Neuroradiology (2020) 62:39–47
https://doi.org/10.1007/s00234-019-02274-1

DIAGNOSTIC NEURORADIOLOGY

A rupture risk analysis of cerebral cavernous malformation associated with developmental venous anomaly using susceptibility-weighted imaging

Shuo Zhang[1,2] · Li Ma[1,2] · Chunxue Wu[3] · Jun Wu[1,2] · Yong Cao[1,2] · Shuo Wang[1,2] · Jizong Zhao[1,2]

Received: 3 June 2019 / Accepted: 29 July 2019 / Published online: 3 September 2019

Abstract

Purpose To search for the risk factors closely related to cerebral cavernous malformation associated with developmental venous anomaly (CCM-DVA) lesions rupture, laying foundations for the development of reasonable individual treatment plans for patients.

Methods In this retrospective study, we collected CCM-DVA patients who met the inclusion criteria in our outpatient department from 2014 to 2017, MRI scans were performed including susceptibility-weighted imaging (SWI) and contrast-enhanced imaging, characteristics and basic clinical information were collected then statistically analyzed, CCM-DVA lesions were divided into 3 types according to the location and quantitative relationship between CCM and DVA.

Results A total number of 319 adult patients were identified with 41.2 ± 11.9 years on average, though univariate and multivariate regression analysis, ruptured presentations were more common in patients with prior hemorrhage ($p = 0.003$), type III CCM-DVA lesions ($p = 0.001$), lesions volume > 1 cm^3 ($p < 0.001$), infratentorial lesions especially located in midbrain ($p = 0.019$), pontine ($p = 0.007$), medulla ($p = 0.015$). Caplan-Meier curve shows a lower Hemorrhage-free survival rate on patients with type III CCM-DVA lesions (log-rank, $p = 0.0222$), functional area lesions (log-rank, $p < 0.001$), lesions volume > 1 cm^3 (log-rank, $p < 0.001$), infratentorial lesions (log-rank, $p = 0.0002$).

Conclusion The classification based on the relationship between CCM and DVA may be meaningful to predict the risk of lesion rupture and CCM lesions next to DVA distal branches showed a higher risk of rupture.

Keywords Cerebral cavernous malformation · Developmental venous anomaly · Risk factors · Susceptibility-weighted imaging

Introduction

Cerebral cavernous malformation (CCM) is a common cerebral vascular malformation with a prevalence about 0.5% in the population biopsy [1], accounting for about 5–13% of the total number of vascular malformation patients [2]. CCM mostly occurring in supratentorial, recurrent intracranial hemorrhage can be one of its main manifestations. The average hemorrhage rate of unruptured lesions was about 0.4–0.6% per year, and the re-hemorrhage rate of ruptured lesions was significantly increased, about 4.5–22.9% per year [3–6]. Compared with other forms of cerebrovascular malformations such as arteriovenous malformation (AVM) and CCM, developmental venous anomaly (DVA) has a higher prevalence rate of about 3% [1]. It is considered having a benign natural history, most patients have no obvious clinical symptoms and the hemorrhage rate is as low as 0.22–0.68% per year [7, 8], but when combined with other intracranial vascular malformations such as CCMs, it often speeds up its progression.

CCM associated with DVA (CCM-DVA) is thought to be the most common type of intracranial vascular malformation, which first reported by Roberson in 1974 then classified by McCormick in 1984 [1, 9]. About 8–33% of DVA and CCMs exists in combination [10–14], and CCM-DVA is more common in a sporadic patient than familial cases. Previous studies

✉ Jizong Zhao
zhaojz205@163.com

[1] Department of Neurosurgery, Capital Medical University Affiliated Beijing Tiantan Hospital, Beijing, People's Republic of China

[2] China National Clinical Research Center for Neurological Diseases, Beijing, People's Republic of China

[3] Department of Neuroradiology, Capital Medical University Affiliated Beijing Tiantan Hospital, Beijing, China

and analyses on the rupture factors of CCM indicated that female sex [2, 3, 15, 16], young age [17, 18], infratentorial location [17], eloquent location [19], and the presence of a DVA [10, 11, 17, 20–23] are associated with a higher risk of hemorrhage.

Since a considerable part of CCM was associated with DVA, different positional structural relationships may have different effects on lesion rupture at the hemodynamic level. In prior studies, the combined DVA was more discussed as a potential risk factor for CCM rupture, but there is no large-scale study on the patient population in which CCM and DVA are combined, and few studies have used the relationship as an indicator of hemorrhage occur. Taking these into consideration, we decided to discuss the CCM-DVA relationships including the number and structural characters in adult patients based on risk factors discussed in previous studies, primarily expected to predict the risk of hemorrhage under the premise of covering the most comprehensive influencing factors and provide evidence for future clinical decisions.

We retrospectively reviewed 319 adult patients suffering from CCMs associated with DVAs in our institute, by analyzing the neuroimaging features (ruptured/unruptured, multiplicity, location, lesion volume, CCM-DVA relationship, etc.) and basic characteristics of the patient (gender, age, chief complaint, prior hemorrhage history), mainly to identify risk factors for rupture of CCM-DVAs and lay the foundation for subsequent long-term prognosis and natural history studies.

Material and methods

Patient population

We analyzed 319 consecutive adult CCM-DVA patients' outpatient records and imaging data admitted to our institution from 2014 to 2017 retrospectively. All patients received MRI examination at 1.5 t or 3.0 t field strength, including T1- and T2-weighted images + susceptibility-weighted imaging (SWI); additional contrast-enhanced T1-weighted images were performed in 243 patients. Among them, 32 patients received surgical treatment because of recurrent symptomatic hemorrhage, progressive aggravation of neurological dysfunction, or epileptic seizure, and the remaining patients received a conservative observation. Patient's demographic information including age, sex, initial symptoms, prior hemorrhage history, lesions location, and multiplicity were collected. The CCM lesion location was classified as supratentorial and infratentorial. Supratentorial included frontal lobe, temporal lobe, parietal lobe, occipital lobe, insula, corpus callosum, ventricle, basal ganglia, and thalamus. Infratentorial group includes cerebellum, midbrain, pontine, and medulla. CCM lesion volume was calculated from the T2 images of the sagittal, coronal, and axial positions. Al-shahi et al. raised standard published in 2008 were used to evaluate whether the CCM-DVA lesion was ruptured [24]. For patients with multiple CCM lesions confirmed by imaging, only symptomatic lesions/ruptured lesions were selected to include in the study. For asymptomatic and unruptured lesions, the analysis was conducted according to the order of priority from a close relationship with DVA to lesion size.

Diagnostic criteria using MRI

Contrast-enhanced T1-weighted images (T1WI) have long been used as the gold standard for diagnosing CCM-DVAs. With the application of SWI sequence, a non-invasive neuroimaging tool to identify DVA, the higher sensitivity to identify microhemorrhage of CCM lesions is gradually valued [25]. The main advantage of SWI is that it can detect the presence of microbleeds more sensitively and small CCM lesions could be visualized directly; this is especially important for finding and observing DVA-related CCM lesions because the associated recurrent microhemorrhage are considered to be the beginning stage of CCM formation [26, 27]. Therefore, an MRI examination with a contrast-enhanced T1WI + SWI sequence were used in our study to provide the greatest extent diagnosis of existing lesions and a clear relationship between CCM lesions and DVA. (Fig. 1).

Depending on types, DVA can be expressed in a variety of forms on MRI: the most common feature is the stellate or linear abnormal vein that can be discerned on the contrast-enhanced T1WI and/or SWI sequence, which merges into the central vein and flows into the cortical vein or deep.

According to different Zabramski types, CCM also presents different imaging features. The most classical features are sponge-like structures, and the central area on T2-weighted images is the mixed signal, surrounded by low-signal hemosiderin deposition area, namely, "popcorn" sign. Traces of bleeding at different stages can be seen within the contour of the lesion and related hemorrhages can be more sensitively observed in the SWI sequence.

In addition, before the diagnosis of CCM, imaging features and previous medical history characteristics should be used to identify and exclude the diagnosis of similar intracranial space-occupying lesions/hemorrhagic lesions such as AVM and tumor stroke hemorrhage by enhanced scanning when necessary.

Image analysis

All patients' image data were gathered in picture archiving and communication systems (PACS) and evaluated by experienced neuroradiologist. The lesions were divided into ruptured/unruptured group according to the characteristics of the lesions presents on MRI images, and the lesions location, number, and maximum diameter of each axis were determined then calculated by volume formula.

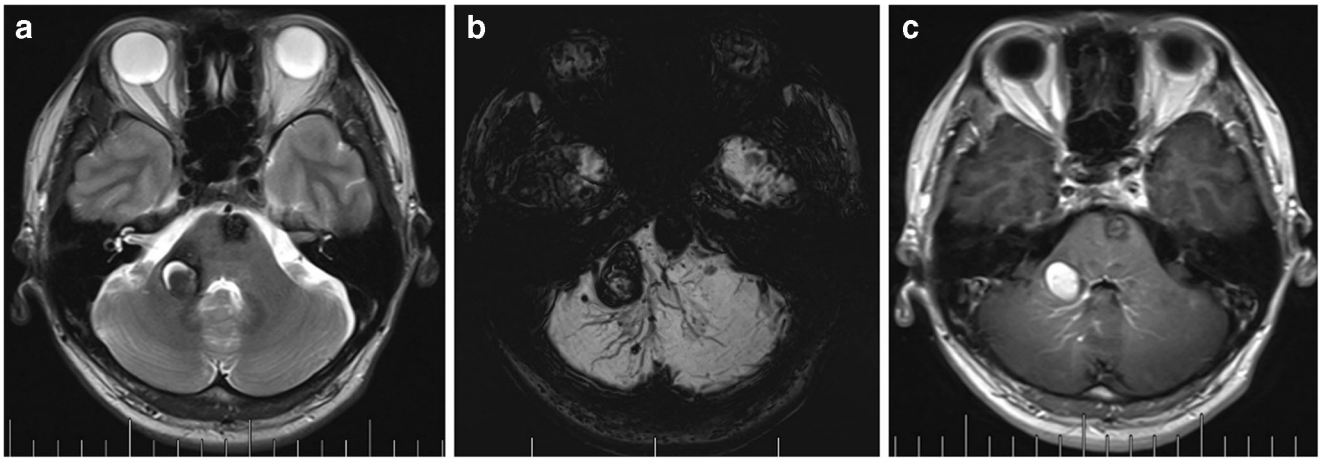

Fig. 1 Patient with multiple CCM-DVA, right cerebellar cavernous malformation has the closest relationship with DVA. DVA can not be recognized on T2WI (**c**), but can be clearly identified in SWI (**b**) and T1-enhanced images (**a**), multiple microhemorrhages at DVA distal branches can be seen by SWI scan and CCMs were highly suspected, which cannot found in enhanced images

Supratentorial deep location refers to CCM located around thalamus, basal ganglia. Location was divided into two groups: supratentorial and infratentorial. Zabramski scale were used for the imaging classification of CCM, and the classification of the hematoma outside the lesion was classified separately and ranked as the third group; however, since the CCM classification based on Zabramski and the exlesional hemorrhage had an obvious direction for the lesions hemorrhage, it was not included in the major discussion affecting the lesions rupture.

CCM-DVAs relationship was valued using contrast-enhanced T1-weighted images and SWI sequences and was classified into three types according to the structural position and quantity relationship between CCM and DVA:

Type I: CCM associated with a single abnormally thickened and distort venous drainage.
Type II: CCM located in the trunk of the DVA with multiple abnormal vein drainage surrounded.
Type III: Typical DVA of the caput medusa-like appearance especially on the SWI/T1-enhanced images, CCM located around the distal radical branch of DVA. (Fig. 2)

Statistical analysis

Statistical analysis was performed using the IBM SPSS version 22.0; univariate analysis using X^2 test (Fisher's exact test when necessary) and independent t tests were used in continuous data, then multivariate analysis confirming the independent risk factors using logistic regression and confirmed by Cox regression. We determined that the CCM-DVA lesion was congenital; Kaplan-Meier curve was drawn to evaluate the relationship between certain risk factors and Hemorrhage-free survival with years. Continuous variables are presented as the mean ± SD, p values less than 0.05 were considered statistically significant.

Result

Demographics of CCMs associated with DVAs

A total number of 319 patients with CCM-DVA were enrolled, with an average age of 41.2 ± 11.9 years (range 18–77 years), of which 176 (55%) were female patients. Eighty-four patients had a prior hemorrhage history (84/319.26%), chief complaint of patients including symptomatic hemorrhage ($n = 148$, 46%), epileptic seizure ($n = 48$, 15%), non-hemorrhage headache/dizziness ($n = 50$, 15%), neurologic deficit ($n = 54$, 17%), and asymptomatic ($n = 19$, 6%). Among them, there were 78 patients with multiple CCMs ($n = 78$, 24%) and 161 patients with infratentorial lesions ($n = 161$, 50%). One hundred ninety-nine patients had lesions adjacent to the eloquent area ($n = 199$, 62%); the lesions diameter measured using T2WI, SWI, and T1-enhanced scan were 15.64 ± 8.56 mm, 18.43 ± 8.27 mm, and 15.53 ± 8.26 mm. The relationship between CCM and DVA is most seen in type III ($n = 230$, 72%), and more detailed demographics are described in Table 1.

We assume that all CCM-DVA lesions are present at birth, there were totally 13,143 patient-years of follow-up of 319 patients (41.2 years average); 140 patients who had 197 ruptured lesions were included, among them, 53 patients had a prior hemorrhage history; the estimated first time hemorrhaging rate was about 0.93% per patient-years.

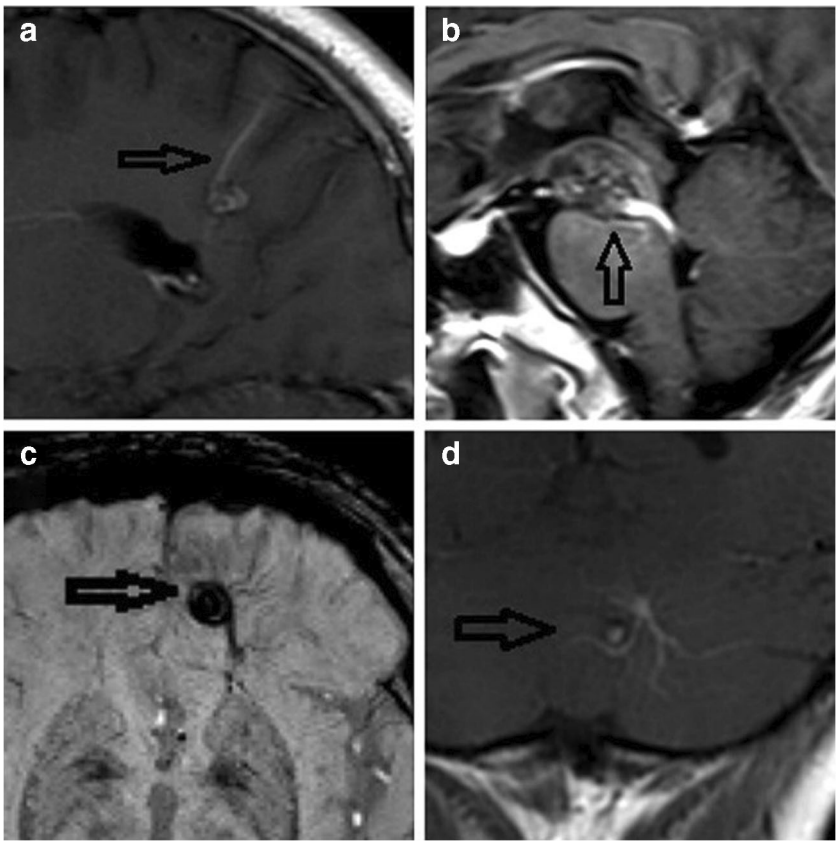

Fig. 2 Different CCM-DVA relationship classification. **a** (Type I) CCM located in occipital lobe with a single drainage vein to the cortex. **b**, **c** (Type II) CCM located in the DVA trunk, surrounded by multiple veins drainage. **d** (Type III): DVA composed of multiple tortuous draining veins that eventually converge into a collecting vein, CCM is located at the turning point of the DVA distal branch, and multiple drainage veins eventually merge into the collecting vein

Risk factors for hemorrhage

Tables 2, 3, and 4 list the following possible hemorrhage-related factors: gender, age, multiple lesions, infratentorial lesions, eloquent lesions, and the diameter of the lesion as well as the type of CCM-DVA. Through univariate analysis and then multivariable analysis, patients with prior hemorrhage history shows a significant higher rupture risk than those ruptured lesions first discovered ($p = 0.003$ [OR] 2.5, [HR] 2.3), which presents more significant in infratentorial lesions ($p = 0.002$, [OR] 4.0) but no differences was found in supratentorial lesions. Compared to CCM-DVA lesions type I and II, type III has a significant higher rupture rate ($p = 0.001$ [OR] 2.8; $p = 0.026$ [HR] 1.6), which was effective for both supratentorial and infratentorial lesions. Besides, CCM lesion volume > 1 cm^3 ($p < 0.001$ [OR] 3.3, [HR] 2.2) were presented to be the independent risk factor of CCM-DVA lesions hemorrhage. On the other hand, female sex, multiple lesions, functional zone, and left side lesion did not show any differences between ruptured/unruptured lesions.

To further explore the factors associated with lesion bleeding, we conducted a subgroup discussion and analysis of several significant factors as followed:

CCM-DVA lesion types

There was a significant difference between the rupture probabilities of the three CCM-DVA types. Type I and type II lesions had a similar rupture rate, while type III lesions harbor a higher rupture risk ($p = 0.001$ [OR 2.8], $p = 0.026$ [HR 1.6]) (Table 2), which is significant for both supratentorial ($p = 0.031$, [OR 2.8]) and infratentorial ($p = 0.009$, [OR 3.0]); furthermore, a shortened hemorrhage-free survival time was found in type III lesions than the other two types (Fig. 3a).

Functional zone

Unfortunately, the statistical results do not reflect the inevitable risk relationship between the functional area and the lesion rupture. Although lesions located in non-functional areas are more likely to achieve longer hemorrhage-free survival ($p = 0.002$, [HR 1.9], Table 2) (log-rank, $p < 0.001$, Fig. 3b), statistical results are insufficient to support their prediction of lesions rupture.

Size of the lesion

In general, ruptured lesions tend to have a relatively large volume, the volume > 1 cm^3 was closely related to lesions

Table 1 Summary of demographic information of patients with DVA associate with CCM ($n = 383$). Summary of demographic information of patients with DVA associate with CCM ($n = 319$)

Variables		No. (%)
Age (year)		41.2 ± 11.9
Sex	Male	143 (45%)
	Female	176 (55%)
Multiple CMs		78 (24%)
Eloquence		199 (62%)
Infratentorial location		161 (50%)
Initial manifestation		
Symptomatic hemorrhage		148 (46%)
Epileptic seizure		48 (15%)
Non-hemorrhage headache/dizziness		50 (16%)
Neurologic deficit		54 (17%)
Asymptomatic		19 (6%)
CM-DVA type		
I		62 (19%)
II		27 (8%)
III		230 (72%)
CCM scale		
Zabramski I		109 (37%)
Zabramski II		154 (46%)
Zabramski III		39 (11%)
Extralesional hemorrhage		17 (5%)
Maximum lesion size (mm)		
T2WI		15.64 ± 8.56
SWI		18.43 ± 8.27
Enhanced T1WI		15.53 ± 8.26

rupture ($p < 0.001$, [OR 3.3] [HR 2.2]), and a larger lesion volume are associated with a shortened hemorrhage-free survival time (log-rank, $p < 0.001$) (Fig. 3c), this conclusion is still valid when it is divided into separate discussions: supratentorial and infratentorial (Tables 3 and 4).

Infratentorial locations

From the statistical data of 319 consecutive patients, the incidence of infratentorial CCM-DVA was basically the same as that supratentorial, but the infratentorial lesions had a significantly higher rupture ratio ($p < 0.001$; OR 3.1). The Kaplan-Meier curve also shows a higher hemorrhage tendency of infratentorial lesions ($p = 0.0002$, Fig. 3d) Specific to each zone, the patients number of pontine lesions was the highest, so does the probability of lesion rupture (58/95, 61%). In addition, we found it that midbrain lesions ($p = 0.019$), pontine lesions ($p = 0.007$), and medulla lesions ($p = 0.015$) were more likely to present with symptomatic hemorrhage (Table 5).

Discussion

DVA is one of the most common cerebrovascular malformations in the brain. It is thought to be formed during embryonic development [28, 29]; however, CCM-DVA is thought to be acquired and developed. The latest study by Waleed et al. found that the incidence of CCM-DVA increased with age [30]. Meng et al. found that the prevalence of CCM in DVA patients was significantly higher than that in the normal population (11.1% vs 2.3%) [12]. In addition, several studies suggested that CCM with DVA had higher annual hemorrhage rate which is inseparable from the formation mechanism [31, 32]: DVA is thought to be more likely to be a congenital vascular malformation that precedes cavernous vascular malformations. A number of pathological studies

Table 2 Results of risk factors for hemorrhage in patients with CCM-DVA

Variables	Ruptured ($n = 141$)	Unruptured ($n = 178$)	Univariate analysis	Multivariate analysis			
			p value	*p* value	OR	*p* value	HR
Female sex	79 (56%)	96 (47%)	0.657	–	–	–	–
Age	40.2 ± 11.1	42.0 ± 12.5	0.189	–	–	–	–
Prior hemorrhage	53 (38%)	31 (17%)	< 0.001	0.003*	2.5 (1.4–4.4)	< 0.001*	2.3 (1.6–3.3)
Multiple CMs	36 (26%)	42 (24%)	0.689	–	–	–	–
Functional zone	107 (76%)	92 (52%)	< 0.001	0.113	–	0.002*	1.9 (1.3–2.8)
Infratentorial	91 (65%)	70 (39%)	< 0.001	< 0.001*	3.1 (1.9–5.1)	0.084	–
Lesion size > 1 cm^3	88 (62%)	57 (32%)	< 0.001	< 0.001*	3.3 (2.0–5.4)	< 0.001*	2.2 (1.6–3.2)
CM-DVA association type							
Type I	18 (13%)	33 (19%)	0.004	1	1	1	1
Type II	8 (6%)	30 (17%)					
Type III	115 (82%)	115 (65%)		0.001	2.8 (1.5–5.1)	0.026*	1.6 (1.1–2.5)

*Statistically significant

Table 3 Results of risk factors for hemorrhage in patients with infratentorial CCM-DVA

	Ruptured ($n = 90$)	Unruptured ($n = 70$)	Univariate analysis p value	Multivariate analysis p value	Odds ratio
Female sex	54 (60%)	42 (60%)	0.933	–	–
Age (years)	40.4 ± 10.5	42.0 ± 11.4	0.371	–	–
Prior hemorrhage	40 (44%)	13 (19%)	0.001	0.002*	4.0 (1.7–9.5)
Multiple CMs	20 (22%)	15 (21%)	0.933	–	–
Functional zone	87 (97%)	61 (87%)	0.051	0.281	–
Lesion size > 1 cm^3	53 (59%)	19 (27%)	< 0.001	0.003*	2.9 (1.4–5.8)
CM-DVA MRI appearance					
Type I	11 (12%)	17 (24%)	0.065	1	1
Type II	8 (9%)	9 (13%)			
Type III	72 (80%)	44 (63%)		0.009*	3.0 (1.3–6.8)

*Statistically significant

suggest that it is abnormally dilated venous plexus interspersed with normal insular brain tissue without arterial or arteriovenous short circuit, the area lacks normal draining veins. For the presence of DVA, changes in hemodynamics resulted in increased venous pressure, vascular torsion, and deformation with times, local thrombosis formed, and vascular lumens were further damaged and stenosis, which affected DVA angioarchitecture, leading to microbleeding and angiogenic proliferation. New blood vessels were less stable and therefore more prone to microhemorrhage, forming a cavernous structure with clustered vascular masses and accompanied by bleeding from different periods [13, 33, 34].

Previous studies have generally agreed that CCM-DVA have a higher rupture risk than those with CCM alone, and that infratentorial lesion is an important risk factor for predicting the bleeding of the lesion, natural history reported that the annual bleeding rate of unruptured CCM around the brain stem was about 2.3–8.7% [35–38], significantly higher than that of supratentorial lesions. However, it is still unclear whether the hemorrhage event of CCM-DVA is related to the infratentorial location. According to our results, the supratentorial lesions did not present any high incidence but with a lower rupture risk than infratentorial lesions, although some multiple lesions may have been neglected. The reasons may vary. First of all, the distribution of infratentorial functional areas are relatively concentrated and the distribution of drainage veins are dense, any enlargement of the lesion caused by a small amount of bleeding may lead to a sudden neurological dysfunction due to the occupying effect, a same bleeding event may remain asymptomatic at supratentorial or may

Table 4 Results of risk factors for hemorrhage in patients with supratentorial CCM-DVA

	Ruptured ($n = 50$)	Unruptured ($n = 108$)	Univariate analysis p value	Multivariate analysis p value	Odds ratio (95% CI)
Female sex	26 (40%)	24 (54%)	0.815	–	–
Age < 40 (years)	39.8 ± 12.1	42.0 ± 13.2	0.332	–	–
Prior hemorrhage	13	18	0.169	0.449	–
Multiple CMs	16 (24%)	27 (30%)	0.358	–	–
Functional zone	20 (38%)	31 (27%)	0.158	0.272	–
Deep	10 (21%)	15 (15%)	0.328	–	–
Left side	31 (48%)	47 (50%)	0.031	0.133	–
Lesion size > 1 cm^3	35 (58%)	38 (30%)	< 0.001	< 0.001*	4.0 (1.9–8.3)
CM-DVA MRI appearance					
Type I	7 (11%)	27 (33%)	0.014	1	1
Type II	0 (2%)	10 (8%)			
Type III	43 (73%)	71 (72%)		0.031*	2.8 (1.1–7.0)

*Statistically significant

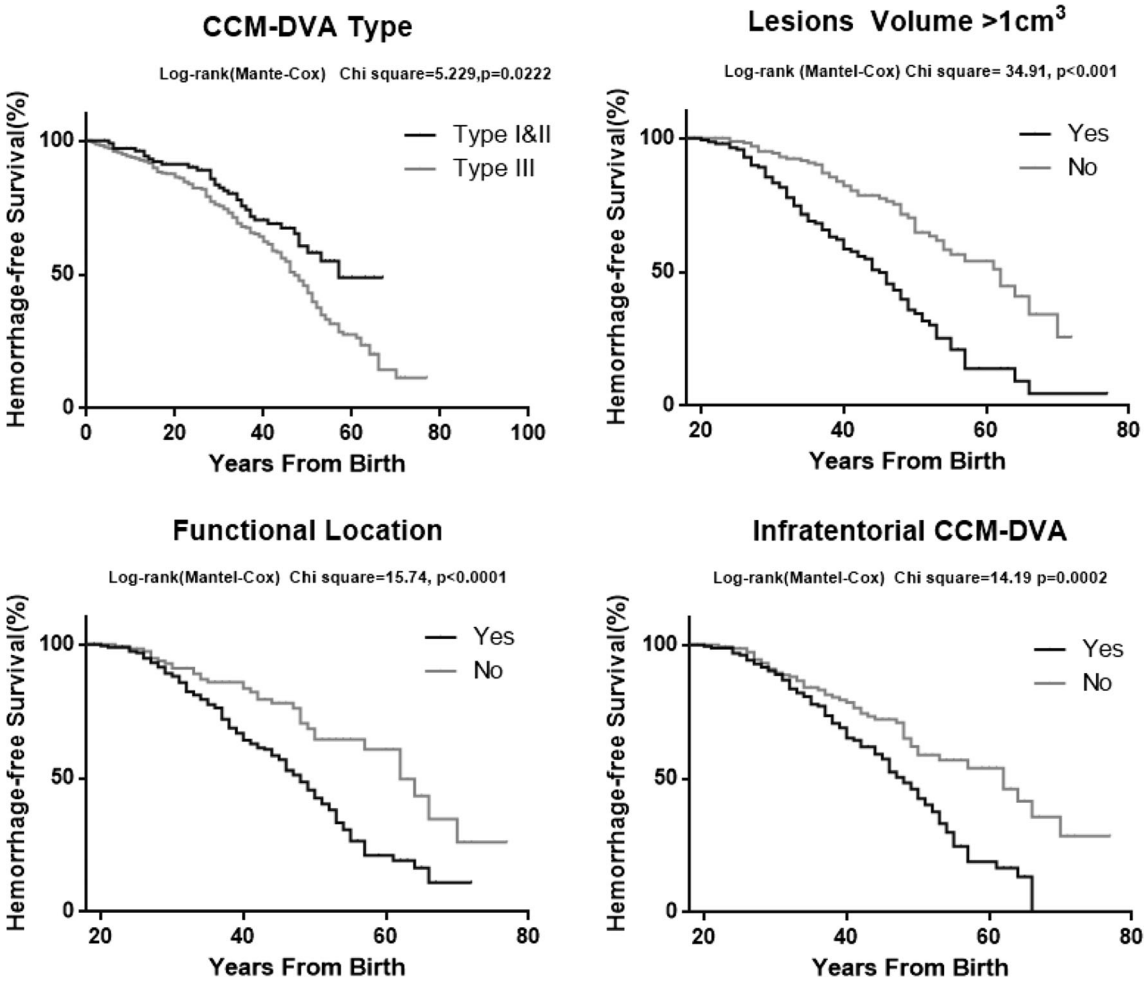

Fig. 3 Kaplan-Meier curves demonstrating the Hemorrhage-free survival rate between CCM-DVAs with certain characteristics

appear as a headache without detection. In addition, the presence of more complex venous components may keep the CCM lesion in a relatively active state for a long time, further exacerbating the progression of bleeding events.

Prior hemorrhage history has been recognized by many studies as one of the most important reasons leading to the rupture of CCM lesions [4, 5, 15, 24]. As mentioned earlier, the combination of DVA is also recognized by most researchers as one of the risk factors that increase the hemorrhaging rate of CCM lesions; however, for CCM lesion associated with DVA, there is still no definite conclusion whether the history of prior hemorrhage has an effect on the progression of the lesion. From our statistical result, for patients with infratentorial CCM-DVA lesion, previous hemorrhaging will probably increase the risk of lesion rupture.

Measures of lesion size vary from study to study, and there is no uniform answer whether lesion size is a predictor of rupture [17, 38, 39]. Recently, Sepide et al. found that >1 cm^3 was significant in predicting supratentorial lesions rupture, while it was not significant in supratentorial lesions [17]. Similarly, lesion volume >1 cm^3 is also an important

Table 5 Comparison of rupture risk in different lesions location

Location	Ruptured ($n=197$)	Unruptured ($n=215$)	Total ($n=412$)	p value
Frontal	18 (9%)	41 (19%)	59 (14%)	NS
Temporal	13 (7%)	28 (13%)	41 (10%)	NS
Parietal	9 (5%)	26 (12%)	35 (8%)	NS
Occipital	6 (3%)	3 (1%)	9 (2%)	0.024*
Insular	2 (1%)	5 (2%)	7 (3%)	NS
Basal ganglia	10 (5%)	11 (5%)	21 (5%)	NS
Thalamus	12 (6%)	10 (5%)	22 (5%)	NS
Corpus callosum	0 (0%)	3 (1%)	3 (1%)	NS
Ventricle	1 (0%)	2 (1%)	3 (1%)	NS
Cerebellum	16 (8%)	19 (9%)	35 (8%)	NS
Midbrain	31 (16%)	19 (9%)	50 (12%)	0.019*
Pontine	58 (29%)	37 (17%)	95 (23%)	0.007*
Medulla	21 (11%)	11 (5%)	32 (8%)	0.015*

*Statistically significant

NS not significant

focal rupture predictor for ruptured lesions in our study, the CCM-DVA lesions volume was larger than unruptured lesions with statistical significance ($p < 0.001$), which effective for both supratentorial and infratentorial lesions.

In previous studies, CCM-DVA was not clearly classified. Ruiz et al. classified it into three types according to the characteristics of DVA in different stages of DSA [40]. Unfortunately, except for a small number of inpatients, most of our subjects did not have the appropriate opportunity to have DSA examination. In addition, the more authoritative CCM classification proposed by Zabramski is only used to describe the statistical data and is not applied to the final risk factor analysis in this study, mainly considering that this classification has a clear tendency for hemorrhage events. The classification methods of CCM-DVAs have been debated differently in previous studies mainly according to the anatomical structure relationship of CCM-DVA and hemodynamics of venous drainage [41, 42]. Hong YJ et al. considered the occurrence of CCM-DVA may be related to anatomical angioarchitectural factors such as stenosis of DVA lumen and degree of vascular tortuosity [43].

Considering these factors, we divided CCM-DVA lesions into three types to evaluate the influence on hemorrhage through careful observation and induction of MRI imaging data of the technicians. From the statistic results, we considered that CCM lesions located around the distal radical branch of typical DVA will possess a relatively high probability of rupture (type III) than CCM associated with single abnormally venous drainage (type I) and CCM located in the trunk of the DVA with multiple abnormal vein drainage surrounded (type II).

As the most common type of CCM-DVAs, DVA-related CCM lesion is often prone to appear in the excessive vascular distortion zone, which is often small distal branches with a poor tolerance to blood flow, as shown in Figs. 1 and 2d. We think it's mostly due to the special structure of these lesions which have a significant pressure changes and irregular blood flow will consistently impact venous pressure during the running, combined with the high venous pressure existing in DVA itself. Under such unstable hemodynamic conditions, CCM structure developed from abnormal drainage vein lacks integrity and is more prone to rupture.

Limitations

This study is limited to the study and analysis of the patient's imaging features. It is difficult to completely determine the existence and exact relationship of CCM venous drainage during the screening process. The patients in the group are all clearly defined in the venous drainage component. Some patients will be missed during the statistical process which makes it difficult to carry out statistics on the exact incidence rate. Besides, with 3T MRI, some small DVA cannot be clearly identified and the relationship with CCM cannot be determined; a higher MRI field strengths MRI may improve accuracy to some extent.

Conclusion

CCM combined with DVA is a common form of intracranial vascular malformation; infratentorial lesions with prior hemorrhage history, brainstem lesions, lesion volume > 1 cm^3, type III lesion are closely related to the rupture of CCM-DVA lesions.

The discovery of these risk factors will provide a basis for subsequent treatment strategies and surgical indications and it is necessary to conduct long-term follow-up of patients to observe disease progression and conduct more research on their natural history.

Funding This study was funded by the National Natural Science Foundation of China (81371292) and the National Natural Science Foundation of China (H0906 81801140).

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

Ethical approval For this type of study formal consent is not required.

Informed consent Informed consent was obtained from all individual participants included in the study.

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