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Technical Note

A Case of Scalp Arteriovenous Malformation Treated by Transvenous Embolization Using Onyx

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Objective: We treated a case of scalp arteriovenous malformation (sAVM) by transvenous embolization using Onyx.
Case Presentation: We describe the case of a 17-year-old woman with a pulsatile mass at the right temporal area. DSA identified sAVM with the venous pouch between the right occipital artery (OA) and the right two occipital veins (OVs), which was also fed by multiple branches of the right posterior auricular artery (PAA) and superficial temporal artery (STA). The shunts were completely occluded by the reverse pressure cooker technique (RPCT), which involves navigating the balloon catheters just distal to the shunt point in the OVs approaching from the right external jugular vein (EJV) and injecting Onyx to each feeder retrogradely with balloons inflated.
Conclusion: This technique may be useful for treating sAVM with venous angioarchitecture enabling a transvenous approach.

Keywords ▶ scalp arteriovenous malformation, transvenous embolization, Onyx

Introduction

Scalp arteriovenous malformation (sAVM) is relatively rare disorder. It forms diverse networks between the scalp arteries and veins without capillary. It is noticed as a palpable pulsatile mass in many cases. Symptoms, such as vascular murmurs, tinnitus, headache, local pain, epilepsy, hemorrhage, and necrosis of the scalp, are observed in some cases.[1,2] Surgical resection, endovascular treatment, or their combination is performed, but treatment guidelines have not been established. Treatment is selected in accordance with patients.

In this study, we report a case of sAVM with a large number of tortuous feeders and several drainers, which was completely cured by transvenous embolization using the reverse pressure cooker technique (RPCT) in which Onyx (Medtronic, Irvine, CA, USA) is retrogradely infused under occlusion of drainers with balloon catheters.

Case Presentation

Patient: A 17-year-old girl.
Medical history: Lumbar disc herniation and childhood asthma.
Present illness: A palpable pulsatile mass was noted in the right temporal region 2 months previously. Vascular murmurs in the right ear were heard, sometimes leading to insomnia. She consulted a local hospital and was referred to our department because MRI demonstrated an abnormality.
Physical examination on admission: Consciousness was clear. There was no abnormal neurological finding. Palpation revealed a pulsatile mass measuring 3 cm in an area posterior to the right ear. There was no redness or tenderness (**Fig. 1**).
Imaging findings: CTA revealed a vascular structure suggestive of sAVM under the skin posterior to the right ear (**Fig. 1**). Cerebral angiography demonstrated sAVM involving the right occipital artery (OA) as a main feeder and two

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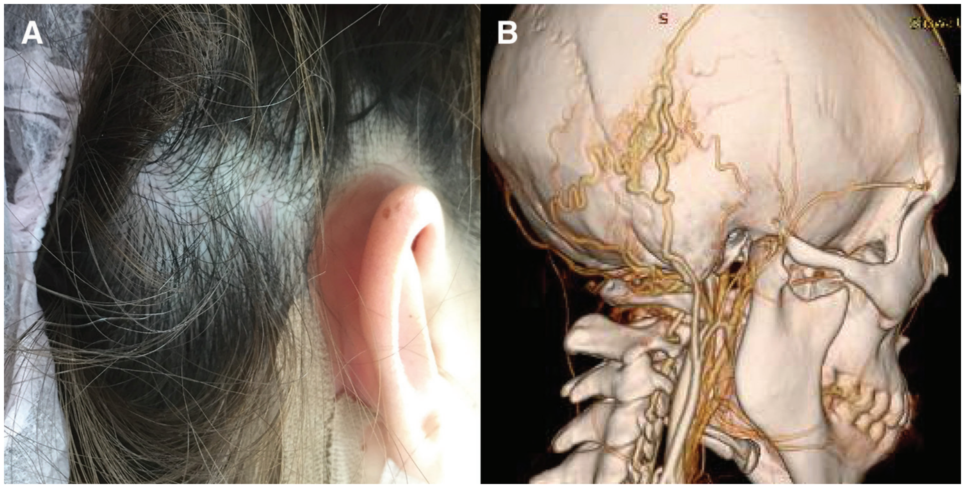

Fig. 1 The clinical photo (**A**) shows a 30-mm pulsatile mass behind the right ear with bruit at night. There was no redness or tenderness. CTA (**B**) shows abnormal angioarchitecture, suggesting sAVM. sAVM: scalp arteriovenous malformation

right occipital veins (OVs) as drainers. Feeders from the right posterior auricular artery (PAA) and right superficial temporal artery (STA) were also observed. The two right OVs had become confluent, exhibiting outflow to the right external jugular vein (EJV) (**Fig. 2**). After receiving informed consent, treatment using Onyx was performed, although it corresponded to off-label use. Furthermore, an application for highly difficult medical technologies was submitted.

Endovascular treatment course: Under general anesthesia, the procedure was started. A 6-Fr sheath was inserted into the right femoral artery and a 4-Fr JB2 (Medikit, Tokyo, Japan) was placed at the origin of the right OA. To improve microcatheter navigability, the right EJV was directly punctured and a 5-Fr sheath was inserted into the right EJV. Heparin at 5000 units was intravenously injected for systemic heparinization. Subsequently, the activated clotting time (ACT) was measured every 30 minutes to maintain a value of ≥250. After external carotid angiography, two Scepter C 4 × 10 mm balloon catheters (Terumo, Tokyo, Japan) were guided to an area adjacent to a venous pouch through the two right OVs, respectively, using a CHIKAI 0.014 200 cm (Asahi Intecc, Aichi, Japan) under a roadmap. The two Scepter C balloon catheters were inflated to occlude the drainers and Onyx34 was infused through the Scepter C balloon catheter placed below the venous pouch (**Fig. 2D**, white arrowhead). Initially, the venous pouch was filled with Onyx, which was allowed to retrogradely reach each feeder. When 3 mL was infused, angiography was performed. The shunt slightly remained and 1.5 mL of Onyx18 was infused. Subsequent angiography confirmed the disappearance of the shunt. After deflating the Scepter C balloon catheter placed below the venous pouch (**Fig. 2D**, white arrowhead), 0.5 mL of Onyx18 was infused to occlude the OV below the venous pouch. Subsequently, 1 mL of Onyx18 was infused after deflating the Scepter C balloon catheter placed above the venous pouch (**Fig. 2D**, white arrow) to occlude the OV above the venous pouch. The total volume of Onyx was 6 mL (**Fig. 3**). The Scepter C balloon catheters were able to be removed without problem. Right external carotid angiography confirmed the complete disappearance of the shunt and the procedure was completed (**Fig. 4**). Immediately after treatment, swelling at the lesion site and mild tenderness were observed, but they disappeared the day after treatment. Murmurs and pulsation also disappeared. There was no change in the skin tone, but the Onyx mass was hard and palpable. After 6 months, cerebral angiography confirmed the disappearance of the sAVM (**Fig. 5**). During the 2-year postoperative follow-up, recurrence on physical or MRI findings has not been observed.

Discussion

In 1764, Hunter first reported AVM of the elbow.[3,4] In 1833, Brecht termed scalp vascular anomalies cirsoid aneurysms.[3,4] Thereafter, several names, such as aneurysmal serpentinum, aneurysm cirsoides, plexiform angioma, arteriovenous fistula, and arteriovenous malformation,[5] have been used.

Various scalp blood vessels may be involved, but the STA or OA functions as a feeder in many cases.[5] Intracranial blood vessels rarely function as a feeder.[6] Etiological

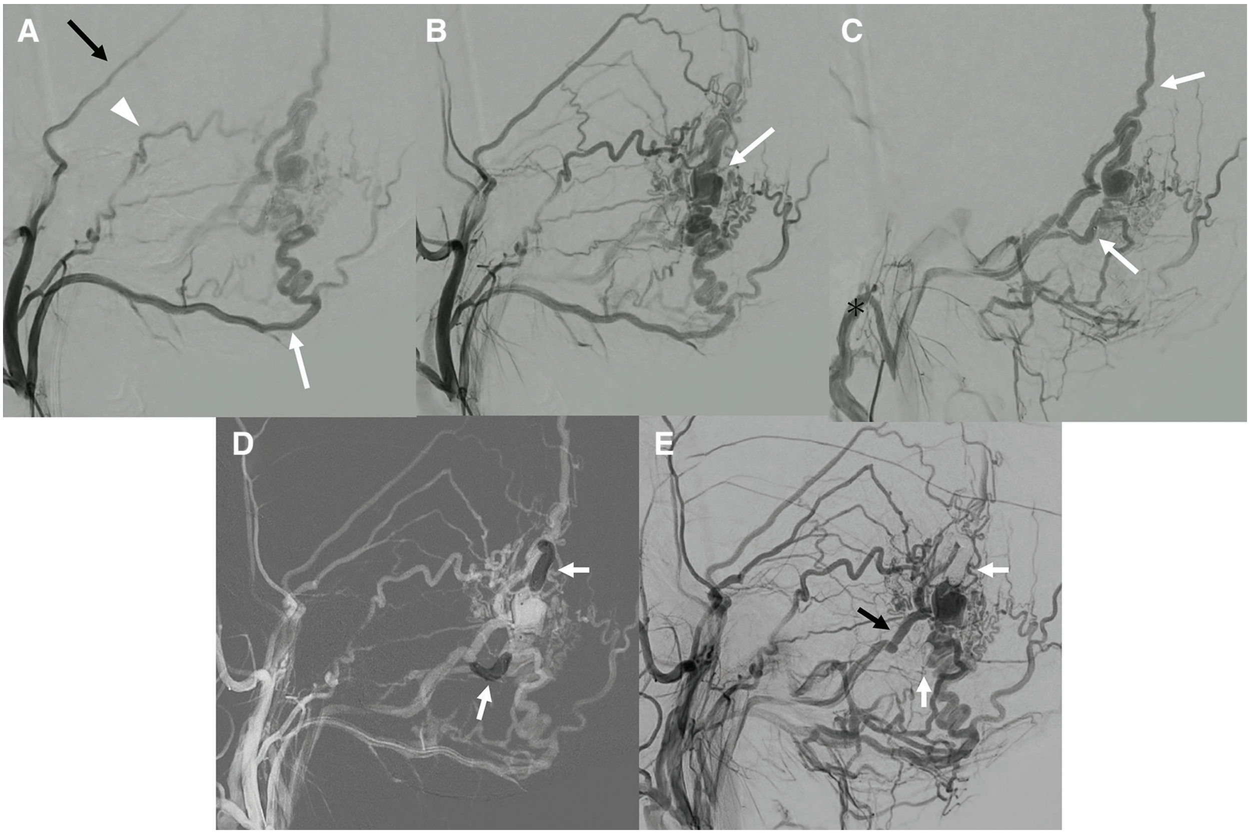

Fig. 2 Lateral DSA of the right external carotid artery in the early (**A**), capillary (**B**), and late (C) phases shows sAVM supplied from mainly the OA (**A**, white arrow) and the PAA (**A**, white arrowhead), the STA (**A**, black arrow), and through the venous pouch (**B**, white arrow), draining to the two OVs (**C**, white arrows) connecting to the EJV (**C**, asterisk). The two inflated Scepter C balloons in the OVs (**D**, white arrows) placed just distal to the venous pouch prevented the migration of the Onyx into the pulmonary artery and helped to pull out the Onyx into each feeder retrogradely. External carotid angiography with the inflated Scepter C balloons shows flow interruption of the OVs (**E**, white arrows) except the minor outflow veins (**E**, black arrow) to the EJV. EJV: external jugular vein; OA: occipital artery; OVs: occipital veins; PAA: posterior auricular artery; sAVM: scalp arteriovenous malformation; STA: superficial temporal artery

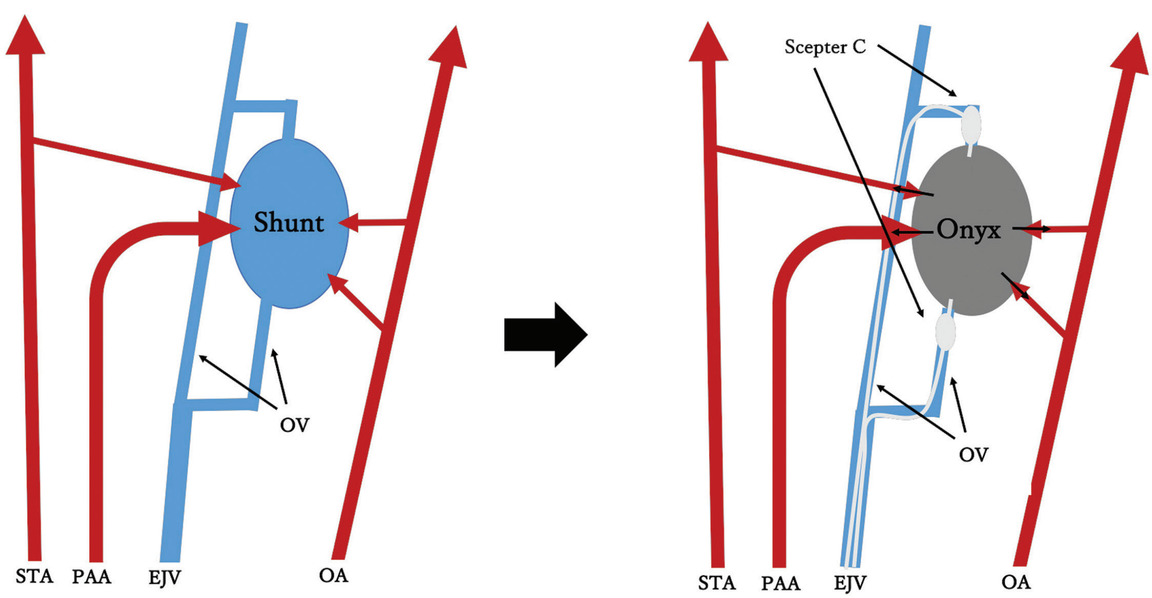

Fig. 3 Schematic diagram of the sAVM. sAVM: scalp arteriovenous malformation

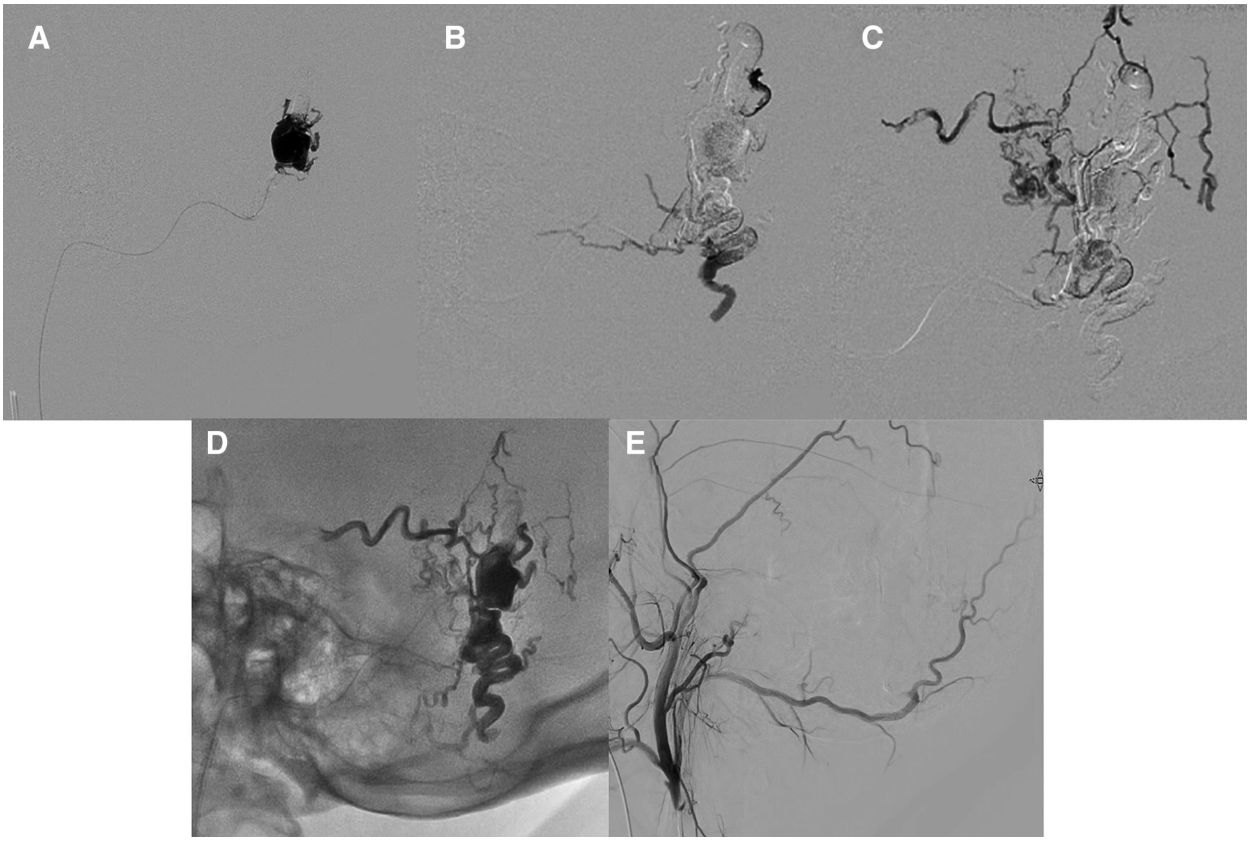

Fig. 4 Onyx 34 was injected through the inflated Scepter into the venous pouch (**A**), and entered into the OA and the STA retrogradely (**B**). Onyx 18 passed through the venous pouch and entered into the distal portion of the OA, STA, and PAA (**C**). DSA of the right external carotid artery after injecting Onyx shows complete occlusion of the AVM (**D**) and injected Onyx (**E**). AVM: arteriovenous malformation; OA: occipital artery; PAA: posterior auricular artery; STA: superficial temporal artery

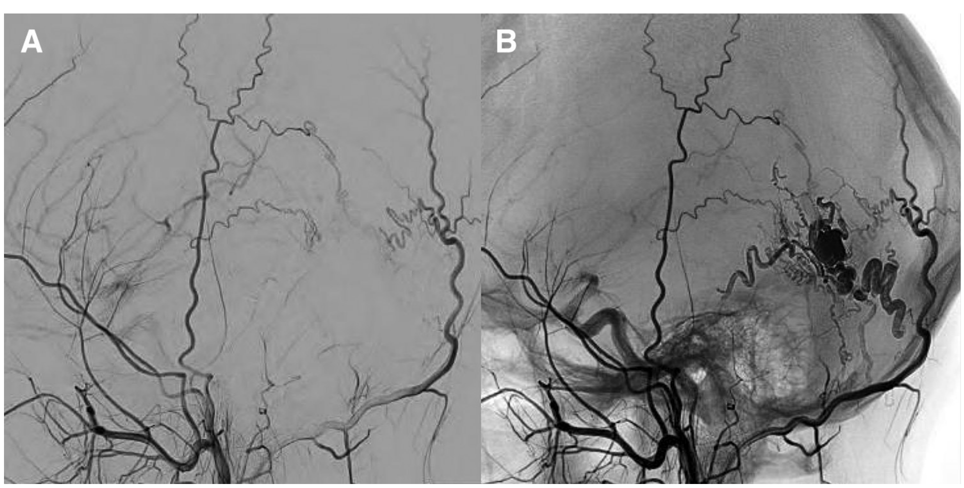

Fig. 5 Angiographic examination at 6 months after embolization confirms no sign of arteriovenous malformation (**A**) and shows the Onyx mass (**B**).

factors are classified into congenital and acquired. As acquired factors, there are many cases of blunt trauma, but scalp transplantation,[7)] craniotomy,[8)] and arthroscopy for temporomandibular joint disorder[9)] have been reported. Congenital factors remain to be not clarified. In the present case, a history of trauma was unclear. Concerning the course of sAVM, small subcutaneous swelling initially occurs, and a pulsatile mass and dilated drainer become palpable with the development of a drainer, leading to the appearance of cosmetic problems.[1,2)] Treatment is indicated

Table 1 Case summaries of scalp AVM treated using Onyx

Report	Age (years)	Sex	Etiology	Approach	Assist technique	Outcomes	Complications
Zheng J[2]	38	Male	T	TAE	None	Cure	None
	43	Female	C	TAE (+surgery)	None	Cure	None
Youn SW[17]	75	Male	C	TAE	Coil + NBCA	Cure	None
	63	Female	C	TAE	Coil + NBCA	Cure	None
Ni W[1]	19	Male	T	TVE	Coil	Cure	None
	34	Male	C	TVE	Coil	Cure	None
	54	Male	C	TVE	Coil	Cure	None
Dalyai RT[15]	Early 30s	Male	I	DP	None	Cure	None
	Early 40s	Male	I	TAE	None	Cure	None
	Early 60s	Female	I	TAE	Coil	Cure	None
Úrat A[19]	13	Male	T	TAE	Coil	Cure	None
	15	Female	U	TAE (+surgery)	None	Cure	None
	43	Female	T	TAE	None	Cure	None
Thiex R[13]	60	Male	C	TVE	Balloon	Cure	None
Kuwajima A[20]	25	Male	T	TAE	Coil	Cure	U
Corr PD[16]	7	Male	U	TAE	EC	Cure	None
	9	Male	U	TAE, TVE, DP	Balloon, EC	Cure	None
	25	Female	U	TAE	Balloon	Cure	None
	26	Female	U	DP	EC	Cure	None
	33	Male	U	DP	EC	Cure	None
	24	Male	U	DP	EC	Cure	Blackish discoloration at injection site
Whiteside OJ[11]	40	Male	T	TAE	Coil	Cure	None

AVM: arteriovenous malformation; C: congenital; DP: direct puncture; EC: external; I: iatrogenic; NBCA: n-butyl-2-cyanoacrylat; T: traumatic; TAE: transarterial embolization; TVE: transvenous embolization; U: unknown

for the prevention of hemorrhage, pulsatile mass-related cosmetic problems, the deterioration of local pain or tinnitus, or A-V shunt-related heart failure deterioration.[2,10,11]

Treatment methods include surgical resection and endovascular treatment. The respective treatments alone or combination therapy is selected in accordance with the vascular structure of a lesion. After 2000, surgical resection was performed in 62 patients, endovascular treatment in 40, and combination therapy in 9. Previously, surgical resection was selected as a first-choice procedure. Although the cost and recurrence rate were low, complications, such as scalp necrosis at the resected site and massive hemorrhage, were reported.[12] Furthermore, there is a report that postoperative vasospasm may lead to misrecognition of complete resection of aAVF.[2] Recently, endovascular treatment has been increasingly selected with advances in devices, such as microcatheters that may reach the periphery, embolic materials, and embolization techniques. Endovascular treatment methods include transarterial approach, transvenous approach, and direct puncture. In the above studies, transarterial approach was adopted in 13 patients, transvenous approach in 4, direct puncture in 25, and transarterial approaching + direct puncture in 4 patients. Of these patients, Onyx was used as an embolic material in 22 (**Table 1**),[13,14] n-butyl-2-cyanoacrylat (NBCA) in 24, coils in 12, and others (thrombin, polidocanol, and gelatin) in 5 patients. To achieve complete cure by endovascular treatment alone, it is necessary to occlude all feeders and shunt points. In cases with a single feeder in which it is easy to approach to an area adjacent to a shunt point, transarterial approach facilitates complete cure. However, the branch of the external carotid artery that works as a feeder is markedly flexed or tortuous in many cases. So, when performing embolization at a position distant from a shunt point, an embolic material may not penetrate all feeders and drainers, leading to insufficient embolization, which not only causes recurrence due to the development of a collateral pathway but also makes a more complex shape in comparison with that before embolization, making approaching more difficult. This may cause necrosis of the scalp or alopecia.[1,15] On the other hand, drainers are often dilated and may be useful as an effective access route. In addition, they may not make microcatheter removal difficult. However, because most of the drainers for sAVM consist of the scalp veins such as the superficial temporal vein (STV),[2] regurgitation to the venous side may result in outflow to pulmonary circulation, so caution is needed. There is also a report of outflow to the transverse or sigmoid sinuses via the bone marrow.[16] Direct puncture is simple and the cost is low because no microcatheter is used. This method can approach lesions with a tortuous feeder. Pressure insertion into each feeder is also possible while preventing regurgitation to pulmonary circulation by manually compressing the venous side. However, postoperative pain, swelling, and infection at the site of puncture were reported.[17]

Our patient was a young female, and endovascular treatment was selected considering the degree of invasiveness and risk of hemorrhage. A complex vascular structure consisting of several, markedly tortuous feeders and shunt points was observed, and transarterial approach to an area adjacent to a shunt point was considered to be difficult. However, it was considered possible to guide a microcatheter to a venous pouch via two drainers, the dilated OVs; therefore, a transvenous approach was selected. Although direct puncture was also considered possible, it was not selected because of scalp invasiveness as well as surgical resection. To achieve complete cure, it is necessary to retrogradely occlude a venous pouch and all feeders; therefore, Onyx was selected as an embolic material. Onyx has a property of non-adhesiveness, differing from NBCA, and it can be continuously infused based on a plan, improving the AVM embolization rate.[1,13,18] No previous study regarding the use of Onyx for sAVM has reported recurrence. As a complication, black coloration of the skin related to the subcutaneous leakage of Onyx (1 case) was noted. In this case, resection was performed afterwards. Furthermore, two cases in which resection was performed on the Onyx mass remaining under the skin were reported.[19] In the present case, a palpable Onyx mass also remained. Dimethyl sulfoxide (DMSO), the solvent for Onyx, is known to be cytotoxic, and its capable daily volume is limited as ≤4.5 mL. To date, no study has reported DMSO toxicity to the scalp. In the present case, the volume of Onyx was 6.0 mL, but there was no adverse event. Concerning the precipitation time of Onyx, hardening after an interval of ≥3 minutes was regarded as complete. In the present case, the balloon was deflated 7 minutes after initial Onyx infusion and there was no regurgitation to the venous side. It is also necessary to explain the off-label use of Onyx for sAVM and receive informed consent.[20]

As a method to infuse Onyx, we selected the RPCT in which Onyx is retrogradely infused into a feeder under proximal balloon protection. A balloon catheter was guided to an area adjacent to a venous pouch to occlude a drainer, facilitating pressure glue insertion into each feeder and embolization while preventing regurgitation to pulmonary circulation. As a balloon catheter, we used a Scepter C with a double-lumen structure. A previous study also reported transvenous approaching with balloon protection,[13] but a HyperForm (Medtronic, Minneapolis, MN, USA) with a single-lumen structure was used. When adopting a balloon catheter with a single-lumen structure, another microcatheter for Onyx infusion is necessary. However, a Scepter C alone allows balloon protection and Onyx infusion; therefore, the procedure is simple and Onyx can be infused while occluding several large drainers, as demonstrated in the present case. The maximum outer diameter of a Scepter C is 2.8 Fr; therefore, three drainers may be theoretically simultaneously managed if an 8-Fr guiding catheter can be inserted. Furthermore, the outer diameter of its tip is 2.1 Fr, being thinner than those of other balloon catheter tips; a Scepter C is useful for navigating to distal. However, if marked dilation of a drainer beyond the diameter of a balloon makes occlusion impossible, the use of a coil or direct puncture must be selected. A previous study reported a method to prevent Onyx regurgitation by inserting a coil to an area adjacent to a venous pouch, but not under balloon protection.[1] However, this method requires two microcatheters per drainer to be occluded, so procedure is complex. In addition, coil placement on the venous side has a risk of distal migration. The RPCT using a Scepter C, which we performed in the present case, may be simple and effective as a transvenous approach method for sAVM.

Conclusion

We reported a patient with sAVM in whom transvenous Onyx embolization using RPCT was successful.

This method may be simple and effective even for patients with a complex vascular structure that are difficult to cure with transarterial approach, if there is an approachable venous structure.

Disclosure Statement

The authors declare no conflict of interest.

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