

# Noncavernous arteriovenous shunts mimicking carotid cavernous fistulae

Chai Kobkitsuksakul  
Pakorn Jiarakongmun  
Ekachat Chanthanaphak  
Sirintara (Pongpech) Singhara Na Ayudya

## PURPOSE

The classic symptoms and signs of carotid cavernous sinus fistula or cavernous sinus dural arteriovenous fistula (AVF) consist of eye redness, exophthalmos, and gaze abnormality. The angiography findings typically consist of arteriovenous shunt at cavernous sinus with ophthalmic venous drainage with or without cortical venous reflux. In rare circumstances, the shunts are localized outside the cavernous sinus, but mimic symptoms and radiography of the cavernous shunt. We would like to present the other locations of the arteriovenous shunt, which mimic the clinical presentation of carotid cavernous fistulae, and analyze venous drainages.

## METHODS

We retrospectively examined the records of 350 patients who were given provisional diagnoses of carotid cavernous sinus fistulae or cavernous sinus dural AVF in the division of Interventional Neuroradiology, Ramathibodi Hospital, Bangkok between 2008 and 2014. Any patient with cavernous arteriovenous shunt was excluded.

## RESULTS

Of those 350 patients, 10 patients (2.85%) were identified as having noncavernous sinus AVF. The angiographic diagnoses consisted of three anterior condylar (hypoglossal) dural AVF, two traumatic middle meningeal AVF, one lesser sphenoid wing dural AVF, one vertebro-vertebral fistula (VVF), one intraorbital AVF, one direct dural artery to cortical vein dural AVF, and one transverse-sigmoid dural AVF. Six cases (60%) were found to have venous efferent obstruction.

## CONCLUSION

Arteriovenous shunts mimicking the cavernous AVF are rare, with a prevalence of only 2.85% in this series. The clinical presentation mainly depends on venous outflow. The venous outlet of the arteriovenous shunts is influenced by venous afferent-efferent patterns according to the venous anatomy of the central nervous system and the skull base, as well as by architectural disturbance, specifically, obstruction of the venous outflow.

The cavernous sinus collects the venous blood from the tributaries of the ophthalmic venous system, the superficial middle cerebral vein and sphenoparietal sinuses and exits through the petrosal sinuses, the veins of the foramen ovale, as well as the skull base emissary veins. In the cavernous arteriovenous (AV) shunt, the arterIALIZED veins usually reverse the normal direction of their existing inlets and frequently reflux into the ophthalmic venous system resulting in clinical presentation of exophthalmos, eye redness, and pulsatile tinnitus in the vast majority of the cases. On rare occasions, the AV shunt locations are not harbored in the cavernous sinus, but drain toward the cavernous sinus and ophthalmic venous systems. Therefore, the clinical and/or noninvasive imaging imitates a cavernous AV shunt. We would like to present these locations of the AV shunt, which mimic the clinical presentation of the cavernous sinus AV fistula (AVF).

## Methods

We retrospectively examined the medical records of 350 patients who were given provisional diagnoses of carotid cavernous sinus fistulae or cavernous sinus dural AVF in the division of Interventional Neuroradiology, Ramathibodi Hospital, Bangkok, Thailand, between 2008 and 2014. All patients underwent cerebral angiographic studies. Patients with cavernous AV shunt were excluded. The clinical presentations were analyzed along with computed tomography (CT), magnetic resonance imaging (MRI), and angiography findings.

From the Division of Interventional Neuroradiology, the Department of Radiology (C.H. ✉ *dr.chai.kobkitsuksakul@gmail.com*) Ramathibodi Hospital, Mahidol University, Bangkok, Thailand.

Received 20 February 2016; revision requested 17 March 2016; revision received 13 April 2016; accepted 15 April 2016.

Published online 21 October 2016.  
DOI 10.5152/dir.2016.16073

Each patient gave informed consent before diagnostic cerebral angiography and intervention. Our institutional review board approved this retrospective study.

## Results

Of 350 patients, 10 (2.85%) were identified as having noncavernous sinus AV shunts at angiographic exams. The mean age of the patients was 40.3 years (range, 11–68 years). Four patients (36.4%) were female. All patients had redness of the eye as the presenting symptom (100%). Proptosis was present in nine patients (90%). Subjective bruit was also another frequent symptom found in 63.6% of the patients. Four patients had an AV shunt at the foramen magnum level; three of these were anterior condylar (hypoglossal canal) dural AVFs and one was a vertebro-vertebral fistula (VVF) (Figs. 1, 2). The other locations comprised two middle meningeal AVF, one lesser sphenoid wing dural AVF, one intraorbital AVF, one direct dural artery to cortical vein AVF, and one transverse-sigmoid dural AVF. We observed venous outlet obstruction/stenosis in six cases (three cases of anterior condylar AVFs, one case of VVF, one case of direct dural artery to cortical vein AVF, and one case of transverse-sigmoid dural AVF). Only two cases of middle meningeal AVF were clearly associated with craniofacial trauma.

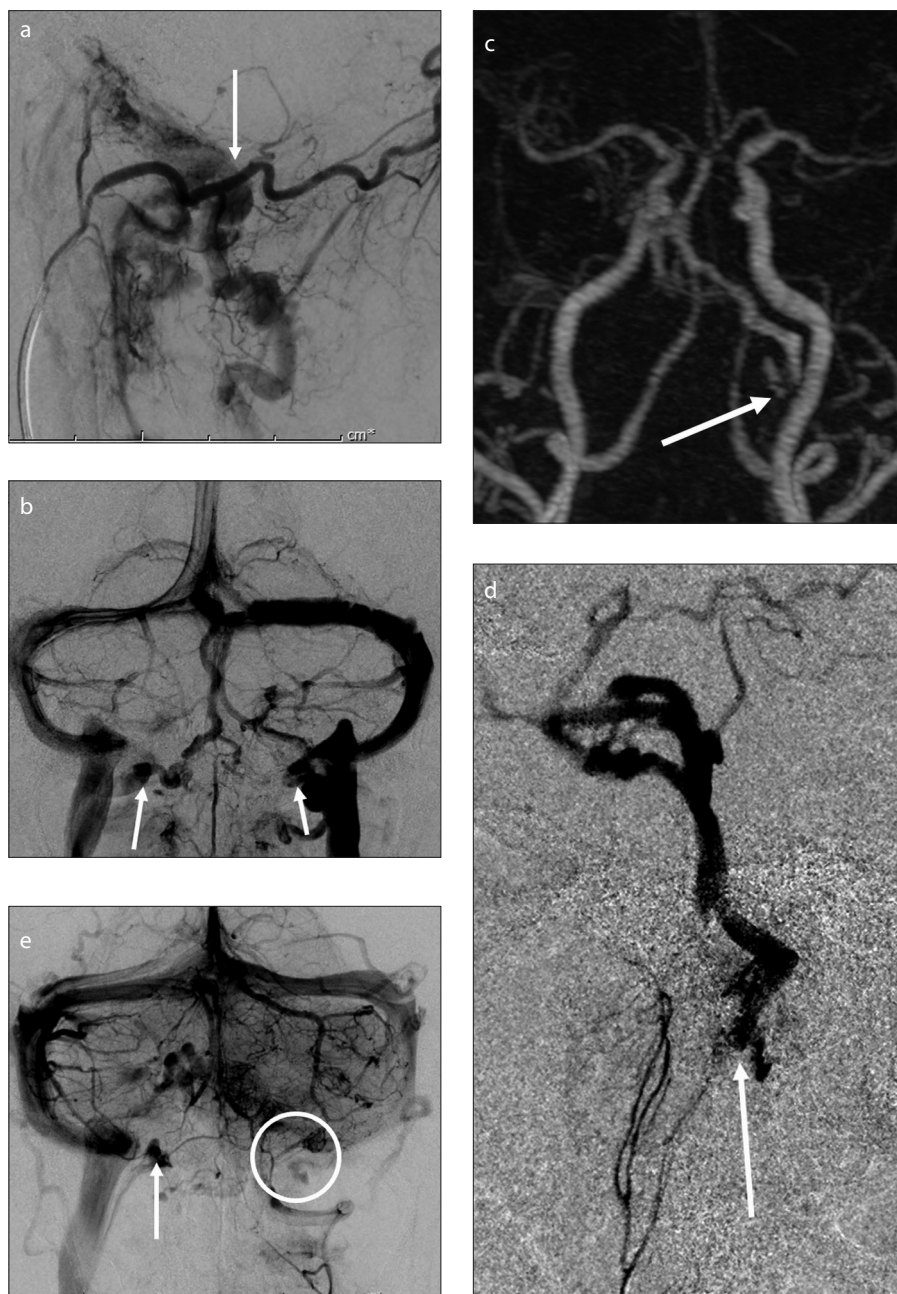
## Discussion

AV shunts mimicking cavernous AV shunts are rare. They are usually presented in small case series (1, 2). The venous interconnection of the brain and the skull base are complex. The cavernous sinuses are centrally located dural sinuses at the skull base and connect with other dural sinuses and subarachnoid vein in various ways from

the supratentorial location to the posterior fossa and the brain stem (3). Hence, theoretically, any AV shunt where the venous drainage has a connection with the cavernous sinus is able to drain within this network including the cavernous sinus.

Theoretically, in shunts located below the cavernous sinus, such as the anterior condylar dural AVF, the arterialized vein should

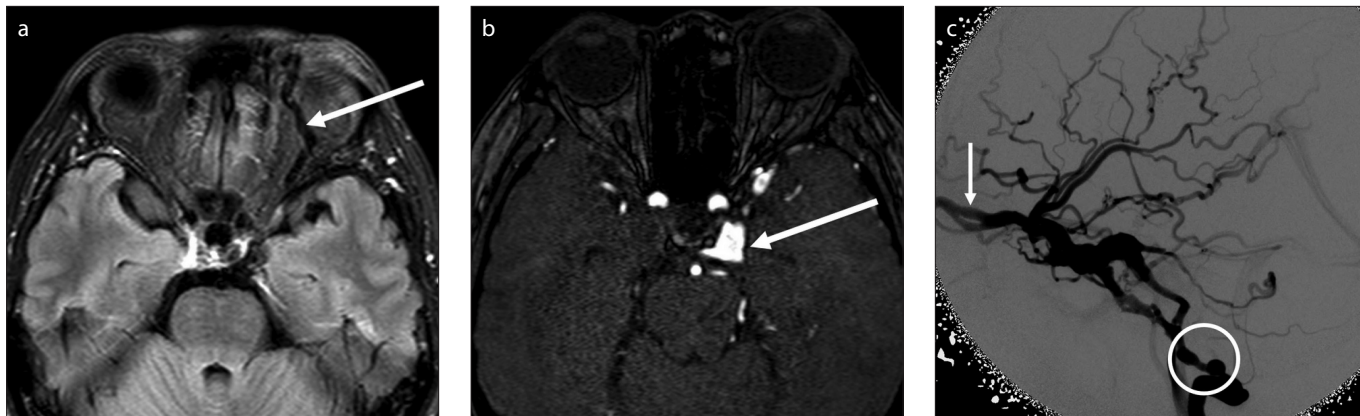
drain toward the marginal sinus, internal jugular vein, or the vertebral venous plexus that is supported by several venous outlets, as shown in human and animal studies (2, 4–9). In contrast, the vein drains upward toward the cavernous sinus. This is presumably the consequence of downstream venous outflow obstruction (Figs. 1, 2). This venous drainage pattern was also reported



**Figure 1.** a–e. The angiography images (a) and (b) show an example of anterior condylar dural arteriovenous (AV) shunt (a, arrow) in which its downstream venous drainage (marginal sinus) is patent (b, arrow). In a similar location, in one of the anterior condylar dural arteriovenous fistulae (AVF) cases in our series, time-of-flight magnetic resonance angiography (c) shows the abnormal vessels (arrow). Ascending pharyngeal arteriogram (d) shows the AV shunt with subsequent drain toward posterior cavernous sinus. Venography in the posterior circulation (e) shows left downstream venous obstruction (e, circle) created in the upward arterialized vein of left anterior condylar dural AVF.

### Main points

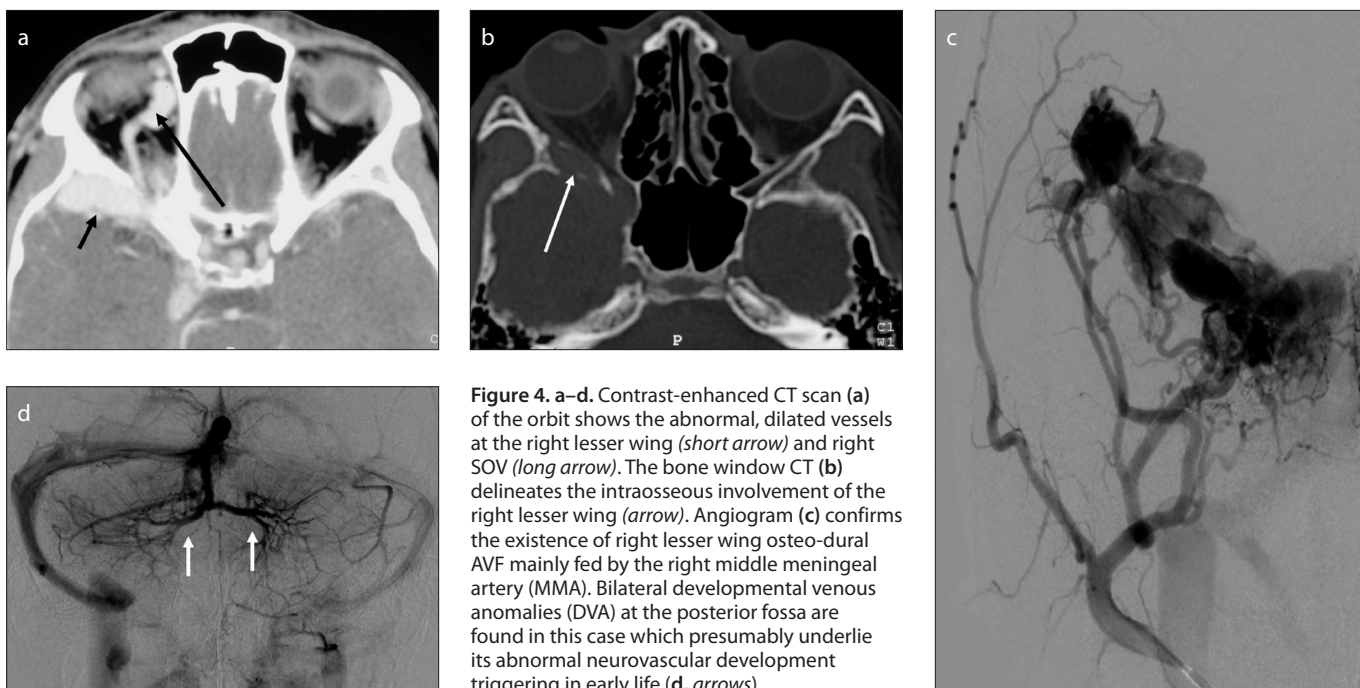
- The clinical presentation of the dural arteriovenous shunt mainly depends on the venous outflow.
- The venous outlet of the arteriovenous shunts is influenced by venous afferent-efferent patterns according to the venous anatomy of the central nervous system, as well as architectural disturbance, specifically, venous outflow obstruction.
- Infrequently, the shunts locating outside the cavernous sinus can mimic symptoms and radiography of cavernous arteriovenous shunts.



**Figure 2.** a–c. Axial T2-FLAIR image (a) and axial time-of-flight (TOF) magnetic resonance angiography (b) show the arterialized vein in left cavernous sinus and left superior ophthalmic vein (SOV) dilatation (a, b, arrow). The vertebral angiogram (c) reveals the vertebro-vertebral fistulae (c, circle). The shunt drains upward to the cavernous sinus and SOV (c, arrow).



**Figure 3.** a–c. Axial contrast-enhanced CT scan (a) shows right SOV dilatation (white and black arrows). Angiogram images (b, c) clearly demonstrate the right transverse sinus dural AVF (b, arrow) with evidence of venous reflux via the right SOV (c, arrow). Note that bilateral sigmoid sinuses are occluded.

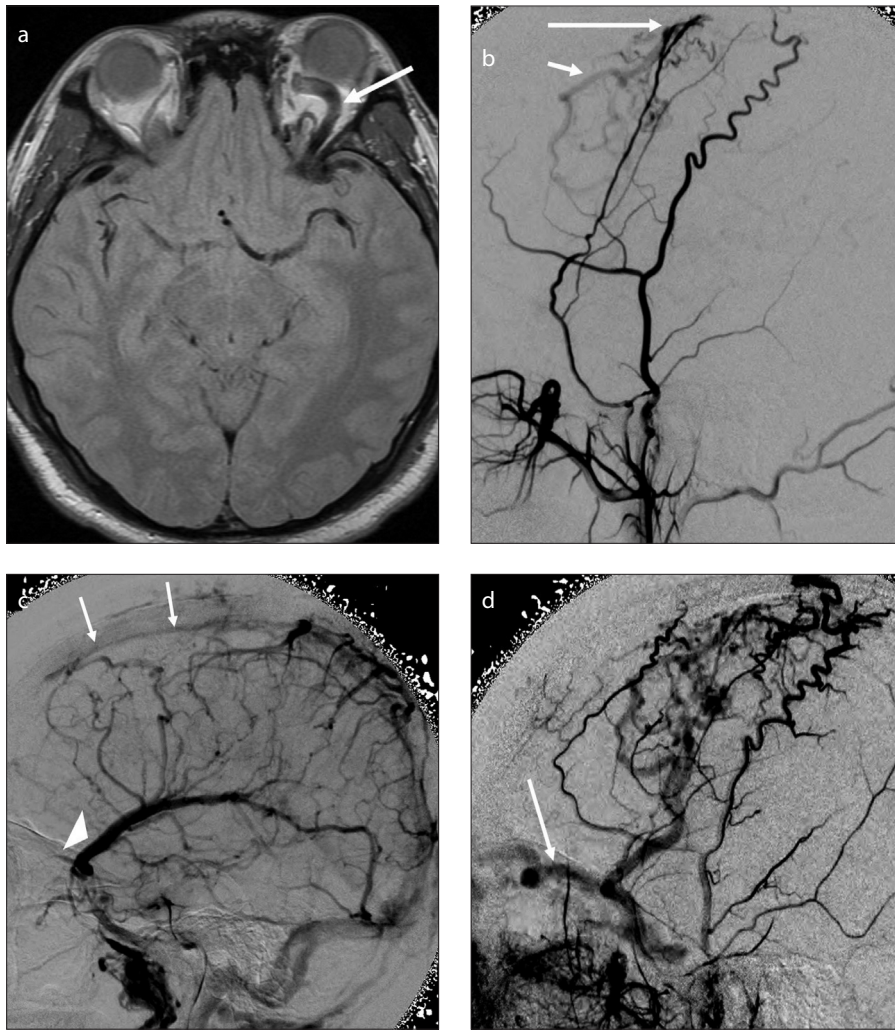


**Figure 4.** a–d. Contrast-enhanced CT scan (a) of the orbit shows the abnormal, dilated vessels at the right lesser wing (short arrow) and right SOV (long arrow). The bone window CT (b) delineates the intraosseous involvement of the right lesser wing (arrow). Angiogram (c) confirms the existence of right lesser wing osteo-dural AVF mainly fed by the right middle meningeal artery (MMA). Bilateral developmental venous anomalies (DVA) at the posterior fossa are found in this case which presumably underlie its abnormal neurovascular development triggering in early life (d, arrows).

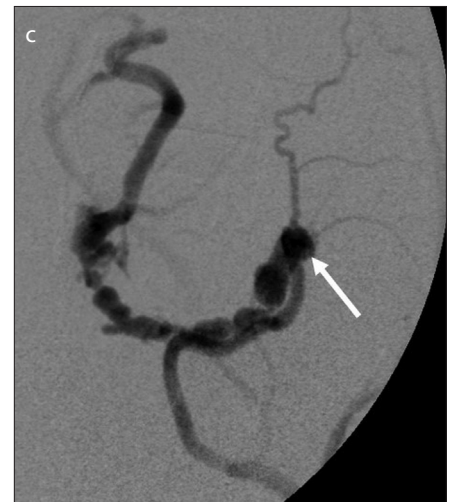
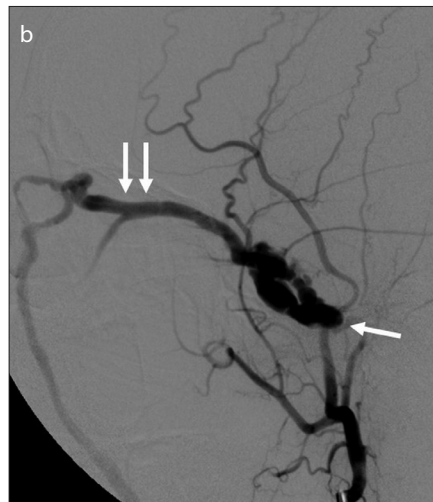
in other cases (2, 6). The venous efferent in transverse sigmoid sinus in our series also revealed downstream obstruction, which

propelled the arterialized vein toward the cavernous sinus and superior ophthalmic vein (SOV) (Fig. 3).

A young teenager who was diagnosed with lesser sphenoid wing dural AVF was successfully treated with transvenous coil em-



**Figure 5.** a–d. Axial proton density-weighted magnetic resonance image (a) shows the enlarged left SOV (arrow) as well as mild exophthalmos. External carotid angiogram (b) depicts the direct AV shunt from middle meningeal artery (long arrow) to the cortical veins (short arrow). Venous phase angiogram (c) shows small caliber of the anterior one-third to the middle two-thirds of the superior sagittal sinus (SSS). Normal variant hypoplasia of the anterior one-third of the SSS could be excluded in this case since the abnormality also involves the middle two-thirds of the SSS (arrows). The sylvian vein takes over the normal venous drainage of the brain (arrowhead). The arterialized vein drains downward and finally toward the SOV (d, arrow)

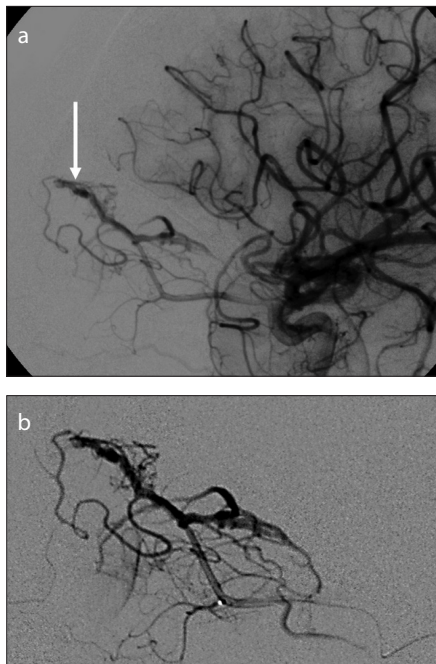


**Figure 6.** a–c. Axial CT scan of the brain (a) shows dilated left ophthalmic vein (arrow). The selective lateral (b) and anteroposterior (c) external carotid arteriography images show the middle meningeal arteriovenous fistulae (b, c, arrows). Its vein drains toward the ophthalmic vein (b, double arrows).

bolization (Fig. 4). The CT scan and angiography showed osteo-dural shunt involving the osseous structure and draining toward cavernous sinus and ophthalmic vein without cortical venous reflux. Shi ZS et al. (10) also demonstrated five cases of lesser sphenoid wing dural AVFs. One case had venous drainage into the superficial middle cerebral vein (SMCV). In two cases, the venous shunt emptied into the cavernous sinus and SOV, mimicking a clinical cavernous AV shunt. In their series, the greater sphenoid wing dural AVFs had more aggressive angiographic features according to SMCV reflux in four of six cases. They presumed that the close approximation of the lesser wing dural AVF to the cavernous sinus led to subsequent ocular symptoms and early medical attention, while the greater wing dural AVFs were more closely related to the paracavernous sinus which communicated with the SMCV, resulting in earlier cortical venous reflux or late ocular presentation (10).

Geibprasert et al. (11) have classified the sphenoid dural AVF and the basi-occipital AVF (including anterior condylar dural AVF) in the ventral epidural group, also known as the osteocartilaginous epidural group. They stated that these lesions are in direct contact with the osseous structures and normally have epidural outflow; therefore, they avoid cerebral venous reflux unless some factors precipitate venous outflow obstruction.

A direct dural artery to cortical vein AVF is the most aggressive type of dural AVF, and according to Cognard classification it bears a risk of hemorrhage of more than 50% (12). In our case, the shunt was located in the superior frontal convexity close to the supe-



**Figure 7. a, b.** Internal carotid angiogram (a) and superselective ophthalmic angiogram (b) show the intraorbital arteriovenous shunt (arrow).

rior sagittal sinus. Instead of draining toward superior sagittal sinus, the arterialized vein flowed downward toward the left cavernous sinus following with SOV mimicking a cavernous dural AVF. The venographic phase of the angiogram demonstrated thrombosis of superior sagittal sinus from the anterior one-third to the middle two-thirds, which presumably interfered with the venous return to superior sagittal sinus (Fig. 5).

Only two cases were clearly associated with head injury from motorcycle accident, which subsequently produced audible bruit and proptosis for several months thereafter. They were initially diagnosed as traumatic carotid cavernous fistulae and finally found to be middle meningeal AVF (Fig. 6). This

kind of AV shunt is relatively rare and has been reported in only 4% (18/446 patients) in an angiographic study of head injury patients (13). None of those in the series were reported to have clinical cavernous AV shunt.

We had one case of intraorbital AVF. It had arterial feeders associated with the ophthalmic artery, which was chosen to be the route of endovascular treatment (Fig. 7). This case was exceptionally rare, and has been found in few case reports in the literature (14).

The small number of patients and the retrospective nature are the main limitations of our study.

In conclusion, an AV shunt that mimics the cavernous AVF location is rare, with a prevalence of only 2.85% in our series. The clinical presentation of the patient depends mainly on venous outflow. The venous outlet of the AV shunts does not drain by chance, but is influenced by venous afferent-efferent patterns according to the venous anatomy of the central nervous system and skull base, as well as by architectural disturbance, specifically, obstruction of the venous outflow.

#### Conflict of interest disclosure

The authors declared no conflicts of interest.

#### References

1. Sedat J, Kominami S, Siriwimonmas S, Pongpech S, Suthipongchai S, Alvarez H. Extracavernous arteriovenous fistulae. Report of five cases. *Interv Neuroradiol* 1999; 5:235–243.
2. Turner RD, Gonugunta V, Kelly ME, Masaryk TJ, Fiorella DJ. Marginal sinus arteriovenous fistulas mimicking carotid cavernous fistulas: diagnostic and therapeutic considerations. *AJNR Am J Neuroradiol* 2007; 28:1915–1918. [\[Crossref\]](#)
3. Kobkitsuksakul C, Jiarakongmun P, Chanthanaphak E, Pongpech S. Radiographic evaluation and clinical implications of venous connections between dural arteriovenous fistula of the cavernous sinus and cerebellum and the pontomedullary venous system. *World Neurosurg* 2015; 84:1112–1126. [\[Crossref\]](#)

4. Alperin N, Lee SH, Sivaramakrishnan A, Hushek SG. Quantifying the effect of posture on intracranial physiology in humans by MRI flow studies. *J Magn Reson Imaging* 2005; 22:591–596. [\[Crossref\]](#)
5. Batson OV. The function of the vertebral veins and their role in the spread of metastases. *Ann Surg* 1940; 112:138–149. [\[Crossref\]](#)
6. McDougall CG, Halbach VV, Dowd CF, Higashida RT, Larsen DW, Hieshima GB. Dural arteriovenous fistulas of the marginal sinus. *AJNR Am J Neuroradiol* 1997; 18:1565–1572.
7. Pearce JM. The craniospinal venous system. *Eur Neurol* 2006; 56:136–138. [\[Crossref\]](#)
8. Tubbs RS, Ammar K, Liechty P, Wellons JC, 3rd, Blount JP, Salter EG, et al. The marginal sinus. *J Neurosurg* 2006; 104:429–431. [\[Crossref\]](#)
9. Zippel KC, Lillywhite HB, Mladinich CR. New vascular system in reptiles: anatomy and postural hemodynamics of the vertebral venous plexus in snakes. *J Morphol* 2001; 250:173–184. [\[Crossref\]](#)
10. Shi ZS, Ziegler J, Feng L, et al. Middle cranial fossa sphenoidal region dural arteriovenous fistulas: anatomic and treatment considerations. *AJNR Am J Neuroradiol* 2013; 34:373–380. [\[Crossref\]](#)
11. Geibprasert S, Pereira V, Krings T, et al. Dural arteriovenous shunts: a new classification of craniospinal epidural venous anatomical bases and clinical correlations. *Stroke* 2008; 39:2783–2794. [\[Crossref\]](#)
12. Cognard C, Gobin YP, Pierot L, et al. Cerebral dural arteriovenous fistulas: clinical and angiographic correlation with a revised classification of venous drainage. *Radiology* 1995; 194:671–680. [\[Crossref\]](#)
13. Freckmann N, Sartor K, Herrmann HD. Traumatic arteriovenous fistulae of the middle meningeal artery and neighbouring veins or dural sinuses. *Acta Neurochir (Wien)* 1981; 55:273–281. [\[Crossref\]](#)
14. Caragine LP, Jr., Halbach VV, Dowd CF, Higashida RT. Intraorbital arteriovenous fistulae of the ophthalmic veins treated by transvenous endovascular occlusion: technical case report. *Neurosurgery* 2006; 58:ONS-E170.