Low-Flow Direct Carotid-Cavernous Fistula Caused by Rupture of an Intracavernous Carotid Aneurysm

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Summary

Direct carotid-cavernous fistulas (CCFs) secondary to a ruptured intracavernous carotid aneurysm (ICCA) are usually high-flow lesions. On very rare occasions, a ruptured ICCA may present as a low-flow CCF, which poses a diagnostic and therapeutic dilemma whether the aneurysm and the observed fistula are causally related. Herein, we describe a rare case in which a ruptured ICCA resulted in a low-flow CCF. We demonstrated our approach to clarify this clinical scenario, and also propose a possible pathomechanism to explain the existence of low-flow direct CCF.

Introduction

Intracavernous carotid aneurysms (ICCA) account for 2-9% of intracranial aneurysms. They are usually regarded as benign lesions because they rarely cause a subarachnoid hemorrhage¹. Clinically they may present either with cranial nerve palsies due to mass effect on the lateral wall of the cavernous sinus (CS), or with aneurysm rupture resulting in a direct carotid-cavernous fistula (CCF)². Aneurysmal CCFs are almost invariably associated with a high-flow arteriovenous shunt². Ruptured ICCAs, on very rare occasions, may present as low-flow CCFs, and the reasons this occurs are not clear³. In this scenario, the question whether the fistula arises from the aneurysm may become a clinical dilemma. Herein, we describe a case in

which our endovascular approach proves the true causal relationship between a ruptured ICCA and a low-flow CCF and we propose a possible pathomechanism to explain this rare finding.

Case Report

A 61-year-old woman presented with an acute onset of severe headache, followed by left painful ophthalmoplegia for five days. There was mild chemosis and exophthalmos, but no orbital bruit. Computed tomography (CT) scan of the brain did not show subarachnoid hemorrhage. CT angiogram (CTA) showed an 8mm lateral projecting broad-necked aneurysm arising from the left intracavernous internal carotid artery (ICA) (Figure 1A,B). There was no significant asymmetric filling of the left CS, or enlargement of the left superior ophthalmic vein (SOV). Clinical considerations included an aneurysmal CCF, a CS thrombosis, or a sudden enlargement of the aneurysm resulting in cranial nerve compression. A cerebral angiogram for diagnosis and potential endovascular treatment was arranged.

The anteroposterior, lateral and three-dimensional rotational angiograms revealed an aneurysm arising from the horizontal cavernous segment of the left ICA (Figure 1C,D). The distal intracranial circulation showed normal opacification and contrast transit times. Early contrast filling of a cavernous venous pouch was seen next to the aneurysm, with a linear

Figure 1 Axial (A) and coronal (B) CTA shows an aneurysm arising from the horizontal segment of the left intracavernous ICA (star sign). The aneurysm wall was highlighted as a filling defect (arrowhead) between the aneurysmal pouch (solid arrow) and an adjacent vascular pouch (dotted arrow). Four venous compartments (outlined by dashed lines) were labeled for anatomical correlation on the contralateral CS (B). Note that the CS is not clearly opacified, and the aneurysm mainly occupies the lateral compartment, which is the narrowest venous space of the CS. At this time point, we did not consider a CCF to be present. Injection of the left ICA in anteroposterior (C) and lateral (D) views demonstrates a low-flow CCF which drained into the inferior petrosal sinus (double arrows) and superior ophthalmic vein (dashed arrow). The lesion morphology in the anteroposterior view is similar to that in coronal CTA [same figure labels as in (B)]. Note the slow appearance of the CS fillings and normal visualization of the ipsilateral hemispheric arteries in the early arterial phase.

filling defect separating both structures (Figure 1C). There was also early filling of the ipsilateral CS, SOV, and inferior petrosal sinus (IPS) in mid-to-late arterial phase, with no associated venous enlargement and with a rather slow flow pattern atypical for a direct CCF (Figure

1D). Because of these flow dynamics, we were in equipoise as to whether an indirect cavernous dural arteriovenous fistula (DAVF) supplied by the inferolateral trunk (ILT) was present with the “aneurysm pouch” rather representing a venous outpouching of the CS that

was inseparable from the ICA due to volume averaging on rotational angiograms, or whether the aneurysm pouch presented an arterial aneurysm that had ruptured into the cavernous sinus.

Bilateral external carotid arteries (ECAs) and contralateral ICA angiograms were all negative. While treatment was deemed necessary given the clinical presentation in the presence of an arteriovenous shunt with presumed compressive effects on the cranial nerves, the treatment approach for the two possible entities (indirect CCF versus direct CCF) would have to be different: in case of an indirect CCF we would typically choose a transvenous approach to occlude the primary venous pouch with thrombogenic coils, while in a direct CCF the approach should be transarterial. In order to determine the nature of the lesion, we advanced through a 6-French guiding catheter placed in the distal left cervical ICA a 0.014" microcatheter into the pouch and performed a superselective injection which demonstrated slow filling of the posterosuperior and antero-inferior compartments of the CS (Figure 2A). To verify that this microcatheter had reached the venous compartment via the aneurysm pouch rather than through the ILT, we then advanced a second 0.014" microcatheter through a more inferior route into the pouch, so access to both the venous and arterial components of the fistula was secured. Intra-aneurysmal microcatheter injections demonstrated the direct communication with the venous pouch (Figure 2B). As the ILT is too small to accommodate two microcatheters (Figure 2E), we were sure that there was no contribution to the CCF via ILT. Angiographic findings were therefore consistent with a true aneurysmal CCF which had ruptured into a CS venous compartment (i.e. Barrow classification type A) with restricted outflow which would explain the low-flow nature of the shunt. Retrospectively, we were able to depict this lesion configuration also on coronal CTA (Figure 1B).

Through both microcatheters, five bare platinum detachable coils were placed in both the venous and arterial sides of the lesion (Figure 2C,D), resulting in complete closure of the shunt (Figure 2F). We deliberately packed the aneurysm sac rather loosely in order not to cause additional mass effect on the CS cranial nerves. The ocular symptoms partially resolved during the week after the procedure. Nearly two months after the procedure, the patient

showed a significant recovery and maintained clinical improvements. Follow-up imaging demonstrated stable occlusion of the fistula.

Discussion

Based on the observed flow rates, CCFs can be divided into high or low-flow. CCFs are regarded as low-flow if the CS opacification is slower than the normal filling of ipsilateral hemispheric arteries^{3,5}. By contrast, CCFs with equal or dominant flow toward the fistula, as compared to that of the hemispheric arteries, are defined as high-flow fistulas^{3,5}. Direct CCFs resulting from spontaneous rupture of ICCAs are almost invariably high-flow mainly because the ruptured aneurysm creates a direct single-hole communication between the high-pressure ICA and the low-pressure CS system. As a result, the sudden rise in intracavernous pressure can lead to variable signs of typical arteriovenous fistulas, such as ocular venous congestions or cortical venous reflux (CVR), depending on the CS outlet involved^{6,7}. The high-flow nature of the shunt also presents typically with an audible bruit, chemosis, and a bulging eye. Ruptured ICCAs may also compress the cranial nerves (III, IV, V1, V2, or VI nerve) which travel through the lateral wall or within the CS^{1,7}.

Low-flow aneurysmal CCFs are rarely encountered. From the literature, we only identified three cases with documented direct low-flow CCF^{3,5}. Based on these limited cases, we found that they had different clinical and angiographic manifestations. In van Rooij et al.'s series of 11 aneurysmal CCFs, for example, two cases were associated with low-flow fistulas, and both fistulas closed spontaneously before treatment³. Spontaneous regression of the high-flow aneurysmal CCFs, in contrast, was rare². In addition, high-flow CCFs refluxed into cortical veins more commonly, but none of the cases with low-flow aneurysmal CCFs (including ours) demonstrated CVR^{3,8}. Though these differences were not statistically significant given the small sample size, one may infer that low-flow variants of the direct CCFs will present with different clinical manifestations and require different treatment planning.

Since ruptured ICCA rarely cause low-flow CCFs, one should exclude an indirect CCF which may present with a similar clinical picture. Indirect CCFs, a type of cavernous DAVFs, are supplied by dural branches of ECA and/or ICA.

Figure 2 A) Four consecutive images (1-4) of a superselective venogram (lateral view) from the venous pouch (arrowhead) demonstrate slow filling of the posterosuperior (solid arrow) and antero-inferior (dotted arrow) compartments of the CS, and subsequent drainage toward the IPS (double solid arrows) and SOV (double dotted arrows) respectively. B) Intra-aneurysmal (solid arrow) microinjection from the second microcatheter (oblique view) directly opacified the venous sac (dotted arrow) and highlighted the aneurysmal wall (arrowhead) in between. C-E) Detachable coils were placed in the venous sac (dotted arrow) and the aneurysm (solid arrow) through two microcatheters. Note that both microcatheters (single and double arrowheads) are spaced apart but enter the aneurysm through the same opening. F) Left ICA angiogram (lateral view) after coil embolization shows complete closure of the fistula.

Feeders from the ECA can be easily evaluated by standard ECA angiograms. Given the low flow and location of the fistula in our case, it was possible that the lesion may have corresponded to a cavernous DAVF fed by the ILT, and the aneurysm was actually the first venous pouch of the fistulous drainage. We excluded this possibility for three different reasons. First, the spherical shape of the first vascular pouch (solid arrow in Figure 1B,C), together with the filling defect outlined by the second venous pouch (dotted arrow in Figure 1B,C), indicates that the first pouch represents a true arterial aneurysm. Second, the ILT is such a small vessel that, even in the presence of a fistula, it will not accommodate two 0.014" microcatheters (Figure 2E). Finally, intra-aneurysmal angiograms demonstrated a direct connection to the adjacent venous pouch, which in turn drained into the CS (Figure 2B). Based on these observations, we are

able to prove that the clinical scenario we encountered is a true ruptured ICCA presenting as a low-flow CCF of Barrow type A⁴.

Even though this entity has been previously described, a pathomechanism to explain a direct low-flow CCF has not been addressed before^{2,3}. We believe that factors such as the shunt volume, orientation of the aneurysm, and the size and outflow of the involved venous space may explain this phenomenon. The CS, based on the relationship to the intracavernous ICA, can be divided into four compartments: antero-inferior, posterosuperior, medial, and lateral⁹. Normally, all venous spaces communicate with each other, and each compartment has its own outlet away from the CS. Among the four described venous spaces, the posterosuperior compartment is the largest one and the lateral compartment is the smallest⁹. The lateral compartment is so narrow that the VI nerve becomes adhered between the

ICA and the lateral wall of the sinus wall, and rarely the CS extends lateral to the VI nerve. Therefore, unlike other compartments, the connections of the lateral compartment with the rest of the CS are narrowed.

The concept of venous compartments is clinically relevant as has been emphasized in targeted compartmental embolization of dural CCFs¹¹. The drainage routes for a direct CCF can also become limited when ICCA rupture into a compartmentalized part of the CS^{7,12}. In our case, we can infer that owing to the lateral projection of the aneurysm its dome was confined to the lateral compartment of the CS. As the aneurysm ruptures, the blood shunts into the first encountered venous space within the lateral compartment. However, the inherent small size and limited outflow of this compartment, together with the mass effect from the aneurysm itself, can block the lateral compartment's outlets. Further restriction to the fistula outflow may be seen if thrombosis of the some other venous spaces occurs. As a result, the blood from the venous pouch cannot drain freely to other compartments of the CS (Figure 2A). This explains why in our case CS filling was slower than that expected for an aneurysmal CCF, and why part of the clinical presentations was more similar to that described for indirect CCFs.

There are reports of spontaneous closure of direct low-flow CCFs, but in our case the acute cranial nerve palsy prompted urgent treatment.

The transarterial route is preferred for endovascular treatment of direct CCFs^{6,13}. The transvenous approach is considered an alternative when the transarterial approach fails or in cases with fragile arterial walls such as Ehlers-Danlos syndrome^{2,14}. Both detachable balloons and coils can be used to occlude the shunt^{2,3,6,13}. When the flow is slower, detachable coils are more suitable to close the shunt². By targeting both the arterial and venous sides of the shunt with coils, the fistula can be closed and the normal drainage of other CS compartments can be preserved. Pure transarterial coiling will necessitate dense packing of the aneurysm which may result in increased mass effect on the cranial nerves and worsening of the cranial nerve palsy. Endoluminal reconstruction using stent grafts or flow diversion may represent an alternative treatment, but long-term results are still pending for these treatment methods^{15,16}.

In conclusion, we describe an unusual case of a direct low-flow CCF secondary to a ruptured ICCA. The management of this type of lesions requires an understanding of the spatial and hemodynamic relationship between a ruptured ICCA and the CS compartments involved.

We hypothesize that the low flow observed in our case is attributable to a combination of aneurysmal (lateral projection) and venous (narrow lateral CS compartment and limited connections to other CS spaces) factors.

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