Sonographic Clue in Non-traumatic Carotid-cavernous Fistula: Report of a Case and Literature Review

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Abstract

Background: Failure to recognize a carotid-cavernous fistula (CCF) promptly may lead to worse prognosis due to a setback in providing proper treatment. To promote early diagnosis of non-traumatic CCF, we report a case with classic clinical symptoms and signs that was diagnosed and followed up with carotid Doppler sonography (CDS) and transcranial color-coded duplex (TCD).

Case report: A 45-year-old woman developed an intermittent headache, pulsatile tinnitus, and double vision sequentially within ten days. Progressive left retro-orbital pain, continuous ringing in the left ear, sensory impairment of trigeminal nerve and abducens nerve palsy were also noted on examination. Despite insignificant findings on computed tomography (CT) of the brain, TCD revealed an aberrant flow pattern with high velocity and low resistance at the left carotid siphon. Digital subtraction angiography (DSA) later confirmed a left direct type CCF by illustrating a quick opacification of left cavernous sinus via the internal carotid artery.

Conclusion: In addition to invasive DSA, non-invasive CDS and TCD may serve as useful apparatus during the initial evaluation and subsequent follow-ups. The positive sonographic clues, including abnormal turbulent and hemodynamic parameters, are quite exhibitive in the existence of CCFs.

Keywords: Carotid-cavernous fistula, Carotid Doppler sonography, Digital subtraction angiography, Ophthalmoplegia, Tinnitus, Transcranial color-coded duplex

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INTRODUCTION

Carotid-cavernous fistula (CCF) refers to abnormal communication between the internal or external carotid arteries (or its branches) and the cavernous sinus (CS). While the incidence is not documented in the literature, trauma injuries account for 70%-75% of all CCFs [(1)]. According to the origin of the fistula, whether from internal carotid artery (ICA) or meningeal branches, CCFs can be classified into direct and indirect types. Typical clinical manifestations include subjective bruit, headache, diplopia, proptosis, chemosis, conjunctival injection and

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cranial nerve III-VI involvement. Because the disorder often masquerades as conjunctivitis, thyroid eye disease or other common ocular conditions, timely diagnosis is important to avoid irreversible complications from treatment delay [2]. While digital subtraction angiography (DSA) is the gold standard for diagnosis, transcranial color-coded duplex (TCD) remains a useful tool in the suspicion of CCF. Here we report a case of left direct CCF successfully diagnosed and followed-up with non-invasive TCD.

CASE PRESENTATION

A 45-year-old Asian woman with hypertension, poorly controlled dyslipidemia, and right high-pitch tinnitus for 3 years presented to the Emergency Department due to intermittent headache, constant pulsatile tinnitus, and double vision for about ten days.

According to the patient's statement, the symptoms began with an episodic, moderate to severe dull pain (visual analog scale 7 out of 10) over the left retro-orbital and forehead region that might last for thirty minutes. While the tenderness could subside spontaneously at first, the condition worsened with increased severity and prolonged duration in the following few days. Approximately three days later, the patient started to suffer from a continuous pulsatile ringing in the left ear without specific exacerbating and relieving factors. After another three days, acute onset of poor sight related to bi-ocular double vision developed. The diplopia persisted all day long and was more bothersome when looking leftwards. Throughout the course, she denied fever, rhinorrhea, nasal stuffiness, facial palsy, choking episode, limb weakness, recent trauma, and related family history. Apart from periorbital and carotid bruits heard over the left side, physical examination was otherwise normal without proptosis, chemosis, and conjunctival injection. Neurologic examination revealed a limitation of extraocular motion (EOM) over left eye (Figure 1A) with mild retro-orbital tenderness during an attempted lateral gaze (CNVI palsy), and a decreased pinprick sensation over ophthalmic, maxillary and mandibular (CNV1-3) region.

Laboratory workup including thyroid function and HbA1c were all within the normal range, except for the

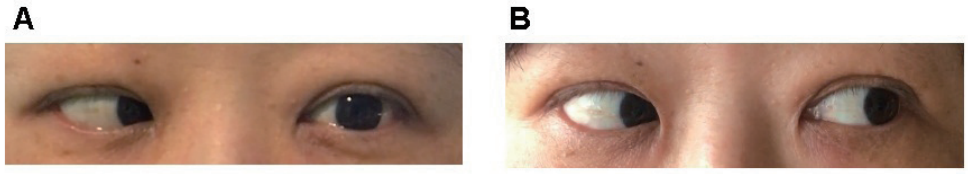

Figure 1. Extraocular motion toward left side. (A) Before the second transarterial embolization (TAE); (B) After the second TAE. The ophthalmoplegia was much improved.

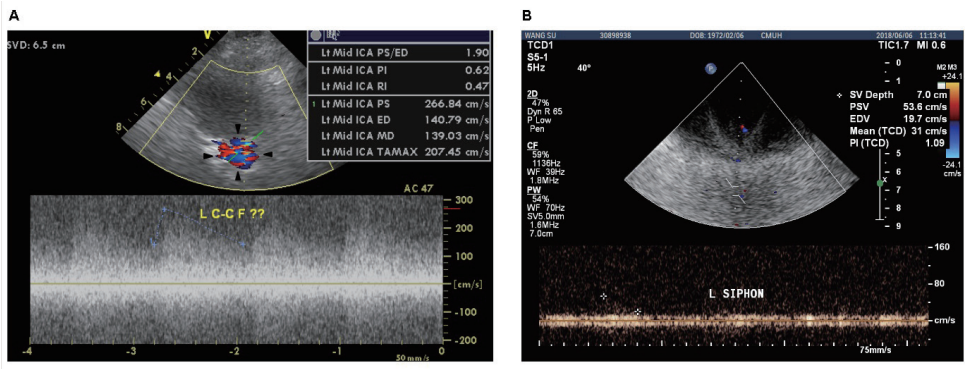

Figure 2. Transcranial color-coded duplex. (A) Before the TAE intervention, a mosaic flow flash around the left carotid siphon (arrowheads). Spectral analysis within the flash demonstrated a high-velocity, low-resistance turbulent flow pattern. (B) Eight months after the second TAE, there was no significant flow disturbance at the left carotid siphon. Spectral analysis of the hemodynamic parameters showed within a normal range.

abnormal lipid profile with elevated total cholesterol (~220 mg/dL) and LDL (~150 mg/dL). In addition to the unremarkable findings on autoimmune screening tests, there was no obvious aneurysm, no intracranial hemorrhage, nor hypodense lesion shown on the computed tomography (CT) of the brain. TCD, however, exhibited abnormal flow disturbance at left distal ICA and middle cerebral artery (MCA) (Figure 2A). Moreover, cerebral angiography demonstrated early opacification of the left cavernous sinus with retrograde filling of the engorged superior ophthalmic vein as well, thus confirming the diagnosis of a left direct (Barrow type A) CCF (Figure 3A).

The patient was then referred to interventional radiology for transarterial embolization (TAE) of the CCF. The procedure was believed to be successful as left tinnitus had significantly improved. Nevertheless, despite the administration of an anti-inflammatory agent (Prednisolone 5mg twice a day), double vision remained with deteriorated EOM limitation (from -1 to -3) during the

Table 1. Role of transcranial and carotid Doppler ultrasonography in carotid-cavernous fistulas

Authors & Year	Pt population			Signs & Symptoms	Type of CCF	Pre-intervention TCD & CDS	
	M	F	Age (yr)			Direct signs	Indirect signs
Kiliç T et al, 2001	5	3	27.5±13.9	Pulsatile exophthalmus, pain, redness, chemosis, diplopia, loss of vision, ophthalmoplegia, increased intra-ocular pressure	6 direct, 2 indirect	Abnormal flow shunt	Increased Vm (Mean= 170.4cm/s), decreased PI (mean=0.37) in all patients
Chen et al, 2000	8	7	43.3±10.8	Mainly ocular symptoms, hemorrhage may result with a superimposed aneurysm	9 direct, 8 indirect, 2 mixed	Mosaic flash (53.3%), aneurysm (15.8%)	Increased FV (78.9%), decreased RI (84.2%)
Chen et al, 1996	5	2	39.3±9.9	Proptosis, pulsatile tinnitus, and chemosis (100%); diplopia (28.6%); blurred vision, limited EOM, and headache (14.3%)	7 direct, 2 indirect, 1 mixed	Heterogenous color flash (71.4%), engorged OV (100%)	Increased flow volume (66.7%), decreased RI (66.7%)
Lin et al, 1994	2	2	40.3±7	Tinnitus and orbital bruit (75%); proptosis (50%), headache, vision loss, chemosis, ophthalmoplegia, and CN III palsy (25%)	3 direct, 2 indirect	Pulsatile turbulent flow (100%), congested OV (75%)	Increased flow volume (100%), decreased RI (80%)
Muttaqin et al, 1992	3	2	61.6±4	Chemosis (100%), orbital headache (40%), exophthalmos (40%), double vision (40%), bruit (40%), ptosis (40%)	5 indirect	Arterialization of superior OV (60%), posteriorly directed abnormal flow (40%)	Increased Vm (mean =42.4cm/s; normal 23.6cm/s) , decreased PI (mean= 0.62; normal 1.32)
Sommer et al, 1992	9	7	54.6±13.8	Bruit (31.3%), chemosis (31.3%), proptosis (31.3%), ophthalmoplegia (31.3%), spastic hemiparesis (18.8%), normal findings (12.5%)	5 direct, 11 indirect	Turbulent flow (37.5%), pendulum-like flow (6.3%), arterialized veins (25%)	Increased Vm (50% above normal reference values) in all partients

* Pt=Patient; M = Male; F = Female; CCF = Carotid-cavernous fistula; TCD = Transcranial color-coded duplex; CDS = Carotid Doppler sonography; Vm = mean flow velocity; FV = Flow volume; PSV = Peak systolic velocity; EDV= end-diastolic velocity; PI = Pulsatility index = (PSV-EDV)/Vm ; RI = Resistivity index = (PSV-EDV)/PSV

next three weeks. Consistent with enduring abnormal flow disturbance at left distal ICA and MCA found by follow-up TCD, angiography indicated a recurrent left direct CCF with early opacification of both cavernous sinuses and bilateral inferior petrous sinus during left ICA injection, which probably resulted from the displacement of coil compaction. Therefore, the patient received a repeated embolization, where an almost complete obliteration of the fistula tract with preservation of the parent artery was achieved (Figure 3B). During the postoperative visits, the patient has been free from ophthalmoplegia (Figure 1B), diplopia, eye pain and headache for two years. Also, there was no disturbing flow on the follow-up TCD checked every six months (Figure 2B), suggesting that the TAE was an effective treatment for fistula closure.

Treatment	Prognosis	Follow-up TCD & CDS
6 via endovascular balloon application, 2 via particle embolization	One developed recurrence with throbbing headache	Both Vm (Mean= 69.5cm/s) and PI (0.67) returned to normal and remained for 3 months posttreatment
-	-	-
4 of 7 patients documented, 2 underwent surgery, 2 via embolization	One developed total occlusion of ICA	Less heterogenity, improvements with diminution in flow and increased RI (66.7%)
-	-	-
-	-	-
Embolization (37.5%), operation (18.8%); spontaneous recovery (37.5%), refuse treatment (12.5%)	One developed left hemiparesis	-

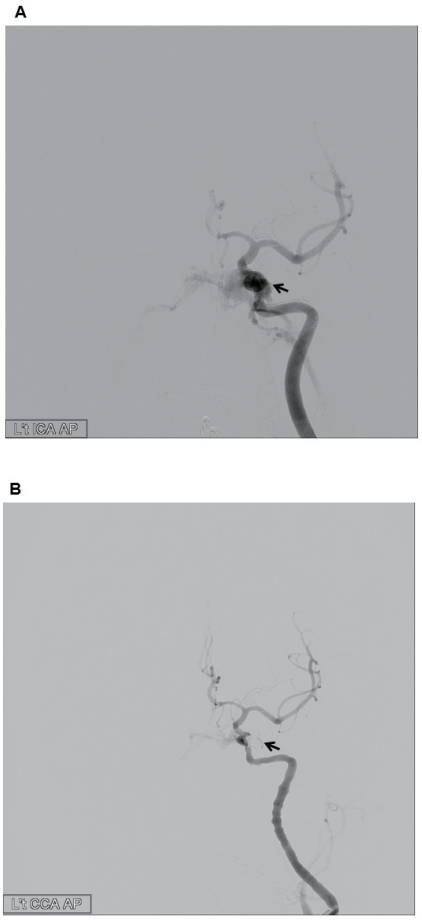

Figure 3. Antero-posterior view of the digital subtraction angiography (DSA). (A) There showed an early opacification of left cavernous sinus, implying a large fistula (arrows) at the left distal cavernous segment of the internal carotid artery (ICA). (B) After the second TAE procedure, the detachable coils (arrow) with nearly complete obliteration of the cavernous sinus fistula was demonstrated.

DISCUSSION

In general, CCFs are found evenly in men and women in any age group, with the predominance of direct type in young men after head injuries, and indirect type in middle-aged to elderly (over 50 years old) women. Although

patients with either type of CCFs may suffer from frontotemporal headache, retrobulbar pain, and pulse-synchronous bruit, those with indirect fistulas usually have a less fulminant presentation. On top of that, clinical scenario varies depending on the draining system, with ocular symptoms often caused by anterior drainage and neurologic symptoms usually resulted from posterior drainage.

In spite of an uncommon vascular malformation, a poor outcome with permanent visual and cranial nerve deficits may occur if no adequate management provided in time. With the advent of non-intrusive equipment such as CT, MRI, carotid Doppler sonography (CDS), and TCD, DSA is no longer the first and sole option to evaluate CCFs. Because of the fewer contraindications among newly developed imaging instruments, researchers began to focus on the efficacy of transcranial ultrasound in assessing and monitoring the recurrence of CCFs since the 1980s. Consistent consequences were obtained (Table 1). When there are CCFs, particularly direct fistulas, heterogenous color mosaic flashes with turbulent flow and enlarged ophthalmic veins with arterialization are the main signs seen on the TCD. The spectral analysis further ensures the existence with an elevation in flow velocity (Vm) and flow volume (FV), whereas a decline in pulsatility index (PI) and resistivity index (RI). The characteristics of hemodynamic parameters may also assist in clarifying the feeding arteries and venous drainage, with low-resistance and high-volume indicating arteries, while low-resistance and high-velocity suggesting veins. Even though the degree of alteration differs, both direct and indirect CCFs present abnormal measurements [3-5]. To advance the clinical utility of CDS and TCD, Dr. Chen and his colleague proposed an ultrasonographic classification as an aid for optimal treatment based on the presence or absence of aneurysm, direct, and indirect signs [6].

Besides the importance of CDS and TCD on investigating CCFs, they play crucial roles in long-term follow-ups after the intervention, especially in patients with indolent symptoms, negative findings on CT/MRI from the beginning or patients complicated with aneurysms. As mentioned in Chen's study in 1996, among three patients underwent CDS and TCD both before and after the treatment, two patients were found with less turbulence within the flashes, an apparent diminution in FV and an elevation in RI [7]. Kiliç's research in 2001 thereafter corroborated that the worth of CDS and TCD should not be underestimated, as significant improvement of both Vm and PI at three months post-treatment was noted in all eight patients [8]. Of the clues on ultrasonography, hemodynamic changes are better correlated with clinical status compared to direct imaging features, since the latter is difficult to quantize. As long as the values of Vm, FV, PI, and RI are improved or normalized, a successful therapeutic approach is determined.

Our patient represents a typical example that has greatly benefited from CDS and TCD in not only the initial evaluation but also the post-interventional assessment of CCFs. Since there were no decisive findings exhibited on the brain CT, the positive sonographic evidence became particularly indicative before final confirmation with DSA. In the beginning, TCD gave a crucial hint of underlying CCF by displaying a high-velocity, low-resistance flow disturbance at the left carotid siphon, which was later affirmed by DSA. Furthermore, thanks to the upfront detection of persistent turbulent flash and aberrant hemodynamic parameters on TCD after the first TAE, our patient has refrained from irrevocable sequelae with immediate fixation. Lastly, in accord with previous studies, a stable condition without recurrence was reached via periodic surveillance of CDS and TCD, where flow pattern and spectral analysis were normal.

CONCLUSION

CCF, a relatively rare form of arteriovenous fistula, can lead to severe neuro-ophthalmic morbidity and mortality if curative strategies are suspended. Though DSA remains a necessary modality in establishing the definitive diagnosis of CCF, it has a higher risk of developing complications due to the invasive property. Hence, to minimize potential iatrogenic harm, we recommend CDS and TCD as auxiliary techniques in both tracking the natural history and evaluating the response to treatment of these fistulas based on the promising results in previous research and our experience in this case. All in all, CDS and TCD are effective diagnosing and follow-up tools that can be performed quickly, inexpensively, and reproducibly.

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