CASE REPORT

Primary intraventricular haemorrhage due to rupture of giant varix of the basal vein of Rosenthal in a patient with long-standing direct CCF: angiographic features and treatment considerations

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SUMMARY

Direct carotid cavernous fistulae (CCF) are often detected early and treated promptly resulting in a paucity of literature regarding its long-term evolution. We present a case of high flow post-traumatic direct CCF that was neglected for over 6 years and presented with a rare manifestation of primary intraventricular haemorrhage. Occlusions of the primary venous outlets likely resulted in engorgement of the deep venous system. The segmental anatomy of the shunting basal vein is critical to the clinical presentation and may range from basal ganglia or brainstem oedema/infarctions to uniquely, as in our case, isolated intraventricular haemorrhage secondary to variceal rupture. Treatment in such chronic cases requires a consideration of cerebral hyperperfusion syndrome necessitating deconstructive techniques with subsequent anticoagulation to avoid accelerated thrombosis of the venous varices.

BACKGROUND

Direct carotid cavernous fistulae (CCF) usually present with myriad clinical manifestations, of which distressing ocular symptoms often lead to an early diagnosis and prompt treatment. Intracranial haemorrhage is an uncommon but devastating complication of direct CCF occurring in only 4.5%–10.2% in large case series.^{1 2} We describe a previously unreported presentation of isolated primary intraventricular haemorrhage (IVH) due to the presence of a large venous varix in a long-standing case of direct CCF. We detail the clinical course and imaging findings peculiar to this presentation as well as the technical challenges in treating these cases.

CASE PRESENTATION

A male aged 28 years with cutaneous manifestations of type I neurofibromatosis presented with a history of sudden-onset headache and loss of consciousness. Six years prior to his current presentation, the patient had significant craniofacial trauma with minimal subarachnoid haemorrhage for which he was conservatively managed. A few months later, he developed progressive diminution of vision in the right eye with mild proptosis, followed by proptosis and pain in the left eye. He remained relatively asymptomatic

and medical attention was declined until the onset of the current event. At presentation, his GCS (Glasgow Coma Scale) was 9/15 and he had bilateral proptosis and optic atrophy on funduscopy. Audible bruit was heard over the orbital and temporal regions bilaterally.

INVESTIGATIONS

At imaging, CT revealed acute IVH with associated obstructive hydrocephalus ([figure 1](#)). CT angiography revealed multiple dilated vascular channels in both the paracavernous and suprasellar regions. Invasive cerebral angiography revealed a transection of the anterior genu of cavernous segment of the left internal carotid artery (ICA) with complete high flow shunting into an aneurysmally dilated left cavernous sinus. Venous egress was noted into right superior ophthalmic vein (SOV) across the intercavernous sinus and through left uncal vein into the left basal vein of Rosenthal (BVR) ([figure 2](#)). The mesencephalic segment of the BVR was absent, with resultant venous reflux further into the frontobasal veins anteriorly and through the peduncular vein into the petrosal vein posteriorly. A partially thrombosed venous varix of the striate and peduncular segments of the BVR was noted protruding into the third ventricle and obstructing the foramina of Monroe ([figure 1](#)). Good cross-circulation was maintained across the anterior communicating artery without significant venous delay.

TREATMENT

The patient underwent endovascular trapping and parent artery occlusion of the left internal carotid artery. Two microcatheters were used, the first to initially form a stable coil mass within the giant cavernous sinus so as to prevent coil migration followed by subsequently coiling of the cavernous ICA just proximal to the rent. The second microcatheter was used to coil the supraclinoid ICA distal to the rent to achieve near complete obliteration of the fistula while preserving antegrade flow into the left cerebral hemisphere from across the posterior communicating artery ([figure 3](#)). Significant venous stasis was noted within the dilated cortical veins and varices after embolisation.

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Figure 1 Axial non-contrast CT (A) and T2-weighted MRI (B) show intraventricular haemorrhage with a partially thrombosed venous varix causing obstructive hydrocephalus. CT angiography (C, D) shows opacification of the cavernous venous sacs which communicate via the uncus vein (solid arrow) with a aneurysmally dilated mesencephalic segment of the left basal vein of Rosenthal (arrowhead). The rest of the basal vein is dilated anteriorly until the confluence of the frontobasal veins (*).

OUTCOME AND FOLLOW-UP

In view of the risk of inducing accelerated thrombosis of the cortical veins or giant varices, the patient was heparinised and bridged over to oral anticoagulation. He had an uneventful postoperative period with significant improvement of neurological status at discharge. He remained clinically stable at the last clinical follow-up at 6 months and further anticoagulation was discontinued.

DISCUSSION

Direct CCFs are high flow arteriovenous shunts between the cavernous ICA and the adjacent cavernous sinus, with trauma being the the most common aetiology.³ Most of these fistulae are treated early due to prominent ophthalmic features and hence the long-term evolution of CCF has not been clearly delineated. The protean clinical manifestations and the aggressiveness of CCF are dependent on the degree and routes of venous egress.⁴ The direction of venous egress evolves over time with the development of venous occlusions and venous rerouting as a result of progressive venous restrictive angiopathic changes and gradual thrombosis. Obliteration of primary venous outlets such as SOV or the petrosal sinuses would result in preferential venous

shunting into the cortical veins, which enlarge over a period of time to form giant venous sacculations.

Haemorrhage is an uncommon but ominous complication of CCF occurring in 4.5%–10.2% of patients.^{1,2} Two possible mechanisms exist to explain the occurrence of haemorrhage at various sites. Intraparenchymal haemorrhages occur secondary to cerebral venous hypertension as a result of cortical venous drainage.⁵ On the contrary, rupture of venous varices may result in extra-axial haemorrhages into either the subarachnoid space, or epistaxis if erosion or fracture of the sphenoid sinus walls had occurred. Halbach *et al* suggested that although the presence of venous varices was not specific for the risk of subarachnoid haemorrhage (SAH), the high fatality rate associated with the usually massive amounts of SAH mandated urgent treatment (100% of cases with SAH died before treatment in their series).² To our knowledge, a case of direct CCF manifesting as primary IVH is unique and has never been reported in the past. This likely relates to the anatomical variation of basal vein of Rosenthal in our case.

The three components of the basal vein embryologically develop from three different veins: the anterior striate segment from the deep telencephalic vein, the middle peduncular

Figure 2 Volume rendered CT angiography (A) and diagrammatic representation (B) show the spatial relationships of the cavernous sinus (1=CS), uncus vein (2=UV), the partially thrombosed varix of the striate and peduncular segments of the left basal vein of the Rosenthal (3=BVR) and the peduncular vein (4=PV). Note the lack of communication between the left basal vein and the vein of Galen (5=VOG) in contrast to the normal appearing contralateral basal vein (6=contralateral BVR).

Figure 3 Selective left internal carotid angiography (A) shows the site of rent with rapid filling (B, C) of the cavernous sinuses, the connecting uncal vein (black arrows) and the dilated basal vein of Rosenthal (*) along with reflux into the peduncular vein (white arrow). Vertebral angiography (D) shows steal across the posterior communicating artery while the contralateral internal carotid angiogram (E) showed complete collateralisation of left cerebral hemispheric flow with some steal into the fistula. Post-trapping, the ipsilateral internal carotid (F) and vertebral (G) angiograms show the coil mass within the cavernous sinus sac and cessation of shunting at the proximal and distal aspects of the ICA transection. Right internal carotid angiography (H), showed improved collateral flow into the left hemisphere with cessation of steal into the fistula.

segment from the ventral diencephalic vein and the posterior mesencephalic segment from the dorsal diencephalic vein.⁶ Any of these segments may be hypoplastic resulting in variations of drainage. If only the anterior striate segment is present and decompresses the cavernous sinus via the uncal vein with reflux into striate veins, extensive basal ganglia oedema may result.⁷ When the mesencephalic segment alone is absent, the lateral mesencephalic vein may act as an anastomosis between the peduncular segment and the posterior fossa veins resulting in brainstem oedema or haemorrhagic infarction.^{8,9} In our case, the draining basal vein lacked a mesencephalic segment, and the uncal vein directly emptied into its anterior segments with no communication with striate veins and only minor reflux into the frontobasal veins. The absence of the Galenic drainage or any significant communication to the infratentorial veins in effect made the basal vein a cul-de-sac. This lack of decompression and resultant long-standing engorgement resulted in a giant varix that compressed the brain parenchyma and ruptured into the

adjacent third ventricle. Indeed, this aneurysmal dilatation was noted at the site of entry of the uncal vein into BVR, which may have been secondary to exposure to a constant pulsating high flow jet. The basal vein variation was thus critical to the presentation of intraventricular haemorrhage in our case.

The chronicity of CCF and the presence of large venous varices in our patient presented its own set of challenges with regard to treatment. The occlusion of all accessible venous routes necessitated that we use a transarterial route. A complete occlusion of venous sac is nearly impossible considering its size and the cost incurred. Other reconstructive techniques such as stenting or flow diversion were considered high risk in view of recent haemorrhage. Additionally, long-term arterial steal may have induced a chronically hypoperfused state in the ipsilateral cerebral hemisphere resulting in a risk of cerebral hyperperfusion syndrome with reconstructive techniques.¹⁰ Hence, we opted for a parent artery occlusion and loose coiling of the venous sac to encourage thrombosis. In

view of the large sac size and high flow, two microcatheters were used to prevent coil migration and form a stable coil mass within the sac. Subsequently, the proximal and distal internal carotid ends of the fistula were occluded with coils, effectively trapping the fistula. Postinterventional stasis of flow in the cortical veins and venous sacs could be a harbinger of accelerated thrombosis with propagation leading to a risk of venous infarction or increased mass effect.^{11–13} We hence prophylactically anticoagulated the patient postoperatively.

Many factors may have led to the delay in diagnosis. The lack of prominent orbital symptoms during his initial presentation may have led to the CCF being neglected for a period of 6 years.⁵ A larger rent was also found to be statistically correlated with a longer time to detection in a series by Chi *et al.*¹⁴ Additionally, it is possible that the associated sphenoid wing aplasia allowed unimpeded growth of the venous sacs increasing their capacitance. The longest standing case of direct CCF was reported by Workman *et al.*, in which they described an adult patient with childhood traumatic CCF treated by surgical trapping that recanalised and presented with intracranial haemorrhage 26 years later.¹⁵ Thus, our case provides a window into the untreated natural history of a disease that is rarely neglected.

CONCLUSION

This report highlights an unusual presentation of intraventricular haemorrhage in a long-standing case of high flow direct

CCF with deep venous drainage. Variations in the anatomy of the draining basal vein influence the symptomatology of CCF and its chronicity impacts choices in treatment.

Contributors All authors listed meet authorship criteria, and all authors certify that they have participated sufficiently in the work to take public responsibility for the content, including participation in the concept, design, analysis, writing, and revision of the manuscript. Furthermore, each author certifies that this material or similar material has not been and will not be submitted to or published in any other publication before its appearance in the Journal of Neurointerventional Surgery.

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Learning points

- Progressive venous occlusions occurring in long-standing carotid cavernous fistulae (CCF) may lead to preferential shunting into the deep venous system. Variations in the segmental anatomy of the draining basal vein result in a spectrum of predictable clinical manifestations.
- Subsequent engorgement may lead to the development of aneurysmally dilated venous sacs that result in subarachnoid or intraventricular haemorrhage in contradistinction to the more common intraparenchymal haemorrhage caused by cortical venous hypertension.
- Chronic CCF merits consideration of deconstructive techniques in view of the risk of cerebral hyperperfusion syndrome and may necessitate postoperative anticoagulation to prevent venous infarction or increased mass effect secondary to accelerated thrombosis of the cortical veins or varices.

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