Marginal Sinus Arteriovenous Fistulas Mimicking Carotid Cavernous Fistulas: Diagnostic and Therapeutic Considerations

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SUMMARY: Marginal sinus fistulas (MSFs) are uncommon vascular anomalies. Occasionally, the dominant venous drainage is forced retrograde up the inferior petrosal sinus and into the cavernous sinus, causing chemosis, proptosis, and ocular hypertension, mimicking a carotid cavernous fistula. This atypical clinical presentation may lead to misdiagnosis and inappropriate hazardous treatment of an MSF. Identifying the site of the fistula and understanding the anatomy of the venous drainage are critical in providing appropriate, safe, and efficacious endovascular treatment.

The marginal sinus surrounds the foramen magnum, communicating anteriorly with the venous plexus of the clivus and posteriorly with the occipital sinus (if present). Correspondingly, dural fistulas of the marginal sinus typically drain via the sigmoid sinus–jugular bulb. However, in some cases, the jugular venous outflow becomes stenotic or thrombosed, forcing the venous drainage retrograde through the inferior petrosal sinus (IPS) and into the cavernous sinus (CS). Patients with this anatomy may present with clinical findings identical to those of a carotid cavernous fistula (CCF).

The clinical or angiographic misdiagnosis of a marginal sinus fistula (MSF) may lead to inappropriate or dangerous treatment. We review the clinical presentation, imaging findings, and endovascular treatment of 2 patients with MSFs, which presented clinically as CCFs.

Case Reports

Case 1
This 49-year-old man presented with an 18-month history of a pulsatile thrill in the right ear with subsequent decreased hearing and a 6-month history of progressive chemosis and proptosis without associated pain or oculoparesis. MR imaging revealed a dilated and arterialized IPS and superior ophthalmic vein (SOV). External carotid angiography (Fig 1) revealed a coalescence of abnormal arteries in the region of a marginal sinus recipient venous pouch with early venous filling and reflux into the IPS and CS. Selective catheterization of the ascending pharyngeal artery, right posterior auricular artery, and right internal carotid artery (Fig 2). The fistula was accessed and successfully coiled via a transjugular approach. The patient’s bruit resolved postprocedurally, and visual symptoms resolved in several days.

Discussion/Results
MSFs are uncommon vascular anomalies. McDougall et al reported on 290 fistulas, of which 14 were MSFs (4.8%). Most lesions were relatively benign, presenting with pulse synchronous bruit in 11 of 14 cases. However, 3 patients presented with hemorrhage (subarachnoid and intracerebellar) and/or neurologic symptoms (ataxia/tremor).

The anatomy of the venous outflow is the critical factor in patient presentation and treatment planning. In grade 1 lesions, the venous drainage is unrestricted and typically through the adjacent ipsilateral jugular vein. These patients present with a pulse synchronous bruit. Any change in the bruit, such as decreased intensity or resolution, may indicate an upgrading of the fistula. Grade 2 fistulas have a restriction or obstruction of venous drainage via the jugular bulb. In these cases, venous outflow is shunted retrograde through the IPS and into the CS. These patients present with orbital venous hypertension and a clinical syndrome more typical for a CCF. If the IPS drainage becomes stenotic or occludes, the fistula can be further upgraded. Grade 3 lesions demonstrate retrograde cortical venous drainage, and these, correspondingly, have the highest propensity to hemorrhage. Although patients with grade 1 and asymptomatic grade 2 may be observed in some cases, patients with grade 2 with ocular hypertension and all grade 3 lesions warrant intervention.

Treatment strategies previously reported initially centered on surgical ligation, with reports of endovascular strategies being ineffective. However, endovascular management of these and other dural arteriovenous fistulas (DAVFs) is both curative and safe.

In the present series, the diagnosis of MSF was complicated by a clinical presentation similar to that of a CCF. Misdiagno-
sis on the basis of clinical presentation and/or noninvasive imaging may lead to the inappropriate dismissal of an MSF as a benign indirect CCF. Furthermore, conventional angiographic investigation may not be performed, and the high-risk features (not infrequently associated with the MSF) may go undiagnosed. If the neurointerventionist misinterprets an MSF and embolizes the superior ophthalmic vein, CS, IPS, or jugular vein without ablating the recipient venous pouch, the fistula may upgrade with the elimination of these lower risk venous outflow pathways, potentially forcing the drainage into higher risk cortical venous pathways. In addition, these embolizations could block future endovascular access to the fistula site and thereby preclude definitive endovascular treatment targeted at occluding the marginal sinus recipient pouch.

As with all DAVFs, the accurate identification and subsequent endovascular embolization or surgical ligation of the proximal recipient vein (in this case the marginal sinus pouch) is required for safe and efficacious therapy. This is best achieved by selective transvenous catheterization of the fistula pouch with subsequent coil embolization (case 2). As with other DAVFs, this is best facilitated by performing roadmaps or by using an “overlay” fluoro-fade technique in an imaging projection that best separates the marginal sinus pouch to be catheterized from the normal anatomic venous drainage. A more selective roadmap of the pouch is usually best achieved with a 4F or 5F diagnostic catheter positioned within a branch of the external carotid artery, which provides a dominant supply to the fistula. This more selective roadmap reduces the degree of obscuration of the fistula site, often produced with a more proximal external or common carotid injection. Transvenous access to the fistula pouch can usually be achieved either directly from the ipsilateral internal jugular (IJ) through the contralateral IPS-circular sinus-ipsilateral IPS approach or retrograde through an SOV-CS-IPS approach. Liquid embolic agents injected via transarterial access or retrograde through transvenous access can be useful when total transvenous coil occlusion of the fistula has not been achieved and access into the fistula has been lost or becomes tenuous. If a transvenous Onyx injection is performed, this must be done with the microcatheter positioned within the fistulous pouch and is most effective when the outflow has been slowed to some extent by partial coil embolization.

**Conclusion**

MSFs are uncommon vascular anomalies whose symptoms can mimic CCFs. Correctly identifying the location and anat-
omy of the recipient pouch is critical in providing appropriate, safe, and efficacious treatment. This diagnosis should be considered whenever fistulas in the region of the jugular vein are identified. Selective external carotid branch angiography with oblique views can be useful in separating the jugular bulb laterally from the marginal sinus and can aid in the identification, catheterization, and embolization of MSFs. As with all dural fistulas, cure is best achieved by a treatment strategy targeted to achieve complete occlusion of the recipient pouch.

References