115 Basilar Artery Trunk Aneurysm: Concomitant Retroperitoneal and Subarachnoid Hemorrhage, Segmental Arterial Mediolysis (SAM), Dissecting Aneurysm, Treatment by Partial Coil Occlusion and Flow Diversion

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Abstract

"Segmental mediolytic arteriopathy" or "segmental arterial mediolysis" (SAM) is an idiopathic disorder of visceral and intracranial arteries and known as a cause of major abdominal, retroperitoneal, and subarachnoid hemorrhage (SAH). The affected arteries show a non-inflammatory and non-atherosclerotic vacuolization and lysis of the tunica media, smooth muscle degeneration, and serration of the lamina elastica interna, undermining the vessel wall stability. Spontaneous dissection and aneurysm formation followed by aneurysm rupture may occur. SAM is the most likely diagnosis in the case of simultaneous abdominal or retroperitoneal and subarachnoid hemorrhage. This is the case of a patient with simultaneously ruptured dissecting aneurysms of abdominal and intracranial arteries. The evolution and treatment of the dissecting basilar artery aneurysm by endovascular coil-assisted flow diversion is the main topic of this report.

Keywords

Basilar artery · Segmental arterial mediolysis · SAH · Dissecting aneurysm · Partial coil occlusion · Flow diversion

Patient

A 30-year-old, previously healthy, male patient collapsed during his office work after complaining of severe headache. There was no history of trauma.

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Diagnostic Imaging

The patient became hemodynamically unstable, was intubated, and brought to the emergency room. An abdominal, thoracic, and cranial CT examination showed a massive retroperitoneal and subarachnoid hemorrhage (Hunt and Hess IV, Fisher 3) (Fig. 1a, b). The laparotomy showed a rupture of the splenic artery, hepatic and splenic lacerations, and fragile abdominal vessels. The patient underwent emergency splenectomy and external ventricular drainage procedures. DSA of the cervical and intracranial vessels 3 days after the initial event showed remnants of previous dissections of both ICAs (Fig. 1c, d). In the middle section of the basilar artery, a small blister aneurysm was recognized (Fig. 1e). Only 13 days following this first DSA examination, a second SAH occurred (Fig. 1f) and was due to a large saccular aneurysm of the basilar trunk (Fig. 1g). The second DSA examination now showed a large dissecting aneurysm, which had developed from the previous blister of the basilar artery (Fig. 1h).

Treatment Strategy

The primary goal of the treatment was to prevent a recurrent SAH. Neither the location, the size of the aneurysm, nor the patient's condition allowed microsurgical treatment. Also, due to the relatively wide neck of the aneurysm at 3.6 mm, it was not suitable for coil occlusion only.

Treatment,

Procedure, 10.05.2016: coil-assisted flow diversion of a ruptured, dissecting basilar artery trunk aneurysm

Anesthesia: general anesthesia; 1× 3000 IU unfractionated heparin (Heparin-Natrium, B. Braun) IV, 1× 1000 mg thiopental (Trapanal, Nycomed) IV, 1× 40 mg dexamethasone (Fortecortin Inject, Merck Serono) IV

Premedication: 1× 500 mg ASA (Aspirin i.v., Bayer Vital) IV and 2× 90 mg ticagrelor (Brilique, AstraZeneca) PO via nasogastric tube and eptifibatide (Integrilin, GlaxoSmithKline) IV as body weight adapted bolus dose; Multiplate (Roche Diagnostics) confirmed dual platelet function inhibition

Access: right femoral artery, 8F sheath (Terumo); *guide catheter*: 6F Heartrail II 100 cm (Terumo); *microcatheters*: 1× Excelsior XT-17 (Stryker) with J-tip for coil insertion; 1× Excelsior XT-17 (Stryker) for flow diverter implantation

Implants: coils: 2× Deltamaxx 6/25 mm (Codman); flow diverter: 1× p64 3/15 mm (phenox)

Course of treatment: a 6F guide catheter was placed in the V2 segment of the left vertebral artery (VA). DSA confirmed the dissecting basilar artery trunk aneurysm with a diameter of 9 mm. Via the left VA, an Excelsior XT-17 microcatheter was advanced to the center of the aneurysm, and two coils were placed without difficulty. Afterward, the Excelsior XT-17 microcatheter was removed, and an Excelsior XT-17 microcatheter was advanced through the BA to the left posterior cerebral artery (PCA). A p64 3/15 mm flow diverter was safely implanted. A complete coverage of the dissected segment of the basilar artery, including the orifice of the aneurysm was achieved. It was assumed that the vessel wall of the BA was quite fragile. Therefore the flow diverter was slightly undersized in order to avoid excessive impact on this already dissected artery. The final run showed contrast stasis in the aneurysm. The procedure was well tolerated (Fig. 2).

Duration: 1st to 45th DSA run: 1 h 41 min; fluoroscopy time: 50 min

Complications: none

Post medication: An increased dosage of 1× 500 mg ASA and 2× 180 mg ticagrelor, both PO daily, was required to maintain sufficient platelet function inhibition due to thrombocytosis after splenectomy and was combined with

- Low molecular weight heparin: 2× 3000 IU certoparin (Mono-Embolex, Novartis) SC daily for 6 weeks after the treatment

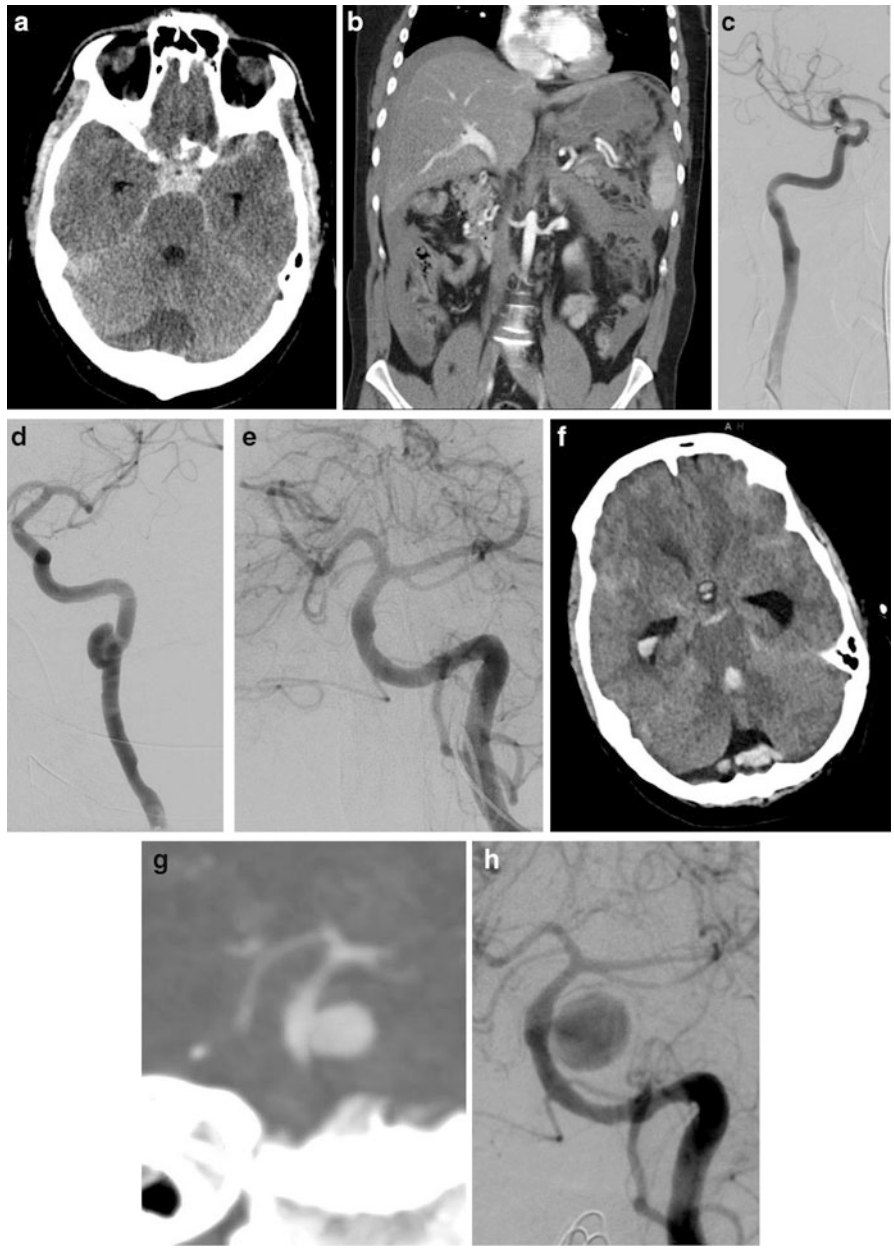

Fig. 1 CT and DSA findings in a case of concomitant abdominal and subarachnoid hemorrhage due to SAM. A cranial CT examination of a 30-year-old male patient with a massive SAH (**a**). An abdominal CT examination of this patient revealed a retroperitoneal hemorrhage due to the rupture of the splenic artery (**b**). The DSA examination of the cervical and intracranial arteries showed remnants of previous dissections on both internal carotid arteries (**c** and **d**) and a blister aneurysm of the basilar artery (**e**). A cranial CT examination 13 days after the DSA examination confirmed the suspected recurrent SAH (**f**) and revealed a saccular dissecting aneurysm of the basilar trunk (**g**). A DSA examination two days later showed a large dissecting aneurysm of the basilar trunk (**h**)

– 3× 4 mg dexamethasone (Fortecortin, Merck Serono) PO daily for 10 days, with tapering off thereafter
– 1× 90 mg etoricoxib (Arcoxia, Grünenthal) PO daily for six weeks
– 2× 150 mg ranitidine (Ranitidin, 1A Pharma) for two weeks

The dosage of the antiplatelet medication was gradually reduced during the course of the following year while maintaining sufficient platelet function inhibition, monitored by repeated Multiplate (Roche Diagnostics) and VerifyNow (Accriva) response tests to 1× 100 mg ASA and 2× 90 mg ticagrelor, both PO daily.

Clinical Outcome

Various issues like small bowel perforation, frontal subdural hematoma after ventricular shunting, and recurrent revision laparotomies dominated the following clinical course. The patient recovered with a Barthel index of 90 only five months after the clinical onset despite the fulminant beginning, the nature of his disease, and a variety of subsequent abdominal complications. After 11 months, the patient presented with mRS 0 for the follow-up examination. A genetic examination of the patient showed a heterozygotic mutation of the COL3A1 gene.

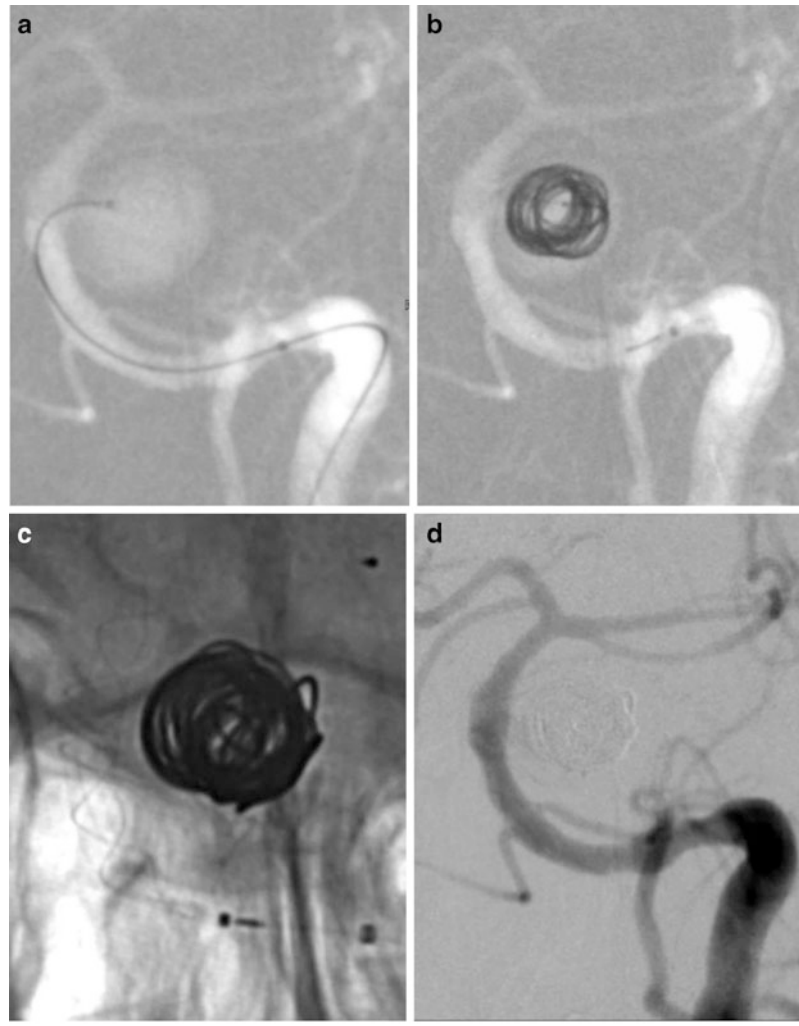

Fig. 2 Treatment of a large dissecting aneurysm of the basilar artery trunk. The aneurysm was gently catheterized (**a**) and then loosely filled with coils (**b**) and covered with a p64 flow diverter (**c**). The final DSA run confirms a significant reduction of blood circulation inside the aneurysm (**d**)

Follow-Up Examinations

DSA and MRI/MRA of the cervical and cranial vessels 11 months after the clinical onset confirmed the complete obliteration of the dissecting basilar artery trunk aneurysm, with an unchanged appearance of the remaining vessels (Fig. 3).

Discussion

The imaging correlates in SAM reported in the literature, include single or multiple dissection(s), intramural hematoma, arterial stenosis and occlusion, and fusiform or saccular aneurysms. Splenic, celiac, mesenteric, and renal arteries are responsible for the abdominal manifestations. SAM has been described for the ICA, ACA, MCA, VA, and BA as well as for spinal arteries (Leu 1994; Welch et al. 2017); the histopathological findings in SAM include patchy vacuolar degeneration of smooth muscle cells of the arterial tunica media, fibrin deposition at the media-adventitia junction, and mucoid material (Slavin and Gonzalez-Vitale 1976; Slavin et al. 1989; Slavin 2009). The tunica media can be missing, bringing intima and adventitia in direct contact (Baker-LePain et al. 2010). Alterations related to vasculitis or atherosclerosis are missing. The relation of SAM to fibromuscular dysplasia (FMD), cystic medial necrosis (CMN), and the vascular Ehlers-Danlos syndrome is a matter of debate.

The presumed or considered diagnoses of the underlying vascular disorders in our patient included vascular Ehlers-Danlos syndrome, Loeys-Dietz syndrome, Erdheim-Gsell cystic medial necrosis, and segmental arterial mediolysis. These diseases are known to show overlapping features (Loeys et al. 2006). The genetic examination of our patient revealed a heterozygotic mutation of the COL3A1 gene, which is known to be associated with type IV (vascular type) Ehlers–Danlos syndrome.

Inflammatory vasculopathies such as polyarteritis nodosa were excluded from the diagnosis because there was no inflammation of the vessel walls histologically observed (Baker-LePain et al. 2010).

The concomitant manifestation of SAM on abdominal and neurovascular arteries is rare. We identified 12 published cases (Welch et al. 2017; Kubo et al. 1992; Fuse et al. 1996; Sakata et al. 2002; Obara et al. 2006; Ro et al. 2010; Stetler et al. 2012; Matsuda et al. 2012; Alturkustani and Ang 2013; Cooke et al. 2013; Pillai et al. 2014; Shinoda et al. 2016). The key features of these reported cases are summarized in Table 1.

There is no general treatment strategy for SAM-associated ruptured aneurysms. For abdominal aneurysms, endovascular treatment or surgery can be considered (Shenouda et al. 2014). Intracranial dissecting aneurysms are usually not ideal surgical targets (Kitanaka et al. 1994). For vertebral artery dissections, parent vessel occlusion with coils is

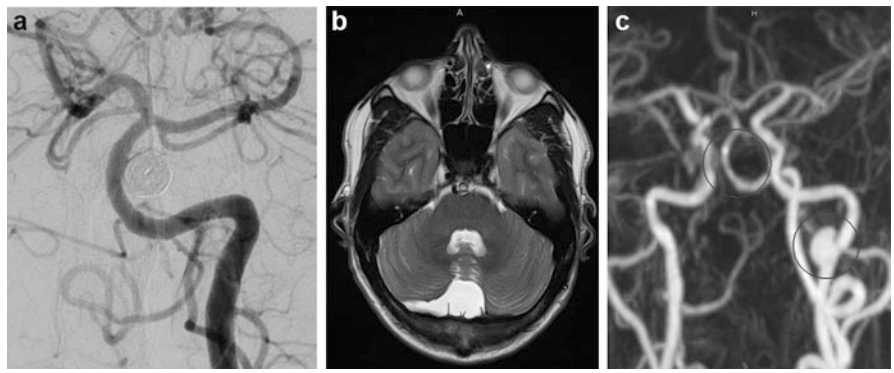

Fig. 3 DSA 16 months after the endovascular treatment of a dissecting basilar artery aneurysm confirmed the reconstruction of the artery with complete occlusion of the aneurysm (**a**). MRI T2WI revealed no damage of the brain stem (**b**). Contrast-enhanced MRA showed sequelae of previous, clinically unobserved dissections of both ICAs (**c**)

Table 1 Segmental arterial mediolysis (SAM) with concomitant visceral or thoracic and neurovascular manifestation. A review in chronological order of 12 previously published cases

Authors	Patient age, gender	Visceral manifestation	Neurovascular manifestation	Histology	Clinical manifestation	Treatment
Kubo et al. (1992)	56, female	Hepatic artery, ruptured aneurysm, splenic artery, several incidental aneurysms	Right cervical ICA, ruptured aneurysm, left VA, incidental fusiform aneurysm	–	Cervical hematoma ⇒ abdominal hemorrhage	Surgery
Fuse et al. (1996)	56, female	Gastroepiploic artery, ruptured aneurysm; gastric arteries, incidental	Left intradural ICA, ruptured aneurysm; right MCA bifurcation aneurysm, incidental	–	SAH ⇒ abdominal hemorrhage	Surgery
Sakata et al. (2002)	48, male	Superior mesenteric artery, bilateral renal artery, left external iliac artery, dissections	Right VA and left ICA, fusiform dilatation, ruptured aneurysm	+	SAH	Conservative
Obara et al. (2006)	52, male	Hepatic, celiac, superior mesenteric artery aneurysms and stenoses	Left ICA dissecting aneurysm, stroke	+	Stroke	Surgery
Ro et al. (2010)	70, male	Right gastroepiploic artery, dissection, ruptured aneurysm; left gastric artery, dissection	Right VA, dissection, asymptomatic	+	Abdominal hemorrhage	Conservative
Stetler et al. (2012)	59, female	Right hepatic artery, ruptured aneurysm*	Right ICA/PcomA, ruptured aneurysm*	–	SAH ⇒ abdominal hemorrhage	Coil occlusion*
Matsuda et al. (2012)	58, male	Splenic, gastroepiploic, gastroduodenal, both renal artery aneurysms	Right ACA (A1* and distal), left VA, ruptured aneurysm	–	SAH	Surgery*
Alturkustani and Ang (2013)	47, male	Aortic dissection, incidental	Left VA (V4), ruptured fusiform aneurysm	+	SAH	Conservative
Cooke et al. (2013)	45–55, male	Right internal mammary, celiac, both renal artery dissecting aneurysms	Left VA*, ruptured aneurysm	–	SAH	Coil occlusion*
Pillai et al. (2014)	?	Celiac artery, dissection	Both ICAs, stroke	?	Stroke	?
Shinoda et al. (2016)	47, male	Middle colic artery, ruptured fusiform aneurysm*	Extracranial VA, thyrocervical artery, incidental dissections; intradural VA, ruptured dissection*	+	SAH ⇒ abdominal hemorrhage	Coil occlusion*
Welch et al. (2017)	61, male	Splenic artery aneurysm, hemorrhage	Posterior spinal artery aneurysm	–	Spinal SAH abdominal hemorrhage	Embolization

Note: *Indicated as for treated aneurysms

widely used (Halbach et al. 1993). For dissected intracranial arteries, which could not be occluded, stent reconstruction (with or without coil insertion) was for many years the only treatment option (Zhang et al. 2016). In the majority of cases, self-expanding stents, developed to assist coil occlusion of aneurysms, had been used. The implantation of flow diverters for this purpose has several advantages; the coverage of the dissected vessel is denser, and the radial force is applied more evenly than that of self-expanding stents. This may improve the readaptation of the separated vessel wall layers. There is very little hemodynamic impact of a self-expanding stent on a covered aneurysm. If, as in our patient, a dissection is the origin of a large saccular pseudoaneurysm, the hemodynamic effect of a flow diverter is advantageous to prevent re-rupture. Meanwhile, flow diversion has become a recognized treatment option for intracranial dissections (Saliou et al. 2016). For this indication, as for many others, the required dual platelet function inhibition is a major drawback. The initial presentation of SAM can be fulminant, as demonstrated by our patient. If this phase is survived, long-term disease-free survival has been reported (Baker-LePain et al. 2010).

Therapeutic Alternatives

Flow Diversion
Parent Vessel Occlusion
Stent-Assisted Coiling
Telescoping Stents

Acknowledgments This case report has been previously published with open access (Hellstern et al. 2017).

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