


Bilateral carotid cavernous sinus fistula: a case report and review of the literature

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Abstract Carotid cavernous fistula (CCF) is an abnormal vascular shunt from the carotid artery to the cavernous sinus. They are commonly classified based on hemodynamics, etiology or anatomically. Hemodynamic classification refers to whether the fistula is high or low flow. Etiology is commonly secondary to trauma or can occur spontaneously in the setting of aneurysm or medical conditions predisposing to arterial wall defects. Bilateral carotid cavernous fistulas are rare. We present a case of bilateral CCF secondary to trauma. Ophthalmology was urgently consulted to assess the patient in the intensive care unit (ICU) for red eye. The patient was found to have decreased vision, increased intraocular pressure, an afferent pupillary defect, proptosis, chemosis, and ophthalmoplegia. Subsequent neuro-imaging confirmed a bilateral CCF. The patient underwent two endovascular embolization procedures. Trauma is the most common cause of CCF and accounts for up to 75% of cases. Most common signs of CCF depend on whether it is high or low flow. High-flow CCF may present with chemosis, proptosis, cranial nerve palsy, increased intraocular pressure, diplopia, and decreased vision. Cerebral angiography is the gold standard diagnostic modality. First-line treatment consists of endovascular embolization with either a metallic coil, endovascular balloon or embolic agent. It is unclear in

the literature if bilateral cases are more difficult to treat or have a different prognosis. Our patient required two endovascular procedures suggesting that endovascular intervention may have reduced efficacy in bilateral cases.

Keywords Proptosis · Red eye · Ophthalmoplegia

Case history

A 17-year-old female was transferred to our hospital 4 days after being involved in a motor vehicle accident, in which she was the only survivor. She was diagnosed with a diffuse axonal injury, right femoral fracture, right peroneal nerve injury, left knee dislocation, mandible fractures, and diabetes insipidus. Ophthalmology was consulted for red eye with limited extraocular movements. Initial computed tomography scan of the orbits was reported as normal. She had no significant past medical or ophthalmic history.

Physical exam

On bedside exam in the ICU visual acuity could not reliably be assessed due to sedation. Subsequent evaluation revealed a Snellen visual acuity of 20/200 in the right eye and 20/30 in the left eye. Pupils were 1 mm bilaterally with afferent defect appreciated in the right eye. Intraocular pressures measured 25 mmHg in the right eye and 21 mmHg in the left eye with Tonopen tonometry. Extraocular movements were severely restricted in all gaze directions in the right eye and full in the left. Exophthalmometry showed 2 mm of proptosis on the right side. Anterior segment examination demonstrated engorged and tortuous vessels in the right eye with segmental subconjunctival hemorrhage. There was

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segmental subconjunctival hemorrhage in the left eye. Posterior examination revealed a cup to disc ratio of 0.2 bilaterally, normal vasculature and normal peripheral retina exam (Fig. 1).

Discussion

Carotid cavernous fistula (CCF) is an abnormal vascular shunt from the carotid artery to the cavernous sinus. They are commonly classified based on hemodynamics, etiology or anatomically. Hemodynamic classification refers to whether the fistula is high or low flow. Etiology is commonly secondary to trauma or can occur spontaneously in the setting of aneurysm or medical conditions predisposing to arterial wall defects. Anatomic classification specifies whether the fistula is direct, arising from the carotid artery, or indirect; arising from one of the branches of the carotid artery.

There are four types of CCF based on Barrow's classification system as seen in the paper by Ellis et al [1]. Type A is defined as direct high-flow lesion, often resulting from a single tear in the internal carotid artery wall. Type A CCF often result from trauma or may arise from an aneurysmal rupture and account for approximately 80% of CCF's [1]. Type B CCF is low-flow indirect lesion arising from the meningeal branches of the internal carotid artery. Type C CCF arises from the meningeal branches of the external carotid artery. Type D is low flow and arises from the meningeal branches of both the internal and external carotid artery.

Trauma is the most common cause of CCF and accounts for up to 75–80% [1]. They predominantly occur in young males, who have a higher incidence of trauma. They are seen in up to 4% of patients who have sustained a basilar skull fracture [2]. CCF are thought to arise from either a direct tear from bony fracture or shear forces secondary to trauma. Spontaneous CCFs typically occur in older females as a result of ruptured internal carotid artery aneurysms or secondary to fibromuscular dysplasia, Ehlers-Danlos, and pseudoxanthoma elasticum [1].

Most common signs of CCF are dependent on whether the CCF is direct or indirect. Direct will commonly present rapidly with signs of chemosis (94%), proptosis (87%), increased intraocular pressure (60%), cranial nerve palsy (54%), diplopia (51%), and impaired vision (28%) [1, 4]. Orbital bruits, headache, and orbital pain may also be presenting features [1, 3]. Indirect CCF tends to present less dramatically with conjunctival injection often being the presenting sign, making diagnosis challenging. Cerebral angiography is the gold standard diagnostic modality. Non-contrast CT, MRI, or angiographic CT or MRI may also help to demonstrate the presence of a CCF [1].

First-line treatment consists of endovascular embolization with either a metallic coil, endovascular balloon or embolic agent. Complications include cerebral infarction, retroperitoneal hematoma, decreased visual acuity and ophthalmoplegia in up to 5% of patients [1]. In one series by Gupta et al., 88.8% of endovascular interventions for CCF were effective in curing the patient [5]. Surgical intervention may

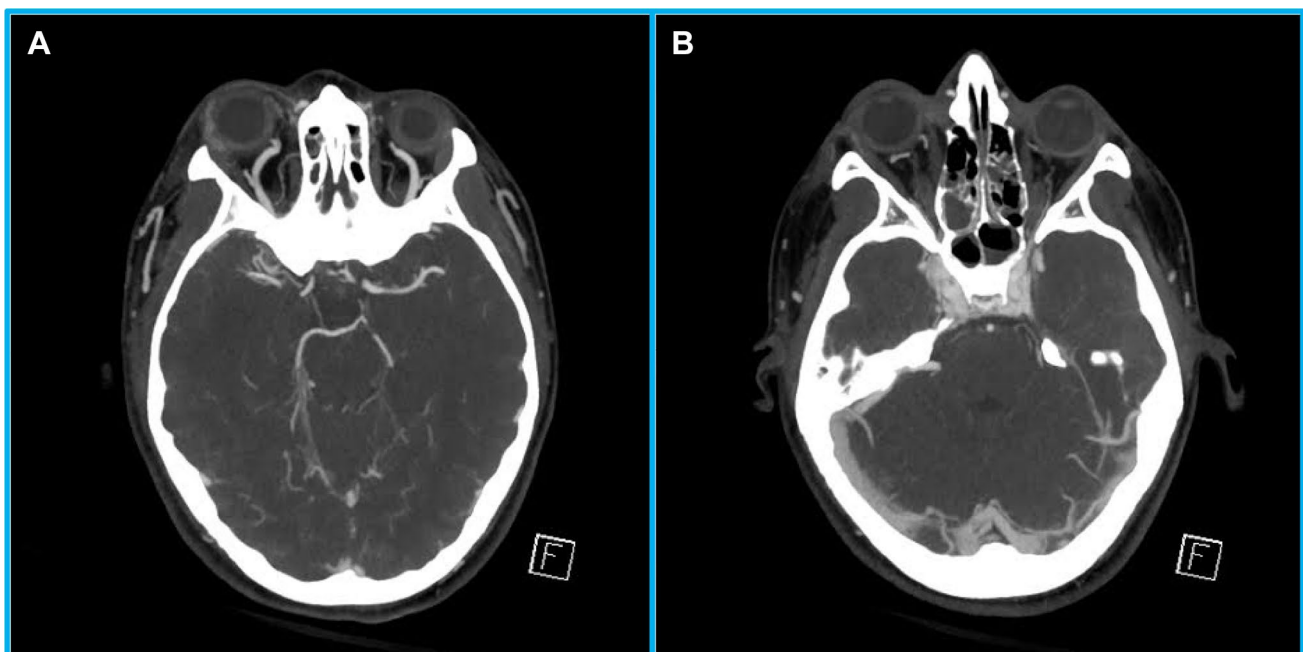


Fig. 1 a Engorged superior ophthalmic veins bilaterally. b Bilateral carotid cavernous sinus fistula with right being greater than left

Table 1 Summary of the reported cases of traumatic bilateral CCF

References	Present age, sex	Initial presentation	Treatment	Outcome
Mason et al. [7]	18, M	Right proptosis, chemosis, 6th nerve palsy, bilateral orbital bruits	Bilateral surgical clipping	CCFs resolved, persistent bilateral 6th nerve palsy
Jamieson [8]	42, M	Bilateral exophthalmos, chemosis, complete external ophthalmoplegia, and decreased visual acuity	Bilateral surgical ligation	Resolved symptoms. Bilateral complete loss of vision
Clemens et al. [9]	51, F	Bilateral exophthalmos, absence of sense of smell and taste and “progressive visual failure”	Unknown	Unknown
Curl et al. [10]	43, F	Left enophthalmos, right exophthalmos, chemosis, 6th nerve palsy	Surgical muscle embolization	CCFs resolved, symptoms resolved
Mullan [11]	19, M	Headache, left 6th nerve palsy, and decreased visual acuity	Bilateral surgical thrombosis and muscle embolization	CCFs resolved, improved vision
Roosen et al. [12]	19, M	Bilateral exophthalmos, and ophthalmoplegia, right chemosis, and facial paresis	Bilateral surgical ligation of carotid arteries, right muscle embolization	CCFs resolved, symptoms resolved
Conley et al. [13]	14, F	Comatose, bleeding from nostrils and ears, right pupil dilated and fixed, left eye inward deviation and nystagmus	Staged surgical ligation	CCFs resolved, persistent right eye blindness and ophthalmoplegia.
Dardenne et al. [14]	51, M	Bilateral ophthalmoplegia, anisocoria, and bruit	Right muscle embolization and carotid ligation, left-controlled surgical embolization	CCFs resolved, resultant motor aphasia, acalculia
Graziussi et al. [15]	37, M	Right complete ophthalmoplegia	Staged cervical and intracranial ligations of the carotids on the right side and digital cervical carotid compressions on the left side	CCFs resolved, residual subtotal ophthalmoplegia
Ambler et al. [16]	64, F	Left proptosis, chemosis, ophthalmoplegia, fixed and dilated pupil	Conservative	Death from injuries
Donnell et al. [17]	39, F	Bleeding from eyes, nose, mouth, bilateral 6th nerve palsy, upper limb weakness	Surgical muscle embolization	Death from injuries
Laws Jr. [18]	25, M	Bilateral chemosis, fixed pupils and ophthalmoplegia	Left surgical muscle embolization, and packing of fistula, right carotid ligation	CCFs resolved, ocular symptoms resolved, impaired from injury
West [19]	8, M	Bilateral proptosis and chemosis, complete bilateral ophthalmoplegia, intracranial bruit	Conservative, death before treatment initiated	Death from injuries
Matsui et al. [20]	39, M	Bilateral pulsating proptosis, chemosis, and bruit, right-sided blindness	Bilateral transarterial detachable balloon embolization	CCFs resolved, unknown patient outcome
vd Vliet et al. [21]	48, F	Right-sided ptosis, chemosis, pulsating exophthalmos, orbital edema, injection, and acute loss of vision	Conservative	Right CCF resolved, complete loss of vision
Kim et al. [22]	29, M	Right ptosis and progressive pulsating protrusion, conjunctival injection and tinnitus	Bilateral transarterial detachable balloon embolization	CCFs resolved, all symptoms resolved
Ng et al. [23]	42, F	Bilateral orbital swelling, chemosis, and periorbital bruits, right exophthalmos, and ophthalmoplegia	Bilateral transarterial detachable balloon embolization	CCFs resolved, symptoms resolved

Table 1 (continued)

References	Present age, sex	Initial presentation	Treatment	Outcome
Alkhani et al. [24]	42, F	Left-sided exophthalmos and chemosis	Conservative spontaneous resolution	CCFs resolved, gradual symptomatic improvement
Kamel et al. [25]	27, F	Complete bilateral external ophthalmoplegia, orbital bruits, bilateral ptosis and dilated unreactive pupils	Unilateral transarterial detachable balloon embolization	CCFs resolved complete symptom resolution on the left, improvement on the right
Churojana et al. [26]	62, F	Left hemiparesis with bilateral exophthalmos with chemosis and subconjunctival hemorrhage	Conservative	CCFs resolved, ocular symptoms resolved, residual neurological deficits
Sanden et al. [27]	17, M	Bilateral exophthalmos, left progressive chemosis, and unreactive mydriatic pupil	Death before embolization treatment initiated	Death before treatment initiation
Oran et al. [28]	19, M	Right periorbital swelling and progressive exophthalmos	Transarterial venous coil embolisation of a low-flow left fistula through right carotid artery tear, with transarterial detachable balloon embolisation of the right high-flow fistula	CCFs resolved, right decreased visual acuity and mild ptosis
Moron et al. [29]	20, F	Proptosis and chemosis, left ophthalmoplegia	Staged, bilateral stent-assisted coil embolization	CCFs resolved, residual ophthalmoplegia
Hantson et al. [30]	68, F	Bilateral periorbital hematoma, with right exophthalmos, dilated and fixed pupil	Bilateral transarterial detachable balloon embolization	Death from post-interventional complications
Luo et al. (7 cases) [31]	21, F	Bruit, chemosis, proptosis	Bilateral transarterial detachable balloon embolization	CCFs resolved, symptoms resolved, no individual report
	23, M	Blurred vision, chemosis, proptosis	Right transarterial detachable coil, embolization, left detachable balloon and coil	CCFs resolved, symptoms resolved, no individual report
	19, M	Bruit, chemosis	Bilateral transarterial detachable balloon and coil embolization	CCFs resolved, symptoms resolved, no individual report
	46, M	Blurred vision, bruit, chemosis	Bilateral transarterial detachable balloon and coil embolization	CCFs resolved, symptoms resolved, no individual report
	28, M	Blurred vision, bruit, chemosis, ptosis	Right transarterial detachable balloon and coil embolization, left detachable balloon	CCFs resolved, symptoms resolved, no individual report
	44, M	Blurred vision, bruit, chemosis, ptosis	Right transarterial detachable balloon embolization, left detachable balloon and coil	CCFs resolved, symptoms resolved, no individual report
	39, M	Blurred vision, bruit, chemosis, proptosis	Bilateral transarterial coil-assisted embolization	CCFs resolved, symptoms resolved, no individual report
Liang et al. [32]	68, F	Intracranial bruit, right-sided exophthalmos, chemosis, and ophthalmoplegia	Bilateral transarterial detachable balloon embolization	CCFs resolved, symptoms resolved

Table 1 (continued)

References	Present age, sex	Initial presentation	Treatment	Outcome
Gao et al. [33]	50, M	Bilateral proptosis, orbital swelling, chemosis, periorbital bruits	Bilateral transarterial detachable balloon embolization	CCFs resolved, de novo type D CCF formation treated with <i>N</i> -butyl-cyanoacrylate, Symptoms resolved
Gierthmuehlen et al. [34]	30, M	Severe headache, fixed dilated right pupil, somnolence, loud bruit on skull	Unilateral complete balloon embolization	CCFs resolved, Symptoms resolved
Cho et al. [35]	48, M	Bilateral proptosis, eyelid swelling, chemosis, bruit over both periorbital regions	Bilateral transarterial balloon-assisted coil embolization	CCFs resolved, Hemorrhages from hyper-perfusion syndrome but resolved
Yu et al. [36]	Unknown	Pulsating tinnitus, proptosis, conjunctival chemosis, Laterality or other symptoms unknown	Staged, Bilateral transarterial balloon-assisted onyx embolization	CCFs resolved, all symptoms resolved
Gapsis et al. [37]	38, F	Periorbital edema, left-dilated pupil, and ophthalmoplegia	Conservative	CCFs resolved, symptoms resolved, transient optic disc edema
Maciej et al. [38]	42, M	Left exophthalmos, 3rd and 6th nerve palsy, decreased visual acuity, pulsating noise, headache	Unilateral endovascular embolization using microembolisation coils	CCFs resolved, all symptoms resolved
Chiriac et al. [39]	57, F	Left exophthalmos, chemosis, eyelid edema, periorbital bruit and loss of vision, mild right exophthalmos	Bilateral detachable balloon embolization	CCFs resolved, persistent left loss of vision and ophthalmoplegia
Ke et al. [40]	60, M	Bilateral exophthalmos, chemosis, ophthalmoplegia, elevated intra-ocular pressure, right decreased visual acuity	Conservative	Unknown CCF outcome, persistent right visual impairment
This study	17, F	Right proptosis, chemosis, decreased visual acuity, ophthalmoplegia, elevated intra-ocular pressure	Unilateral transarterial detachable balloon embolization	CCFs resolved, persistent ophthalmoplegia

be considered in cases where endovascular treatment has been unsuccessful or is not possible. Success rates have been reported between 30 and 80% in the literature [1]. Radio-surgery appears to be effective in patients with indirect, low flow, CCF's [5].

Bilateral CCFs are rare. A recent case report and literature review article of bilateral CCF found a total of 67 cases since 1954, 41 of which were post-traumatic [6]. All the cases with known patient details, had ocular involvement at initial presentation and were type A CCFs. Surgical ligation was employed more frequently in the earlier reported cases. However, it has mainly been replaced with transarterial endovascular embolization, which was first reported in 1987 [20]. An additional three cases were found on our search of literature that were not included in this recent review [28, 38, 40]. Table 1 summarizes all cases of post-traumatic bilateral CCFs with reported patient presentation, treatment and outcome.

In this case, ophthalmic consultation was sought for red eye with decreased eye movements. Ophthalmic exam revealed significant proptosis, chemosis, and elevated intraocular pressures, which in the setting of trauma suggested a possible CCF. Review of neuroimaging by the on-call ophthalmology resident confirmed the diagnosis of a bilateral CCF. The patient underwent endovascular embolization, which, was initially successful in treating her CCF. Unfortunately, she presented 1 month later with recurrent symptoms and endovascular intervention needed to be repeated. Second attempt at endovascular intervention was successful. Her visual acuity returned to 20/25 in the right eye and 20/20 in the left using Snellen visual acuity chart. Intraocular pressure returned to a normal range. Her afferent pupillary defect resolved. Her right eye had a persistent 6th nerve palsy throughout.

We were unable to determine if bilateral CCF's vary in response to intervention compared to unilateral based on the literature available. In this case, two attempts at unilateral transarterial balloon on the most severely affected side were required to resolve the CCF.

Compliance with ethical standards

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

References

- Ellis JA, Goldstein BS, Connolly ES, Meyers PM (2012) Carotid-cavernous fistulas. *Neurosurg Focus* 32(5):E9
- Liang W, Xiaofeng Y, Weiguo L, Wusi Q, Gang S, Xuesheng Z (2007) Traumatic carotid cavernous fistula accompanying basilar skull fracture: a study on the incidence of traumatic carotid cavernous fistula in the patients with basilar skull fracture and the prognostic analysis about traumatic carotid cavernous fistula. *J Trauma* 63:1014–1020
- De Keizer R (2003) Carotid cavernous and orbital arteriovenous fistulas: ocular features, diagnostics, and hemodynamic considerations in relation to visual impairment and morbidity. *Orbit* 22:121–142
- Kirsch M, Henkes H, Liebig T, Weber W, Esser J, Golik S, Kuhne D (2006) Endovascular management of dural carotid-cavernous sinus fistulas in 141 patients. *Neuroradiology* 48:486–490
- Gupta AK, Purkayastha S, Krishnamoorthy T, Bodheey NK, Kapilamoorthy TR, Kesavadas C et al (2006) Endovascular treatment of direct carotid cavernous fistulae: a pictorial review. *Neuroradiology* 48:831–839
- Al-Mufti F, Amuluru K, El-Ghanem M et al (2017) Spontaneous bilateral carotid-cavernous fistulas secondary to cavernous sinus thrombosis. *Neurosurgery* 80:646–654
- Mason TH, Swain GM, Osheroff HR (1954) Bilateral carotid-cavernous fistula. *J Neurosurg* 11:323–326
- Jamieson KG (1964) Bilateral carotico-cavernous fistulae. Hypopituitarism from bilateral carotid ligation for surgical cure. *Aust NZ J Surg* 34:1–10
- Clemens F, Lodin H (1968) Some viewpoints on the venous outflow pathways in cavernous sinus fistulas: angiographic study of five traumatic cases. *Clin Radiol* 19:196–200
- Curl FD, Harbert JC, Luessenhop AD, Di Chiro G, Kamm RF (1972) Radionuclide cerebral angiography in a case of bilateral carotid-cavernous fistula. *Radiology* 102:391–392
- Mullan S (1974) Experiences with surgical thrombosis of intracranial berry aneurysms and carotid cavernous fistulas. *J Neurosurg* 41:657–670
- Roosen K, Grote W (1975) Diagnosis and treatment of bilateral traumatic carotid-cavernous sinus fistulae. *Neurochirurgia* 18:175–189
- Conley FK, Hamilton RD, Hosobuchi Y (1975) Successful surgical treatment of bilateral carotid-cavernous fistulas. *J Neurosurg* 43:357–361
- Dardenne GJ (1975) Bilateral traumatic carotid-cavernous fistulae. *Surg Neurol* 3:105–107
- Graziussi G, Granata F, Terracciano S (1977) Bilateral carotid-cavernous fistula of traumatic origin. A case report. *Acta Neurol* 32:347–353
- Ambler MW, Moon AC, Sturmer WQ (1978) Bilateral carotid-cavernous fistulae of mixed types with unusual radiological and neuropathological findings. *J Neurosurg* 48:117–124
- Donnell MS, Larson SJ, Correa-Paz F, Worman LW (1978) Traumatic bilateral carotid-cavernous sinus fistulas with progressive unilateral enlargement. *Surg Neurol* 10:115–118
- Laws ER Jr, Onofrio BM, Pearson BW, McDonald TJ, Dirrenberger RA (1979) Successful management of bilateral carotid-cavernous fistulae with a transsphenoidal approach. *Neurosurgery* 4:162–167
- West CGH (1980) Bilateral carotid-cavernous fistulae: a review. *Surg Neurol* 13:85–90
- Matsui Y, Yamada K, Hayakawa T et al (1987) Bilateral traumatic carotid-cavernous fistulas. Case report. *Neurol Med Chir* 27:447–450
- vd Vliet AM, Rwiza HT, Thijssen HO et al (1987) Bilateral direct carotid-cavernous fistulas of traumatic and spontaneous origin: two case reports. *Neuroradiology* 29:565–569
- Kim JK, Seo JJ, Kim YH, Kang HK, Lee JH (1996) Traumatic bilateral carotid-cavernous fistulas treated with detachable balloon. A case report. *Acta Radiol* 37:46–48
- Ng SH, Wan YL, Ko SF et al (1999) Bilateral traumatic carotid-cavernous fistulas successfully treated by detachable balloon technique. *J Trauma* 47:1156–1159

24. Alkhani A, Willinsky R, TerBrugge K (1999) Spontaneous resolution of bilateral traumatic carotid cavernous fistulas and development of trans-sellar intercarotid vascular communication: case report. *Surg Neurol* 52:627–629
25. Kamel HA, Choudhari KA, Gillespie JS (2000) Bilateral traumatic carotidocavernous fistulae: total resolution following unilateral occlusion. *Neuroradiology* 42:462–465
26. Churojana A, Chawalaparit O, Chiewwit P, Suthipongchai S (2001) Spontaneous occlusion of a bilateral post traumatic carotid cavernous fistula. *Interv Neuroradiol* 7:245–252
27. Sanden U, Grosse U, Jaksche H (2003) Visualization of bilateral carotid cavernous sinus fistulas with duplex sonography. *J Clin Ultrasound* 31:319–323
28. Oran I, Bozkaya H, Parildar M (2004) Embolisation of both fistulae through the same carotid artery tear in a patient with bilateral traumatic carotidocavernous fistulae. *Neuroradiology* 46:234–237
29. Moron FE, Klucznik RP, Mawad ME, Strother CM (2005) Endovascular treatment of high-flow carotid cavernous fistulas by stent-assisted coil placement. *Am J Neuroradiol* 26:1399–1404
30. Hantson P, Espeel B, Guerit JM, Goffette P (2006) Bilateral carotid-cavernous fistula following head trauma: possible worsening of brain injury following balloon catheter occlusion? *Clin Neurol Neurosurg* 108:576–579
31. Luo CB, Teng MM, Chang FC et al (2007) Bilateral traumatic carotid-cavernous fistulae: strategies for endovascular treatment. *Acta Neurochir* 149:675–680 (**discussion 680**)
32. Liang W, Xiaofeng Y, Weiguo L et al (2007) Bilateral traumatic carotid cavernous fistula: the manifestations, transvascular embolization and prevention of the vascular complications after therapeutic embolization. *J Craniofac Surg* 18:74–77
33. Gao BL, Zhao W, Xu GP (2009) The development of a de novo indirect carotidcavernous fistula after successful occlusion of bilateral direct carotid-cavernous fistulas. *J Trauma* 66:E28–E31
34. Gierthmuehlen M, Schumacher M, Zentner J, Hader C (2010) Brainstem compression caused by bilateral traumatic carotid cavernous fistulas: case report. *Neurosurgery* 67:E1160–E1163 (**discussion E1163-E1164**)
35. Cho K-C, Seo D-H, Choe I-S, Park S-C (2011) Cerebral hemorrhage after endovascular treatment of bilateral traumatic carotid cavernous fistulae with covered stents. *J Korean Neurosurg Soc* 50:126–129
36. Yu Y, Huang Q, Xu Y, Hong B, Zhao W et al (2012) Use of onyx for transarterial balloon-assisted embolization of traumatic carotid cavernous fistulas: a report of 23 cases. *Am J Neuroradiol* 33:1305–1309
37. Gapsis BC, Ranjit RU, Malavade S et al (2013) Spontaneous resolution of ophthalmologic symptoms following bilateral traumatic carotid cavernous fistulae. *Digit J Ophthalmol* 19:33–38
38. Maciej W, Tadeusz P, Pawel B et al (2013) Posttraumatic bilateral carotid-cavernous fistula. *J Int Adv Otol* 9:417–422
39. Chiriac A, Iliescu BF, Dobrin N, Poeata I (2014) One-step endovascular treatment of bilateral traumatic carotid-cavernous fistulae with atypical clinical course. *Turk Neurosurg* 24:422–426
40. Ke L, Yang Y, Yuan J (2017) Bilateral carotid-cavernous fistula with spontaneous resolution: a case report and literature review. *Medicine* 96(19):e6869