CASE REPORT

Successfully treated symptomatic fusiform basilar artery aneurysm in a patient with hindbrain malformation via inverted Y-stenting

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SUMMARY

A double overlapping reverse Y-stent approach to creating flow diversion using traditional open-cell stent technology was evaluated as a treatment option symptomatic fusiform basilar artery aneurysms. A 36-year-old man with a complex hindbrain malformation presented with acute ocular dysmotility due to a rapidly enlarging fusiform basilar artery aneurysm. The aneurysm was treated by insertion of two stents into the vertebrobasilar system in an inverted Y-configuration from the basilar tip to the V4 segments of the bilateral vertebral arteries, essentially creating flow diversion without using a dedicated flow diversion device. This resulted in immediate symptomatic improvement. The stents remained patent and the aneurysm was obliterated at 6 months follow-up. Furthermore, the patient remained free of associated symptoms at 10 months follow-up. Thus, the double stenting technique can be used instead of a flow diversion device to effectively create flow diversion, promote aneurysm sac thrombosis, and lead to resolution of symptoms in large fusiform basilar artery aneurysms.

BACKGROUND

Fusiform aneurysms of the posterior circulation are challenging lesions for both endovascular and open surgical repair. Among various treatment strategies, the implantation of flow-diverting devices is currently preferred for the management of fusiform basilar artery aneurysms.¹ We present a symptomatic partially thrombosed mid-basilar artery aneurysm that was treated with an overlapping reverse Y-configuration stenting technique in a patient with rhombencephalosynapsis.

CASE PRESENTATION

A 36-year-old man with a history of Chiari II malformation and multiple surgeries including cord untethering and ventricular shunting presented with episodes of diplopia and ocular dysmotility.

INVESTIGATIONS

A brain MRI obtained before the visual deficits demonstrated a slightly ectatic mid-basilar artery, dysgenesis of the corpus callosum, rhombencephalosynapsis, brainstem hypoplasia, a small posterior fossa, and cerebellar tonsils extending 12 mm below the foramen magnum (figure 1). A follow-up brain MRI and MR angiography obtained 8 months later when the patient developed diplopia and ocular dysmotility demonstrated marked interval enlargement of the fusiform basilar artery aneurysm (figure 2A)

which, on the coronal maximum intensity projection image (figure 2B), measured approximately 23 mm in length and 7 mm in diameter.

TREATMENT

Due to the patient's symptoms and rapid enlargement of the aneurysm, it was decided to proceed with endovascular intervention. A double stent with reverse Y-configuration exclusion of the basilar artery fusiform aneurysm was performed with a reversed Y-stenting technique using one open-cell design stent (Neuroform-EZ; Stryker, Kalamazoo, Michigan, USA) and one closed-cell design stent (Enterprise; Cordis, Miami, Florida, USA). Using a 014 Synchro microwire (Stryker), an Excelsior XT 27 microcatheter (Stryker) was advanced to the right posterior cerebral artery, P1–P2 segment junction. Next, a Neuroform EZ-3 3.5 mm×30 mm was delivered and deployed from the upper basilar artery to just below the right superior cerebellar artery origin to the distal right vertebral artery, about 5 mm below the vertebrobasilar junction. A Prowler Plus microcatheter (Codman, Raynham, Massachusetts) was then used to deliver a 4.5 mm×37 mm Enterprise stent to cover from the upper basilar artery to the distal left vertebral artery.

OUTCOME AND FOLLOW-UP

Immediate post-procedure digital subtraction angiography demonstrated patency of the stents and

Figure 1 Baseline axial T2-weighted MRI showing a mildly ectatic basilar artery and rhombencephalosynapsis.

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Figure 2 (A) Axial T2-weighted MRI obtained 8 months later at the time of symptom development shows a dysmorphic cerebellum with absence of the vermis and continuity of the cerebellar hemispheres across the midline, compatible rhombencephalosynapsis, hypoplasia of the pons, and a wide basilar artery flow void (arrow). (B) Corresponding three-dimensional maximum intensity projection image time-of-flight MR angiogram showing a long segment fusiform basilar artery aneurysm (arrow).

residual filling of the basilar artery aneurysm (figure 3), albeit with significant flow reduction and stagnation of intra-aneurysmal flow. The patient tolerated the procedure well and there was immediate symptomatic improvement of the ocular dysmotility.

Follow-up CT angiography obtained 6 months after the procedure demonstrated essentially complete obliteration of the aneurysm sac without evidence of in-stent stenosis, as visible on axial and three-dimensional volume-rendered images (figure 4A, B). Furthermore, the patient remained free of associated symptoms at 10 months follow-up.

DISCUSSION

Rhombencephalosynapsis is a rare congenital malformation of the cerebellum that results from an embryologic insult between days 28 and 41 of gestation. It is postulated to be a primary failure of vermian differentiation^{2 3} and consists of fusion of the cerebellar hemispheres, dentate nuclei, and superior cerebellar peduncles, and hypogenesis/agenesis of the vermis. In addition to cerebellar dysmorphism, the condition is associated with other neurologic findings including aqueductal stenosis

leading to congenital hydrocephalus, ventriculomegaly, dysgenesis of the corpus callosum, and various abnormalities of the thalamus, septum pellucidum, temporal lobes, olivary nuclei, optic chiasm, posterior pituitary, and hippocampus.^{2 3} Although rhombencephalosynapsis has also been associated with ophthalmologic abnormalities,² our patient did not exhibit ocular dysfunction early in life and only became symptomatic as the basilar dolichoectasia transformed into a fusiform aneurysm, rather than from the underlying hindbrain malformation itself. However, we speculate that the patient's complex hindbrain malformation with associated vertebrobasilar dolichoectasia may have predisposed to the formation of the basilar artery fusiform aneurysm.

Fusiform basilar artery aneurysms are rare, but are associated with a high morbidity rate, 60% experiencing new symptoms without treatment, and a mortality rate reported to be as high as 60%.⁴ Large fusiform aneurysms without a well-defined neck are among the most difficult type of aneurysms to treat without parent vessel occlusion. However, because patency of the many perforating arteries supplying the brainstem must be maintained, frank occlusion of basilar artery aneurysms cannot be undertaken and alternative methods must be employed.⁵ Numerous endovascular techniques have been attempted with varying success. Traditional treatments that have been used include parent vessel occlusion with surgical ligation or coil occlusion, surgical trapping with or without bypass, clip reconstruction, and stenting with or without coiling, or the newer technology of flow diversion.⁶

Flow diversion devices have had reported success primarily in the anterior circulation. However, caution has been advised for use in the posterior circulation, mainly due to concerns about multiple perforating branches which supply crucial brainstem structures.¹ Conventional open-cell stents have a metal surface area coverage of 10% compared with 30–35% in flow diversion devices such as the Pipeline Embolization Device.⁴ The successful treatment of posterior circulation fusiform aneurysms has been described using flow diversion devices.¹ However, small and tortuous distal vertebral arteries and a rapidly enlarging aneurysm with adjacent brain edema suggested fragility of the aneurysmal wall and mitigated against the ability to safely deliver and deploy a flow diversion device without generating excessive stress against the aneurysm wall. In addition, stenting has only been shown to be a potential alternative treatment option for basilar artery dissection and dissecting aneurysms.⁷ It was therefore decided to use two overlapping non-flow diversion stents to induce flow diversion.

Figure 3 Three-dimensional rotational digital subtraction angiography image obtained at the completion of the procedure showing the presence of the stents in an inverted Y-configuration and residual fusiform basilar aneurysm sac.

Figure 4 (A) Axial and (B) three-dimensional volume-rendered CT angiography images obtained 6 months after inverted Y-stenting showing satisfactory occlusion of the aneurysm and patency of the lumen.

In order to optimize the flow diversion qualities of the overlapping stents, it is important to minimize the cell size of the deployed stents. First, a stent larger than the diameter of the basilar artery was inserted. In addition, a 4.5 mm stent was inserted inside a 3.5 mm stent, thereby limiting the cell size of the larger stent. In this case, blood flow diversion was achieved through the parent vessel using two non-flow diversion devices which subsequently resulted in obliteration of the basilar artery fusiform aneurysm.

Learning points

- ▶ Rhombencephalosynapsis may predispose to the formation of symptomatic fusiform basilar artery aneurysms.
- ▶ The use of an inverted Y-configuration of open-cell stents can successfully create flow diversion within a fusiform basilar artery aneurysm.
- ▶ Inverted Y-stenting appears to be a safe and effective treatment approach for treating symptomatic fusiform basilar artery aneurysms.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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