Although MA cases are quite rare and usually misdiagnosed presurgically, a correct preoperative diagnosis, at least a differential diagnosis, can be rationally achieved via a triad of patients' ages, symptomatic seizure attacks and radiological features (both CT and MR). MA is curable and the prognosis is excellent since most patients became free of seizure and recurrence after surgical treatments.

1. Introduction

Mengioangiomatosis (MA) is a rare hamartomatous lesion which usually involves the leptomeninges and underlying cerebral cortex [1]. MA is not neoplastic and barely grows, so it can be surgically cured [1–3]. It has been nearly 100 years since the first MA case was identified, but little is known about MA because it rarely happens. As a result, MA is usually misdiagnosed, and sometimes mistreated by ineffective methods, e.g. radiosurgical treatment. Two types of MA are broadly accepted [4,5]: neurofibromatosis type 2 (NF2) associated MA and sporadic MA. NF2 associated MA is generally found in autopsy and the patients usually experience no noticeable symptoms. This article specifically focuses on sporadic MA. Ten cases of MA patients surgically treated in the Department of Neurosurgery of Huashan hospital are reviewed. They happen to be the largest MA samples in one single neurosurgical center. The clinical characteristics, radiological features, treatment methods and surgical outcomes are analyzed and demonstrated as follows.

2. Materials and methods

2.1. Clinical materials

All ten MA patients (Table 1) were surgically treated in our department from 2002 to 2011. None of the patients presented any signs of NF2 according to current clinical diagnostic criteria [6,7]. Six patients were males and four were females. Average age at surgery was 19.5 years old (ranging from 8 to 39 years old); average age of seizure onset was 16.7 years old (ranging from 1 to 24 years old), one with history of febrile convulsion attacks. All ten patients were either misdiagnosed as low-grade gliomas (six cases), vascular malformations (one case), cystic encephalomalacia (one case) or calcified lesions (two cases), and they usually suffered from repeated seizure attacks. Four patients presented with generalized tonic-clonic seizures (GTCS), two with complex partial seizures (CPS) and four with simple partial seizures followed by secondary GTCS (SPS–GTCS) attacks. Except for one 8-year-old patient whose

* Corresponding author at: Department of Neurosurgery, Huashan Hospital, Fudan University, 12 Wulumuqi Zhong Road, Shanghai 200040, China.
Tel.: +86 21 5288 7214; fax: +86 21 5288 7214.
E-mail addresses: Xiaoming_che@yahoo.com.cn, neurosurgery2000@yahoo.com.cn (X. Che).
[1] The first two authors contributed equally to this work.

0303-8467/$ – see front matter © 2013 Elsevier B.V. All rights reserved.
http://dx.doi.org/10.1016/j.clineuro.2013.01.021

Table 1
Ten cases of sporadic Meningioangiomatosis treated in Huashan hospital.

Patient no.	Age (years)/sex	Seizure duration (months)	Postop follow-up (months)	Lesion location	Seizure type	Radiology	Outcome
1	18/M	2	10	R ft	SPS–GTCS	CT: calcification; T1WI: iso with hypo; T2WI: hypo with iso/hyper; FLAIR: hypo with iso/hyper; C+: slight spotty enhancement; MRA: normal.	Engel class I
2	18/M	12	8	R ft	GTCS	T1WI: iso; T2WI: iso with hyper; FLAIR: hypo with iso/hyper; DWI: hypo; C+: little enhancement; MRS: Cho/NAA = max 1.359.	Engel class I
3	13/F	2	49	L pt	SPS–GTCS	CT: calcification; T1WI: iso with hypo; T2WI: hypo with iso/hyper; FLAIR: hypo with iso/hyper; DWI: hypo; C+: little enhancement.	Engel class I
4	39/F	180	40	R tp	CPS	T1WI: iso with hypo; T2WI: iso with hyper/hypo; FLAIR: hyper with iso/hypo; DWI: hypo with hyper; C+: little enhancement.	Engel class III
5	8/M	0.5	20	R ft	GTCS	CT: calcification; T1WI: iso with hypo; T2WI: iso with hyper/hypo; FLAIR: iso with hyper/hypo; C+: heterogeneous enhancement.	Engel class I
6	21/M	24	31	L pt	GTCS	CT: calcification; T1WI: iso; T2WI: iso/hypo with hyper; FLAIR: hypo with iso; DWI: hypo; C+: slight heterogeneous enhancement; DSA: normal; MRS: Cho/NAA = max 0.943.	Engel class I
7	14/M	11	108	L ft	SPS–GTCS	Cystic lesion; T1WI: hypo with iso; T2WI: hyper; FLAIR: iso with hyper, incomplete suppressed cystic fluid; C+: no enhancement; MRA: normal.	Engel class III
8	17/F	60	75	R tp	CPS	CT: calcification; T1WI: hypo; T2WI: hypo with hyper/iso; C+: little enhancement.	Engel class II
9	34/F	48	41	L oc	GTCS	Nodule with cystic change; T1WI: hypo with iso; T2WI: hyper with hypo; FLAIR: hypo, incomplete suppressed cystic fluid; DWI: hypo; C+: obvious enhancement.	Engel class I
10	13/M	4	45	R pt	SPS–GTCS	T1WI: hypo; T2WI: hyper with iso; C+: obvious enhancement; MRS: unstable baseline.	Engel class I

All ten patients received gross total resection (GTR) of the MA lesion. Five patients (Nos. 1, 3, 5, 6, 8) received plain CT scans, which present calcifications of the lesions. R = right, L = left, ft = frontal, pt = parietal, tp = temporal, oc = occipital, GTCS = generalized tonic-clonic seizure, SPS = simple partial seizure, SPS–GTCS = simple partial seizure with secondary generalized tonic-clonic seizure, CPS = complex partial seizure, AEDs = antiepileptic drugs. iso = isointensity; hypo = hypointensity; hyper = hyperintensity; Cho = choline; NAA = N-acetylaspartate; MRA: magnetic resonance angiography; DSA = digital subtraction angiography; C+ = Gd-contrast; radiology: A with B = mainly A mixed with B.

family refused any drug-related treatment (he received surgery 0.5 month after his first seizure attack), all the other nine patients received one or multiple AEDs, which appeared poorly effective for controlling, these intractable seizure attacks.

2.2. Radiological materials

Ten MA lesions were all solitary and located in the cortex of supra-tentorium cerebral hemispheres. There were four on the left side and six on the right; four in frontal lobes, two in temporal lobes, three in parietal lobes and one in occipital lobe. All the patients accepted MR scans with different sequences including T1 weighted imaging (T1WI, all ten patients), T2 weighted imaging (T2WI, all ten patients), fluid attenuated inversion recovery (FLAIR, eight patients), diffusion weighted imaging (DWI, five patients) and Gd-DTPA (Gd) enhancement (all ten patients). Most MA lesions were shown on MRI as a solitary gyriform mass, which usually did not seem to demonstrate a clear border with the normal cortex. On T1WI, all ten cases showed iso/hypointensity to gray matter. On T2WI (Fig. 1), the signals expressed diversity: seven were mainly iso/hypointense with or without hyperintensity, three were mainly hyperintense with or without iso/hypointensity (two of which were cystic). On eight FLAIR images, seven were mainly iso/hypointense with or without hyperintensity, one was mainly hyperintense mixed with iso/hypointensity. Five DWIs showed hypointensity of the lesions except for one mixed with