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Cranial Dural Arteriovenous Fistula: Predisposing Factors to Aggressive Symptoms

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ABSTRACT

Objective: To evaluate the clinical and angiographic data of cranial dural arteriovenous fistula in Thai patients and to determine the predisposing factors to aggressive symptoms.

Methods: Retrospective review of 71 patients with cranial dural arteriovenous fistula who underwent cerebral angiography at Siriraj Hospital between July 2002 - February 2006. Their clinical manifestations were classified as benign or aggressive symptoms. The relationship between aggressive symptoms and the following factors were studied: gender, location of the fistula, classification according to venous drainage pattern, presence of cerebral sinus thrombosis, and shunt multiplicity. Linear-by-linear association and chi-square test were used to determine statistical significance.

Results: Sixteen (22.5%) of 71 patients with cranial dural arteriovenous fistula had aggressive presenting symptoms. Factors that significantly correlate with aggressive symptoms were location of the fistula, retrograde leptomeningeal venous drainage, presence of cerebral sinus thrombosis, and multiplicity. Factor which was not significantly correlated with aggressive symptoms was gender.

Conclusion: This study evaluated clinical data and angiographic features of cranial dural arteriovenous fistula in Thai patients. Predisposing factors to aggressive symptoms were location of the fistula, retrograde leptomeningeal venous drainage, presence of cerebral sinus thrombosis, and multiplicity.

Keywords: Aggressive symptoms; cerebral sinus thrombosis; cranial dural arteriovenous fistula; retrograde leptomeningeal venous drainage

Siriraj Med J 2007; 59: 114-118
E-journal: http://www.sirirajmedj.com

Cranial dural arteriovenous fistulas (cranial dAVFs) are abnormal arteriovenous shunts localized to the intracranial dura mater, accounting for 10-15% of all intracranial arteriovenous lesions.[1] The symptoms and prognosis of the patients with cranial dAVFs can be highly variable. Some cranial dAVFs produce only tinnitus or ocular symptoms while others are associated with neurological deficit or intracranial hemorrhage. It has been generally recognized that the venous drainage pattern determines the clinical presentation and prognosis.[1,2,3] The absence of retrograde leptomeningeal venous drainage can be considered a potential predictor of a benign clinical course, and therapeutic effort should be directed toward palliation rather than toward an angiographically verifiable cure.[4] In contrast, the presence of retrograde leptomeningeal venous drainage correlates with aggressive neurological presentation and warrant more vigilant therapeutic strategy.[3] Other predisposing factors to aggressive symptoms in the patients with cranial dAVFs are detected during the study.

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To our knowledge, there have been no published reports that evaluate cranial dAVFs in Thailand. The purpose of our study was to evaluate the clinical and angiographic data of cranial dAVFs in Thai patients and to determine the predisposing factors to aggressive symptoms.

MATERIALS AND METHODS

Angiographic reports of the patients who underwent cerebral angiography at Siriraj Hospital from July 2002 to February 2006 were retrospectively reviewed to identify the patients with cranial dAVFs. Among these, 75 cranial dAVF patients were identified. Their medical records and cerebral angiograms were analyzed for clinical data and angiographic data.

Clinical data

The patient's age and gender were noted. Their presenting symptoms were classified under the same criteria of previous reports into two groups: benign symptoms and aggressive symptoms.[1,3] The benign symptoms include tinnitus, cranial nerve palsy, and ocular symptoms not

TABLE 1. Presenting symptoms in 71 patients.

Presenting symtoms	No. of patients (% within group)
Benign symptoms	55
Ocular symptoms	45 (81.82%)
Tinnitus	24 (43.64%)
CN palsy	19 (34.55%)
Trigeminal neuralgia	1 (1.82%)
Asymptomatic	1 (1.82%)
Aggressive symptoms	16
Non-hemorrhagic neurological deficit	8 (50%)
Seizure	4 (25%)
Intracranial hemorrhage	2 (12.5%)
Increased intracranial pressure	2 (12.5%)

Note: Some patients presented with more than one symptom.

related to intracranial hypertension. The aggressive symptoms include intracranial hemorrhage, non-hemorrhagic neurological deficit other than cranial nerve palsy, seizure, altered mental status, and signs of increased intracranial pressure.

Angiographic data

The locations of cranial dAVFs were recorded and identified as cavernous sinus, transverse-sigmoid sinus, superior sagittal sinus, or other sinuses. The fistulas were classified into type I, II, or III according to Borden classification[2] and into type I, IIa, IIb, IIa+b, III, IV, or V according to Cognard classification[1]. They were also classified by simple classification into presence or absence of retrograde leptomeningeal venous drainage.

Presence of cerebral sinus thrombosis was angiographically defined as: total occlusion of sinus (such as abrupt cessation of contrast medium passage in the dural sinus), reversed flow in the non-involved part of the sinus and reflux into the other sinuses or cortical vein, or stasis of the contrast medium at the point of thrombosis.[5]

Multiplicity of cranial dAVFs was defined as the occurrence of two or more fistulas that are anatomically separated within one patient, either synchronous (occurred at the same time) or metachronous (occurred at different time).[6] Bilateral cavernous sinuses were classified as single lesions.

To assess the association between symptoms (benign or aggressive) and other variables, linear-by-linear association and chi-square test were used. Linear-by-linear association was used for ordinal variables (location, classification according to venous drainage pattern, presence of cerebral sinus thrombosis, multiplicity). Chi-square test was used for nominal variables (gender). SPSS statistical software version 11.5 was used for the statistical analysis. A p value of less than 0.05 was considered significant.

RESULTS

The total of 75 patients with cranial dAVFs who underwent cerebral angiography at Siriraj Hospital between July 2002 and February 2006 were initially included in this study. Four patients were excluded because of incomplete data, thus 71 patients were included in the analysis.

Of the 71 patients, 27 (38%) were male and 44 (62%) were female. The mean age was 53.28 years (ranged 19-80 years; S.D. = 15.40). Fifty-five patients (77.5%) presented with benign symptoms with ocular symptoms as the most common presentation in this group. Sixteen patients (22.5%) presented with aggressive symptoms with non-hemorrhagic neurological deficit as the majority (Table 1).

The most frequent location of cranial dAVFs was the cavernous sinus (40 patients, 56.3%), followed by the transverse-sigmoid sinus (18 patients, 25.4%), and the superior sagittal sinus (5 patients, 7%). The remaining 8 patients had cranial dAVFs at locations other than the three mentioned sites. Their locations were classified into "others" location group. Five patients had cranial dAVFs at more than one location. As for statistical analysis, we used the location that tends to be associated with more aggressive symptoms for statistical evaluation in the following order: others, superior sagittal sinus, transverse-sigmoid sinus, and cavernous sinus; e.g., a patient with a fistula in the cavernous sinus and transverse-sigmoid sinus was evaluated as in "transverse-sigmoid sinus" group; a patient with the fistula in transverse-sigmoid sinus and torcular herophili was evaluated as in "others" location group, etc.

Most of the cranial dAVF patients (45 patients, 63.4%) did not show retrograde leptomeningeal venous drainage. When using Cognard classification, there were 22 patients (31.0%) in type I, 23 (32.4%) in type IIa, 4 (5.6%) in type IIb, 20 (28.2%) in type IIa+b, 1 (1.4%) in type III, and 1 (1.4%) in type IV. None had Cognard type V cranial dAVF (Fig 1). Under Borden classification, there were 45 patients (63.4%) in type I, 24 (33.8%) in type II, and 2 (2.8%) in type III.

Cerebral sinus thrombosis was diagnosed in 17 patients (23.9%) of patients with cranial dAVFs (Fig 2). Among these, the most common location of cerebral sinus thrombosis was the transverse-sigmoid sinus (11 patients, 64.7%), followed by the superior sagittal sinus (4 patients, 23.53%), the inferior petrosal sinus (3 patients, 17.65%), and the jugular vein (1 patient, 5.88%). Two patients had thrombosis in both the transverse-sigmoid and superior sagital sinus. The cerebral sinus thrombosis was located at almost either the sinus around the cranial dAVFs or the downstream venous flow pathways of the cranial dAVFs.

Multiple cranial dAVFs were found in 7 patients (9.9%). Two patients were diagnosed to have synchronous lesions and 4 patients with metachronous lesions. One patient had both synchronous and metachronous lesions. Some patients had two or more fistulas at different sites in the same sinus.

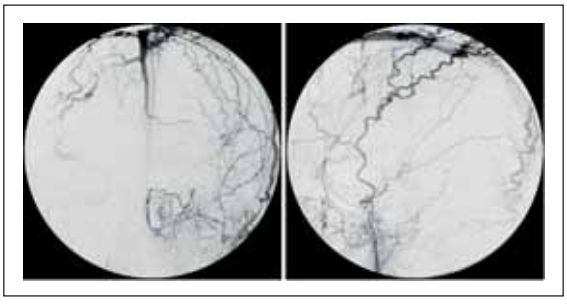


Fig 1. AP (left) and lateral (right) view of left external carotid angiogram in arterial phase, showing a cranial dAVF at superior sagittal sinus with retrograde leptomeningeal venous drainage (Cognard type IIa+b). This 47-year-old male presented with aggressive symptoms; abnormal movement of left arm.

TABLE 2. Correlation between factors and presenting symptom.

Factors	Aggressive symptom (n=16)	Benign symptom (n=55)	p value
Gender: : Male (n=27)	9 (33.3%)	18 (66.7%)	0.088[1]
: Female (n=44)	7 (15.9%)	37 (84.1%)	
Location[3] : CS (n=40)	2 (5%)	38 (95.0%)	< 0.001[2]
: TS-SS (n=18)	6 (33.3%)	12 (66.7%)	
: SSS (n=5)	4 (80.0%)	1 (20.0%)	
Cognard type[4]: I (n=22)	1 (4.5%)	21 (95.5%)	< 0.001[2]
: IIa (n=23)	2 (8.7%)	21 (91.3%)	
: IIb (n=4)	0 (0%)	4 (100%)	
: IIa+b (n=20)	11 (55%)	9 (45%)	
: III (n=1)	1 (100%)	0 (0%)	
: IV (n=1)	1 (100%)	0 (0%)	
Borden type[5]: I (n=45)	3 (6.7%)	42 (93.3%)	< 0.001[2]
: II (n=24)	11 (45.8%)	13 (54.2%)	
: III (n=2)	2 (100%)	0 (0%)	
RLVD: Presence (n=26)	13 (50.0%)	13 (50.0%)	< 0.001[2]
: Absence (n=45)	3 (6.7%)	42 (93.3%)	
Cerebral sinus thrombosis			0.001[2]
: Presence (n=17)	9 (52.9%)	8 (47.1%)	
: Absence (n=54)	7 (13.0%)	47 (87.0%)	
Multiplicity:			<0.001[2]
: Presence (n=7)	5 (71.4%)	2 (28.6%)	
: Absence (n=64)	11 (17.2%)	53 (82.8%)	

[1] Uing chi-square test

[2] Using linear-by-linear association

[3] CS = cavernous sinus
TS-SS = transverse-sigmoid sinus
SSS = superior sagittal sinus
Eight patients had the fistula in "others" location and were not statistically evaluated (5 at torcular herophili, 2 at inferior petrosal sinus, and 1 at posterior fossa vein)

[4] Cognard type I - antegrade sinus drainage
II - insufficient antegrade sinus drainage
IIa - retrograde sinus drainage only
IIb - antegrade sinus drainage with RLVD
IIa+b - retrograde sinus drainage with RLVD
III - RLVD only without venous ectasia
IV - RLVD only with venous ectasia
V - spinal venous drainage
RVLD = retrograde leptomeningeal venous drainage

[5] Borden type I - sinus drainage only
II - sinus drainage with RLVD
III - RLVD only



Fig 2. A 32-year-old male presented with tinnitus at left ear. Upper: lateral (left) and AP (right) view of left external carotid angiogram in arterial phase revealed a cranial dAVF at left transverse-sigmoid sinus, draining via contralateral transverse-sigmoid sinus. Lower: left internal carotid angiogram in arterial (left) and venous (right) phase, showing non-visualized left sigmoid sinus, representing left sigmoid sinus thrombosis.

There was statistically significant relationship between presenting symptoms and some factors (Table 2). Most of the patients with cranial dAVFs in the cavernous sinus (38 from 40 patients, 95%) presented with benign symptoms. The fistulas in the superior sagittal sinus and transverse-sigmoid sinus were significantly associated with aggressive symptoms more than the lesions in the cavernous sinus. Most patients with Cognard type I and type IIa cranial dAVFs presented with benign symptoms (95.5% and 91.3%, respectively), while those with Cognard type IIa+b, type III, and type IV were more likely to present with aggressive symptoms (55%, 100%, and 100%, respectively). There were 4 patients with Cognard type IIb cranial dAVF and all of them presented with benign symptoms. The relationship between Borden type I and benign symptoms was found and there was correlation between benign symptoms and cranial dAVFs that showed no evidence of retrograde leptomeningeal venous drainage. The significant correlation between aggressive symptoms and presence of cerebral sinus thrombosis and multiplicity was also demonstrated. In this study, however, no relationship between the gender and the aggressiveness of the presenting symptoms was found.

TABLE 3. Correlation between some factors and retrograde leptomeningeal venous drainage.

Factor	Presence of RLVD (n=26)	Absence of RLVD (n=45)	p value[2]
Location[1]: CS (n=40)	10 (25.0%)	30 (75.0%)	0.031
TS-SS (n=18)	6 (33.3%)	12 (66.7%)	
SSS (n=5)	4 (80.0%)	1 (20.0%)	
Cerebral sinus thrombosis:			0.006
Presence (n=17)	11 (64.7%)	6 (35.3%)	
Absence (n=54)	15 (27.8%)	39 (72.2%)	
Multiplicity:			0.046
Presence (n=7)	5 (71.43%)	2 (28.57%)	
Absence (n=64)	21 (32.81%)	43 (67.19%)	

[1] Eight patients with "others" location were not statistically analyzed.
[2] Using linear-by-linear association

DISCUSSION

Cranial dAVFs are generally accepted to be acquired lesions that consist of abnormal arteriovenous shunts within the dura.[3] Usually, they develop within the wall of a dural venous sinus, but other sites not directly related to a dural sinus have also been demonstrated.[1,2] Knowledge about natural history of the disease and predisposing factors to aggressive manifestations can help us provide good care to the patients.

Venous drainage pattern

It has been recognized that the venous drainage pattern determines the clinical presentation of cranial dAVFs. Correlation between retrograde leptomeningeal venous drainage and aggressive clinical manifestations has been well established.[1,2,4,7] The mechanism was due to an increase in cerebral blood volume and dural sinus pressure, resulting to venous infarction or diminution of CSF absorption.[1] Cognard[1] and Borden[2] classification that classifies cranial dAVFs according to their venous drainage pattern are widely used nowadays. Our data showed a significant correlation between retrograde leptomeningeal venous drainage and aggressive symptoms and we have been able to reconfirm the utility of Cognard and Borden classification in providing accurate prediction of aggressive behavior. However, none of the patients with Cognard type IIb cranial dAVFs presented with aggressive symptoms although they had retrograde leptomeningeal venous drainage. We supposed that it could be due to relatively small number of patients in this group in this study.

Location

The most common locations of cranial dAVFs in our study were cavernous sinus and transverse-sigmoid sinus and this was similar to previous reports in other countries.[1,5,7,8] Chung et al. reported the relation between cranial dAVFs at the location "other than the large sinuses" and the poor clinical outcome.[8] Awad et al found that no location of cranial dAVFs was free from aggressive neurological behavior. However, an aggressive neurological course was least often associated with cases involving the transverse-sigmoid sinuses and cavernous sinuses and most often associated with cases involving the tentorial incisura.[3] Cognard et al and Davies et al found that some locations were significantly correlated to aggressive manifestation because they had more tendency to coexist with retrograde leptomeningeal venous drainage. When venous drainage pattern was controlled, the location lost association with aggressive symptoms.[1,7] Our data showed that cranial dAVFs at cavernous sinus were significantly correlate to benign symptoms and the lesions in the transverse-sigmoid sinus and superior sagittal sinus had more tendency to present with aggressive symptoms.

One possible explanation was the cranial dAVFs in the cavernous sinus caused ocular symptoms, e.g., exophthalmos, eye redness that forced the patients to seek medical advice earlier in their disease course, before occurrence of retrograde leptomeningeal venous drainage. However, there was evidence that some benign cranial dAVFs, including in the cavernous sinuses, did not develop retrograde leptomeningeal venous drainage throughout their clinical course and could be spontaneously cured without any management.[4] Similar to Davies et al study[7], we found a significant relationship between location and retrograde leptomeningeal venous drainage (Table 3) and this could better explain how the location is correlated to the presenting symptoms.

Cerebral sinus thrombosis

Cerebral sinus thrombosis has been associated with cranial dAVFs. It has been suggested that physiological arteriovenous fistulas that are normally present between perforating arterioles and meningeal veins expand to form pathological fistulas as a result of sinus thrombosis.[2] However, the exact mechanism of this formation has not as yet been described in detail.

Tsai et al. proposed that cerebral sinus thrombosis may force retrograde leptomeningeal venous drainage, predisposing patients to a more aggressive neurological course.[5] Their study on 69 cranial dAVF patients in Taiwan showed that 39% of patients were associated with cerebral sinus thrombosis. However, the relationship between cerebral sinus thrombosis and aggressive manifestation could not be demonstrated in their study and they claimed this could be due to relatively small sample size.

In our study, 23.9% of patients with cranial dAVFs had associated thrombosis. The significant relationship between cerebral sinus thrombosis and aggressive symptoms was found. We also noted correlation between cerebral sinus thrombosis and retrograde leptomeningeal venous drainage (Table 3), supporting the hypothesis of Tsai et al.

Gender

In our study of 71 Thai patients with cranial dAVFs, the female outnumbered the male (F = 62%, M = 38%), similar to study of Chung et al. in Korean patients.[8] This was distinct from the studies of Davies et al.[7] and Klurfan et al.[9], studying in Canadian patients, which showed rather equal amount in each gender. The studies of Cognard et al.[1], Chung et al.[8], and Klurfan et al.[9] presented significant correlation between male gender and aggressive symptoms. In this study, no thus correlation was noted.

Multiplicity

The occurrence of multiplicity in our series was 9.9% (7 from 71 patients). Significant relationship between multiplicity and aggressive symptom was demonstrated. We also noted correlation between multiplicity and retrograde leptomeningeal venous drainage (Table 3).

Vandijk et al. studied in 235 cranial dAVFs and

found that multiplicity was demonstrated in 19 patients (8.1%)[6]. They concluded that multiplicity was associated with a significantly higher percentage of cortical venous drainage and aggressive behavior at presentation similar to our study.

There were some limitations in this study. First, it was a retrospective study, so complete data could not be obtained from all patients and the angiographic data necessarily came from different machines. The second limitation was due to the small number of the patients analyzed, especially in the group that presented with aggressive symptoms.

CONCLUSION

This study provided clinical data and angiographic features of cranial dAVFs in Thai patients. Predisposing factors to aggressive symptoms were, namely: location of the fistula, retrograde leptomeningeal venous drainage, presence of cerebral sinus thrombosis, and multiplicity. However, correlation between the patient's gender and aggressive manifestations was not demonstrated in this study.

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บทคัดย่อ

ปัจจัยที่มีความสัมพันธ์กับอาการรุนแรงในโรค cranial dural arteriovenous fistula

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วัตถุประสงค์: เพื่อศึกษาลักษณะทางคลินิกและลักษณะภาพถ่ายรังสีหลอดเลือดสมองของผู้ป่วยโรค cranial dural arteriovenous fistula ในประเทศไทย และเพื่อหาปัจจัยที่มีความสัมพันธ์กับอาการรุนแรงของโรคนี้

วิธีการ: ศึกษาย้อนหลังในผู้ป่วยโรค cranial dural arteriovenous fistula จำนวน 71 คน ซึ่งได้รับการตรวจหลอดเลือดสมองโดยฉีดสารทึบรังสีที่โรงพยาบาลศิริราช ตั้งแต่เดือนกรกฎาคม 2545 - เดือนกุมภาพันธ์ 2549 อาการทางคลินิกของผู้ป่วยจะถูกแบ่งเป็น 2 กลุ่มคืออาการรุนแรงและอาการไม่รุนแรง ความสัมพันธ์ระหว่างลักษณะต่าง ๆ ของภาพถ่ายรังสีหลอดเลือดสมองกับอาการรุนแรงจะถูกนำมาวิเคราะห์ว่ามีนัยสำคัญทางสถิติหรือไม่

ผลการศึกษา: พบผู้ป่วย cranial dural arteriovenous fistula ที่มีอาการรุนแรงทั้งหมดจำนวน 16 คน (22.5%) ปัจจัยที่มีความสัมพันธ์กับอาการรุนแรงได้แก่ ตำแหน่งของโรค, retrograde leptomeningeal venous drainage, cerebral sinus thrombosis, และ multiplicity การศึกษานี้ไม่พบความสัมพันธ์ระหว่างเพศของผู้ป่วยกับอาการรุนแรง

สรุป: ปัจจัยที่มีความสัมพันธ์กับอาการรุนแรงได้แก่ ตำแหน่งของโรค, retrograde leptomeningeal venous drainage, cerebral sinus thrombosis, และ multiplicity