Clinical Neurology and Neurosurgery 157 (2017) 34–39

Contents lists available at ScienceDirect

Clinical Neurology and Neurosurgery

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

No significant difference between chiari malformation type 1.5 and type I

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ARTICLE INFO

Keywords:
Chiari malformation
Syringomeylia
Tonsillar ectopia
Brainstem herniation
Posterior cranial fossa

ABSTRACT

Objective: Chiari malformation Type 1.5 (CM 1.5) was defined as the association of Chiari malformation Type I (CM I) and brainstem herniation. The objective was to demonstrate the difference of clinical features and surgical outcomes between CM 1.5 and CM I.

Patients and methods: All CM 1.5 and CM I adult patients who underwent posterior fossa decompression with duraplasty at our institution between 2006 and 2010 were retrospectively reviewed. Clinical characteristics, imaging features, and long-term outcomes were compared between CM 1.5 and CM I patients.

Results: A total of 142 adult patients were enrolled, including 27 CM 1.5 and 115 CM I patients. The average follow-up period was 102 months. Age at diagnosis was significantly younger in CM 1.5 group than CM I group ($p = 0.039$). And the degree of tonsillar herniation was significantly more severe in CM 1.5 group than CM I group ($p < 0.001$). There was no significant difference in other clinical and imaging characteristics. Moreover, improvement of symptoms was observed in 21 CM 1.5 patients (77.8%) and 94 CM I patients (81.7%), and no significant difference was detected ($p = 0.637$). There was no significant difference in the resolution of syringomyelia between CM 1.5 (72.7%) and CM I (76.5%) patients, either ($p = 0.710$).

Conclusions: Although CM 1.5 patients presented with brainstem herniation and more severe tonsillar herniation, other clinical and imaging features and surgical outcomes were similar with CM I patients. We think CM 1.5 is just a subtype of CM I, rather than a unique type of Chiari malformations.

1. Introduction

Chiari malformation (CM), also known as Arnold-Chiari malformation, was first described by the Austrian pathologist Hans Chiari in 1891. It is a congenital malformation and characterized by the downward herniation of the cerebellar tonsils into the spinal canal [1–4]. According to different morphological changes, Chiari classified these malformations into four types, thus CM type I (CM I), CM type II (CM II), CM type III (CM III), and CM type IV (CM IV) [5–9]. However, for some patients, the morphological changes of the above four entities do not strictly apply. For example, some patients presented with caudal descent of both cerebellar tonsils and brainstem, without herniation of the fourth ventricle or the vermis. These patients were previously regarded as part of CM I. However, strictly speaking, these patients were neither CM I nor CM II and were recently described as CM type 1.5 (CM 1.5) [5,10,11].

Studies on CM 1.5 were really rare and their clinical manifestations and surgical outcomes were unclear. Moreover, the pathological changes of CM 1.5 and CM I were similar except the brainstem herniation, and no studies have systematically analyzed the differences between CM 1.5 and CM I, especially in adult patients. Therefore, we conducted this retrospective study to demonstrate their differences of clinical and radiologic features and surgical outcome, with a large series enrolled and a long-term follow up performed.

2. Materials and methods

The Institutional Review Board of People's Hospital of Xinjiang Uygur Autonomous Region approved this study. All adult patients with CM I and CM 1.5 who received posterior fossa decompression (PFD) at our institution between January 2006 and December 2010 were enrolled in this study. Patients with a history of lumbar puncture, intracranial lesions, craniosynostosis, meningitis or brain trauma were excluded. Besides, patients who underwent other kinds of surgeries and who lost to follow up were also excluded.

2.1. Clinical presentations

Clinical data were recorded, including sex, age, duration of symptoms and clinical presentations. The various clinical symptoms and

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http://dx.doi.org/10.1016/j.clineuro.2017.03.024
Received 12 February 2017; Received in revised form 26 March 2017; Accepted 28 March 2017
Available online 30 March 2017
0303-8467/

Table 1
Comparison of clinical and imaging characteristics between CM I and CM 1.5 patients.

Characteristics		CMI Group (%)	CM 1.5 Group (%)	p value
Sex (male/female)		32/83	12/15	0.093
Age at diagnosis (years)		40 ± 12	36 ± 10	0.039
Duration of symptoms (months)		62 ± 75	73 ± 71	0.469
Category of chief complaints	Headache	27 (23.5)	6 (22.2)	0.987
	Paresthesia	62 (53.9)	14 (51.9)	
	Motor deficits	19 (16.5)	5 (18.5)	
	Other manifestations	7 (6.1)	2 (7.4)	
Degree of tonsillar herniation (mm)		8 ± 4	14 ± 6	< 0.001
CTD grade	I	64 (55.7)	4 (14.8)	< 0.001
	II	42 (36.5)	7 (26.0)	
	III	9 (7.8)	16 (59.2)	
Hydrocephalus		12 (10.4)	3 (11.1)	1.000
Syringomeylia		102 (88.7)	22 (81.5)	0.489
Klippel-Feil syndrome		1 (0.9)	1 (3.7)	0.345
Occipitalization		7 (6.1)	1 (3.7)	0.984
Basilar ingvagination		12 (10.4)	2 (7.4)	0.908
Scoliosis		9 (7.8)	3 (11.1)	0.867

signs of CM patients were categorized into four groups according to patients' main presentations (Table 1): 1) headache; 2) paresthesia, including numbness, hypoesthesia, pain, and deficits of pain and temperature sensations; 3) motor deficits, such as motor weakness of limbs and muscle atrophy; 4) other manifestations, including hoarseness, dysphagia, dizziness, tinnitus, ataxia, nausea, vomiting and sphincter dysfunctions [12].

Neurological functions were evaluated according to the modified McCormick grading (mMG) system (Table 2) preoperatively, at discharge (usually 1 week after surgery), and at long-term follow-up (80–128 months after surgery, the most recent follow-up)[12,13].

2.2. Radiologic features

All preoperative, post-operative and follow-up brain and cervical magnetic resonance imaging (MRI) and computed tomography (CT) scans were evaluated. The degree of tonsillar ectopia and brainstem herniation was measured on sagittal T1-weighted MRI. The brainstem herniation was exhibited by the inferiorly displaced obex beneath the foramen magnum [5,10,11]. Patients whose tonsillar ectopia was greater than 5 mm without herniation of brainstem, cerebellar vermis and fourth ventricle was defined as CM I. Patients with tonsillar ectopia greater than 5 mm and brainstem herniation without fourth ventricle and vermis herniation was defined as CM 1.5 [5,10,11]. The degree of tonsillar ectopia was classified into three grades according to the cerebellar tonsil descent (CTD) grading scale: 1) Grade I, the tonsil descends over the foramen magnum but does not reach the posterior arch of atlas; 2) Grade II, the tonsil descends to atlas arch level (Fig. 1A, B); and 3) Grade III, the tonsil descends below posterior arch of atlas [12,14–16]. All these measurements and evaluations were performed by two authors independently and consensus was taken.

Moreover, other kinds of abnormalities were also noted in each patient, including hydrocephalus, syringomeylia, basilar ingvagination, occipitalization, scoliosis, and Klippel-Feil syndrome.

2.3. Surgical procedures

For patients with obvious symptoms or syrinx, surgical treatment was recommended to relieve symptoms and to reduce the syrinx. Four patients with hydrocephalus and paraventricular edema on CT scans were treated with ventriculoperitoneal shunt previously. All patients underwent posterior fossa decompression (PFD). A small-bone-window posterior fossa decompression and duraplasy with an autologous graft were performed in each patient, in a left-lateral position. A posterior midline skin incision was made from the external occipital protuberance to the spinous process of the third or fourth cervical vertebra. An autologous graft (2 cm × 2 cm, approximately) was excised from the fascia and reserved. And then the squamous part of the occipital bone (2–3 cm) and C1 lamina (1.5–2.0 cm) was removed. Whether the C2 laminectomy was performed depended on the severity of tonsillar ectopia. The dura mater was opened (1.5–2 cm) without impairing the arachnoid, and then the dura grafting was performed with the previously resected autologous graft. No patient underwent tosillar manipulation or syringomyelia shunting.

2.4. Follow-up

All patients were followed up by outpatient clinical or telephone interviews. Cervical MRIs were performed at one week after surgery, 3–6 months after surgery and then annually. According to patients' main complaints, clinical outcome were classified into two categories: 1) improved: patients experienced marked improvement compared to preoperative symptoms, or complete relief, which significantly improved their quality of life; 2) Not improved: only slight improvement, no change or deterioration of symptoms.

2.5. Statistical analysis

All statistical analyses were performed with SPSS Windows version 22.0 (IBM). The chi-squared test for the R × C contingency tables was used to compare categorical variables between CM I and CM 1.5. Continuous variables were compared using independent *t*-test. Nominal variables (CTD grades, mMG grades) were compared with Wilcoxon rank-sum test. A probability value < 0.05 was considered statistically significant.

3. Results

3.1. Patient population

Between January 2006 and December 2010, a total of 154 adult patients with CMI or CM 1.5 were surgically treated in our institute. However, 12 patients (7.8%) were lost to follow up, thus, 142 patients were enrolled in this study. According to the definition of CM I and CM

Table 2
Modified McCormick grading scale for neurological function.[a]

Grade	Grade Definition
I	Neurologically normal; gait normal; normal professional activity
Ib	Tired after walking several kilometers; running is impossible, or moderate sensorimotor deficit does not significantly affect the involved limb; moderate discomfort in professional activity
II	Presence of sensorimotor deficit affecting function of involved limb; mild to moderate gait difficulty; severe pain or dysesthetic syndrome impairing patient's quality of life; still functions and ambulates independently
III	More severe neurological deficit; requires cane/brace for ambulation or significant bilateral upper extremity impairment; may or may not function independently
IV	Severe deficit; requires wheelchair or cane/brace with bilateral upper extremity impairment; usually not independent