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The clinical features, angiographic and endovascular treatment of indirect carotid cavernous fistulas

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Abstract:

Indirect carotid cavernous fistulas (ICCFs) are rare disease entities triggered by indirect low flow between the cavernous sinus and the meningeal branches of the internal or external carotid artery or from both. To evaluate clinical characteristics, angiography, and relationship to the result of endovascular treatment of indirect carotid cavernous fistulas. A prospective study of 32 patients diagnosed to identify ICCFs through digital subtraction angiography (DSA) and intravascular intervention. Results showed that indirect carotid cavernous fistulas were more common in females of advanced age (68.8%) and occurred spontaneously (87.5%); the most common symptoms included chemosis (93.75%), headache (87.5%), proptosis (53.1%), and tinnitus (37.5%) with type D being the most common (81.2%). Thirty out of thirty-two patients with intervention had complete flow block (83.3%), near complete (13.3%), and failure in one case. Incomplete occlusion interventions and failures were of the type D complication. The complication and number of drainage veins are related (p<0.05). ICCFs have diverse symptoms that depend on the drainage veins but most commonly were red eyes, headache, and protruding eyes. The ICCFs were mostly D-type formation and endovascular treatments were safe and effective. Common complications occurred in the case of many veins draining.

Keywords: **angiography, endovascular treatment, indirect carotid cavernous fistula.**

Classification number: **3.2**

Introduction

An indirect carotid cavernous fistula is a rare condition that is caused by the indirect low flow between the cavernous sinus and meningeal branches of the internal or external carotid artery or from both. This disease is common in females over 50 years old. Indirect carotid cavernous fistulas have atypical symptoms like direct carotid cavernous fistulas [1]. Symptoms depend on the flow rate and drainage vein patterns [2, 3]. Currently, there are many methods to evaluate blood vessels such as ultrasound, computed tomography of multisequence cerebral vessels, brain magnetic resonance imaging, and, especially, DSA, which is the gold standard to detect CCFs. In the past, treatment of ICCFs included symptom treatment and constriction of the carotid artery, which had low success rates and many complications. Endovascular treatments were carried out all over the world in the 1970s [4], followed by further development by other authors. Therefore, the objective of our study is to analyse clinical features, cerebral angiography, and the relationship with the outcome of endovascular treatment in patients with ICCFs.

Subjects and methods

Subjects: 32 patients diagnosed to identify ICCFs through DSA and intravascular intervention at Bach Mai Hospital from July 2018 to August 2019.

Methods: Prospective research. All cases meeting the selected criteria were admitted to the study to collect data on age, sex, clinical features, imaging characteristics of cerebral angiography, results of endovascular treatment, and post - intervention symptom improvement. SPSS 22.0 was used to process data.

Results

Demographic features

The age and sex characteristics of the research's subjects are presented in Table 1.

Table 1. The age and sex features of subjects.

Age	Male	Female	p
<50	1 (14.3%)	6 (85.7%)	0.00
≥50	3 (12%)	22 (88%)	
Mean	59.4±14.6		

Clinical features

Figure 1 showed that the most common symptoms included chemosis (93.75%), headache (87.5%), progressive vision loss (53.1%), proptosis (53.1%), and tinnitus (37.5%).

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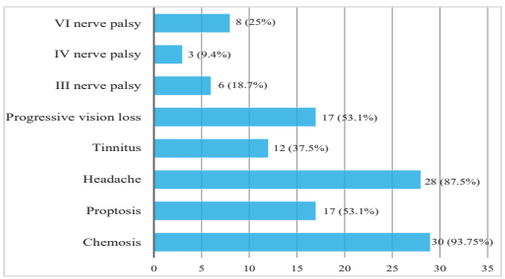

Fig. 1. Clinical features.

Imaging characteristics of cerebral angiography: In 32 cases, 7 cases had bilateral connection (21.9%), 12 cases had connection to the left cavernous sinus (37.5%), and 13 cases had connection to the right cavernous sinus (40.6%).

The features of feeding arterial branches in DSA are presented in Table 2.

Table 2. The features of feeding arterial branches in DSA.

Feeding arterial branches		Number	Rate (%)
Internal carotid artery	Right	4	12.5
	Left	5	15.6
	Bilateral	20	62.5
External carotid artery	Right	7	21.9
	Left	8	25
	Bilateral	14	43.7

In Table 2, most of cases had feeding arterial branches from both sides.

The ophthalmic vein accounted for 96.9% of draining veins, followed by the inferior petrosal sinus (34.4%) and occlusion (21.9%) (Table 3).

Table 3. Other features in DSA.

Draining veins	Number	Rate (%)
Ophthalmic vein	31	96.9
Inferior petrosal sinus	11	34.4
The cortical venous reflux	5	15.6
Contralateral drainage	6	18.7
Occlusion	7	21.9

A 60-year-old female patient was admitted due to left ptosis and tinnitus. Her DSA pictures are described in Fig. 2.

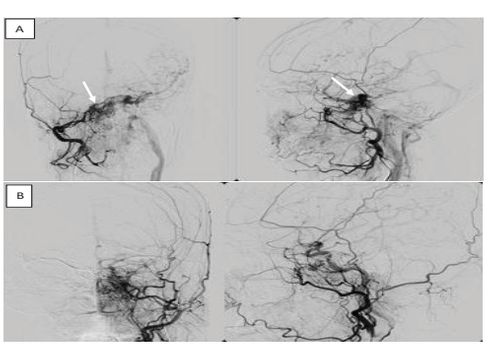

Fig. 2. DSA pictures before and after intervention. (A) ICCFs (arrow): Feeding artery from the external carotid artery bilaterally with cortical venous reflux, **(B)** After intervention: Total occlusion of the left cavernous sinus and disappearance of ICCFs.

Type D had the highest rate with 81.2% while type B and C shared the same proportion, accounting for 9.4% (Fig. 3).

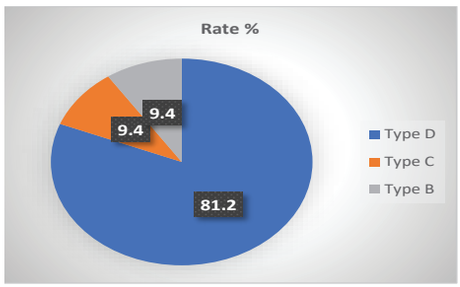

Fig. 3. Classification of flow according to Barrow.

Results of endovascular intervention

In Table 4, most patients had results of complete occlusion (83.3%). Only one case (3.4%) failed due to inaccessibility of the cavernous sinuses as the flow was too small to intervene.

Table 4. Results of endovascular intervention.

Endovascular intervention	Number	Rate (%)
Complete occlusion	25	83.3
Incomplete occlusion	4	13.3
Failed	1	3.4
Total	30	100

The ratios of complete occlusion in one side and bilateral were the same and accounted for 83.3% (Fig. 4).

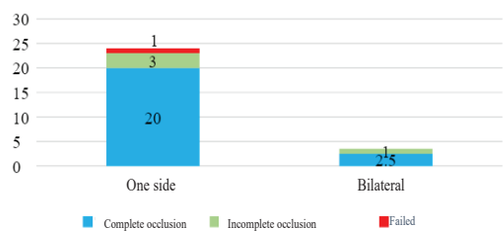

Fig. 4. The results of endovascular treatment by flow location.

Figure 5 proved that, after treatment, all symptoms decreased. Functional symptoms had a higher rate of reduction than physical signs. Chemosis reduced from 29/32 (90.6%) to 5/32 (15.6%) of cases, headache decreased from 87.5 to 12.5%, and proptosis decreased from 53.1 to 6.25% significantly with p=0.00 at a 99% confidence interval (CI).

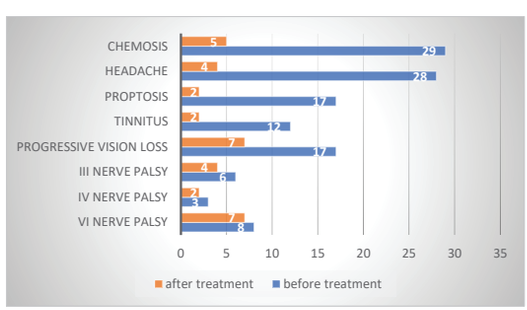

Fig. 5. Improvement of clinical symptoms after treatment.

Post - intervention complications

The most common complication after intervention was ocular palsy with 4/30 (13.33%) of cases. Three patients had VI nerve palsy, one patient had all three nerve palsies (II, IV, VI), and one patient had ischemia.

The relationship between clinical features, imaging characteristics, and results of treatment

In Table 5, symptoms of proptosis and chemosis are related to the ophthalmic venous drainage (with p=0.001 and 0.031). Tinnitus and drainage to the inferior petrosal sinus are related, and statistically significant with p=0.00, 95% confidence, OR=11.33.

Table 5. The relationship between clinical symptoms and draining veins.

Clinical symptoms		Inferior petrosal sinus		Total	p
		Yes	*No*		
Tinnitus	Yes	8 (66.7%)	4(33.3%)	12 (100%)	0.00
	No	3 (15%)	17 (85%)	20 (100%)	
	OR	11.33		2.036-63.082	
Headache	Yes	10 (35.7%)	18 (64.3%)	28 (100%)	0.573
	No	1 (25%)	3 (75%)	4 (100%)	
	OR	1.67		0.152-18.217	
Ocular palsy	Yes	5 (41.6%)	7 (58.4%)	12 (100%)	0.383
	No	6 (30%)	14 (70%)	20 (100%)	
	OR	1.67		0.347-7.424	
		Ophthalmic vein			
		Yes	No		
Proptosis	Yes	17 (100%)	0	17	0.001
	No	14 (93.3%)	1 (6.7%)	15	
Chemosis	Yes	29 (100%)	0	29	0.031
	No	2 (66.7%)	1 (33.3%)	3	
Tinnitus	Yes	11 (91.7%)	1 (8.3%)	12	0.375
	No	20 (100%)	0	20	

*p<0.05, using Fisher's Exact.

Results by types of ICCFs are provided in Table 6. All incomplete occlusion and failed cases were type D.

Table 6. The results by types of ICCFs.

	Complete occlusion	Incomplete occlusion	Failed
Type B	3	0	0
Type C	3	0	0
Type D	19	4	1
Total	25	4	1

Table 7 exhibited that there were no differences between two groups in the number of drain veins.

Table 7. The relationship between drain veins and post-intervention complication.

	Complication	No complication	Mean
The number of drain veins	4.6±0.45	1.5±0.58	2.1±1.37
p = 0.000, mean difference=3.258; 95% conference interval 2.71-3.806			

In Table 8, there was no relationship between cortical drainage veins and complications after intervention.

Table 8. The relationship between cortical drainage veins and complication.

Cortical drainage veins	Complications		Total	p
	Yes	*No*		
Yes	1 (20%)	4 (80%)	5 (100%)	0.642
No	4 (16%)	21 (84%)	25 (100%)	
Total	5	25	30	
OR	1.3		0.168-8.589	

Discussion

Feature of subjects

All patients were in adulthood with ages from 24 to 76 years old. The high prevalence rate of the group over 50 years old, which accounted for 78.1%, was higher than the group below 50 (p<0.05). Our results are similar to other studies around the world [5-8]. The rate of ICCFs in male and female were 12.5 and 87.5%, respectively. The female/male ratio was 7/1. The pathogenetic mechanism is unknown, but hormones may be involved [9, 10].

Clinical features

The clinical features of ICCFs are various. However, the symptoms are usually not as pronounced and typical as in direct CCFs. In our research, the most common symptoms were: chemosis (93.75%), headache (87.5%), progressive vision loss (53.1%), proptosis (53.1%), and tinnitus (37.5%). Symptoms manifesting in one eye accounted for 75% with the bilateral one being 25%. In Vietnam, chemosis was the most popular symptom. It was also the first reason for patients to see their doctor [11]. In a previous research, the rates of chemosis, tinnitus, and diplopia were 92.1, 5.3, and 2.6%, respectively [12]. Thus, clinical symptoms such as chemosis, headache, proptosis, tinnitus, vision loss, and ocular palsy were valuable suggestions for detecting the pathology of ICFFs.

Imaging characteristics of cerebral angiography

DSA was selected to diagnose cavernous carotid fistulas in all cases. This procedure helps to categorize lesions and choose the right treatment. The feeding arteries from the internal carotid artery in 20 cases was bilateral (62.5%), followed by the left side (15.6%) and the right side (12.5%). The feeding arteries from the external carotid artery in 14 cases were bilateral (43.7%), followed by the left side (25%) and the right side (21.9%). Thus, the majority of patients had the bilateral feeding arteries.

The characteristics of flow: 7/32 cases were bilateral (21.9%), right side (40.6%), and left side (37.5%). There were no differences between the three types.

The drain veins: 31/32 were from the ophthalmic vein (96.9%) and 11/32 from the inferior petrosal sinus (34.4%). The proportion of patients with cortical venous reflux on DSA was 15.6%, with contralateral drainage was 18.7%, and with occlusions in drain veins was 21.9%.

The location of the drainage veins was consistent with clinical symptoms. The most prevalent symptom was chemosis (93.75%) and DSA also showed that the majority of patients had venous drainage (96.9%). There was an association between tinnitus symptoms and inferior petrosal sinus drainage (p<0.05). In the study of M.D.

Alexander, et al. (2019) [13], the inferior petrosal sinus was observed and varied among patients (p=0.005). Therefore, we consider that the type of drainage vein will determine a patient's clinical symptoms.

Characteristics of classification of flow by D.L. Barrow, et al. (1985) [14]: Type D had the highest percentage with 26 patients (81.2%), which was followed by types B and C with equal rates (9.4%).

The relation between clinical features, imaging characteristics, and results of treatment

In our study, all 32 patients had DSA performed to diagnose ICCFs. There were two cases of type D that had no indication for intervention because of too small flow and insignificant ophthalmic vein. Thirty patients received intervention and 25/30 of said patients had complete occlusion (83.3%). One case of intervention failure was had right cavernous sinus flow, type D, ophthalmic vein, and inferior petrosal sinus obstruction. The intervention was conducted through the external carotid vein into the facial vein. It was failed when accessing to the ophthalmic vein due to small aperture, zigzag veins, and too small flow. According to some authors, for ICCFs with a low flow rate, the preferred treatment option is gamma knife radiotherapy. This is a minimally invasive method and is used when endovascular or surgical approaches are too dangerous or have failed [15-17]. The rate of complete occlusion in one side was 20/25 and bilateral was 5/6 (83.3%).

Complications after intervention were seen in five patients and accounted for 16.7%. To be specific, there were three cases of VI nerve palsy (6.7%), one case of ischemia (3.3%), and one case of ocular immobilization accompanied by an artery tear that formed aneurysm in the common femoral artery (3.3%). There were no cases of recent recurrence within a week, nor death. Sixth nerve palsy was treated by Solu Medrol and the patient recovered. In the case of the thigh aneurysm, emergency surgery to repair the artery was conducted.

Incomplete occlusion and failed cases were type D, possibly due to the type D flux received by the feeding arteries from both internal and external carotid arteries, thus the source of leakage was greater and the flow was higher.

The mean value of differences in the number of drainage veins of two groups with complication and no complications were 3.258.

The CI of the difference was 2.71-3.806 (p<0.05) and there was no significant difference. This result is similar to K.H. Jung, et al. (2011) [18] study with p=0.011, OR=2.189, and 95% CI from 1.198 to 3.999. The mechanism is unknown, but as can be seen in cases of multiple draining veins, when the intervention abruptly obstructs them decompensation can occur and may contribute to worsening cases.

Conclusions

ICFFs are common in females over 50 years old (68.8%). Clinical symptoms are varied, atypical, and depend on the pattern of draining veins with the most common symptoms including chemosis (93.75%), headache (87.5%), progressive vision loss (53.1%), proptosis (53.1%), and tinnitus (37.5%). Type D was the most common (81.2%). Thirty out of thirty-two patients with intervention had complete occlusion (83.3%), incomplete occlusion (13.3%), and failure in one case. Clinical symptoms related to drainage flow to the ophthalmic veins were proptosis (p<0.05) and chemosis (p<0.05). The occurrence of drainage to the inferior petrosal sinus increased 11.33 times when patients had tinnitus. The cases of incomplete occlusion and failure were type D, and complications were related to the number of drain veins (p<0.05).

COMPETING INTERESTS

The authors declare that there is no conflict of interest regarding the publication of this article.

REFERENCES

[1] A.J. Ringer, L. Salud, T.A. Tomsick (2005), "Carotid cavernous fistulas: Anatomy, classification, and treatment", *Neurosurg. Clin.*, **16(2)**, pp.279-295.

[2] N.R. Miller (2007), "Diagnosis and management of dural carotid-cavernous sinus fistulas", *Journal of Neurosurgery*, **23(5)**, DOI: 10.3171/FOC-07/11/E13.

[3] H.S. Kalish, et al. (2002), "Cavernous sinus dural arteriovenous malformations: Patterns of venous drainage are related to clinical signs and symptoms", *Ophthalmology*, **109(9)**, pp.1685-1691.

[4] F.A. Serbinenko (1974), "Balloon catheterization and occlusion of major cerebral vessels", *J. Neurosurg.*, **41(2)**, pp.125-145.

[5] M.K.S. Heran, et al. (2019), "Imaging-guided superior ophthalmic vein access for embolization of dural carotid cavernous fistulas: Report of 20 cases and review of the literature", *Am. J. Neuroradiol.*, **40(4)**, pp.699-702.

[6] D.J. Kim, et al. (2006), "Results of transvenous embolization of cavernous dural arteriovenous fistula: A single-center experience with emphasis on complications and management", *Am. J. Neuroradiol.*, **27(10)**, pp.2078-2082.

[7] M. Théaudin, et al. (2007), "Diagnosis and treatment of dural carotid-cavernous fistulas: A consecutive series of 27 patients", *J. Neurol. Neurosurg. Psychiatry*, **78(2)**, pp.174-179.

[8] K. Yoshida, et al. (2010), "Transvenous embolization of dural carotid cavernous fistulas: A series of 44 consecutive patients", *Am. J. Neuroradiol.*, **31(4)**, pp.651-655.

[9] R.M. Taniguchi, J.A. Goree, G.L. Odom (1971), "Spontaneous carotid-cavernous shunts presenting diagnostic problems", *J. Neurosurg.*, **35(4)**, pp.384-391.

[10] S. Toya, et al. (1981), "Spontaneous carotid-cavernous fistula during pregnancy or in the postpartum stage: Report of two cases", *J. Neurosurg.*, **54(2)**, pp.252-256.

[11] V.G. Nguyen, M.T. Le (2011), "Clinical evaluation of ophthalmic outcomes of embolization intervention in carotid-cavernous fistula", *Ho Chi Minh city Journal of Medicine*, **15(1)**, pp.106-112 (in Vietnamese).

[12] D.C. Nguyen, C.C. Tran (2014), "Evaluating effectiveness of cavernous sinus dural arteriovenous fistula treatment by transvenous embolisation", *Ho Chi Minh city Journal of Medicine*, **18(6)**, pp.433-440 (in Vietnamese).

[13] M.D. Alexander, et al. (2019), "Relationship of clinical presentation and angiographic findings in patients with indirect cavernous carotid fistulae", *J. NeuroInterventional Surg.*, **11**, pp.937-939.

[14] D.L. Barrow, et al. (1985), "Classification and treatment of spontaneous carotid-cavernous sinus fistulas", *J. Neurosurg.*, **62(2)**, pp.248-256.

[15] H. Baharvahdat, et al. (2020), "Updates in the management of cranial dural arteriovenous fistula", *Stroke Vasc. Neurol.*, **5(1)**, pp.50-58.

[16] C.J. Chen, et al. (2015), "Stereotactic radiosurgery for intracranial dural arteriovenous fistulas: A systematic review", *J. Neurosur.*, **122(2)**, pp.353-362.

[17] T.R. Miller, D. Gandhi (2015), "Intracranial dural arteriovenous fistulae: Clinical presentation and management strategies", *Stroke*, **46(7)**, pp.2017-2025.

[18] K.H. Jung, et al. (2011), "Clinical and angiographic factors related to the prognosis of cavernous sinus dural arteriovenous fistula", *Neuroradiology*, **53(12)**, pp.983-992.