Embolosclerotherapy by the Transvenous Approach for Lower Extremity Arteriovenous Malformation in Cowden Syndrome: A Case Report

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A 69-year-old female with Cowden syndrome presented with pain at rest in the right leg. Arteriovenous malformations (AVMs) of the right lower extremity were detected by computed tomography and magnetic resonance imaging. Angiography indicated arteriovenous fistulae, which were initially treated using a transarterial approach with minimal therapeutic effect. In contrast, excellent outcomes were achieved with a transvenous approach using coil embolization and liquid sclerotherapy for the venous component of the nidus. At 15 months after embolosclerotherapy, no angiographic evidence of AVM recurrence was noted. Embolosclerotherapy by the transvenous approach for AVM in Cowden syndrome was a useful therapeutic strategy for arteriovenous fistulae.

Keywords: arteriovenous malformation, embolosclerotherapy, Cowden syndrome

Introduction

Cowden syndrome is an autosomal-dominant disorder, and associated with multiple hamartomatous and neoplastic diseases.1) This condition was first reported in 1963, and causative mutations have been identified in phosphatase and tensin homolog (PTEN), a tumor suppressor gene located at 10q23. Arteriovenous malformations (AVMs) and hemangiomas are not widely recognized features of Cowden syndrome. There is no mention of other vascular malformations in Online Mendelian Inheritance in Manor in the current diagnostic criteria for Cowden syndrome.2) However, some articles have reported a relationship between Cowden syndrome and AVMs.2–4)

Although the transarterial approach is preferred to embolize AVMs when possible,5) this general approach cannot be applied in all cases. Here, we report a case of AVM in Cowden syndrome that was initially treated using a transarterial approach with minimal therapeutic effect, but that achieved excellent outcomes when approached via a transvenous approach.

Case Report

A 69-year-old female presented with pain at rest in the right leg and in both knee joints. She had been diagnosed with Cowden syndrome that was complicated with esophageal polyposis, oral papilloma, uterine cancer, and goiter. She also had osteoarthritis of the knee that caused the knee joint pain. AVMs of the right lower extremity were detected by computed tomography and magnetic resonance imaging that were performed because of the knee joint pain. Transcatheter embolization for AVM was planned, for the purpose of pain control in the right leg and to decrease bleeding at surgery for osteoarthritis of the knee.

Angiography and three-dimensional digital subtraction angiography indicated that the AVM in the right lower leg consisted of multiple feeding arteries from the dilated peroneal artery/anterior tibial artery and three drainage veins, which were two dilated peroneal veins and a superficial vein (Fig. 1). We diagnosed the patient with arteriovenous fistulae. The nidus was located between the tibia and fibula, and there was erosion on the tibia.

Initially, the AVM was treated over three sessions via a transarterial approach using N-buty-2-cyanoacrylate (NBCA)-lipiodol as an embolic material, but the therapeutic effect was minimal.

Next, endovascular therapy by the transvenous approach was undertaken. A retrograde approach from the right common femoral vein using a 5F sheath and an
Antegrade approach from the right common femoral artery using a 4F sheath were used. Angiography before and after endovascular therapy by a transarterial approach showed that the number of feeding arteries from the peroneal artery/anterior tibial artery were decreased slightly and that a superficial drainage vein had disappeared, but the therapeutic effect remained minimal. A 5.2F balloon catheter (Selecon MP, Terumo Clinical Supply Co., Ltd., Gifu, Japan) was inserted through the sheath of the common femoral vein and advanced to the dilated peroneal vein using a 0.035 inch guidewire. Then, a 2.5F microcatheter was advanced and placed in another dilated peroneal vein. After the balloon was inflated, superficial femoral artery angiography demonstrated that blood flow was reduced and that the dilated drainage vein had disappeared. The dilated peroneal vein and venous sac were embolized using coils through a microcatheter.

Then, 6 mL of 1% polidocanol were slowly injected through a balloon catheter. After embolosclerotherapy, pain at rest in the right leg disappeared. Endovascular therapy was performed for residual AVMs of knee and plantar over three sessions at the patient’s request. At 1 year and 3 months after

Fig. 1 Pretreatment angiography of the right lower leg shows a plexiform arteriolar component (arrows) and a large venous component (arrowheads). (a) arterial phase, (b) late arterial phase, (c) venous phase.

Fig. 2 Pre-treatment angiography shows two dilated peroneal veins and a large venous component. A balloon placed at one of the dilated peroneal veins was inflated (a). Another dilated peroneal vein and the venous sac were embolized using coils through a microcatheter (b, c). Then, 6 mL of 1% polidocanol were slowly injected through a balloon catheter.
Ethanol is more effective than 1% polidocanol, but it is often associated with local complications compared with polidocanol. Polidocanol is useful for small, low-flow and superficial lesions. On the other hand, ethanol is useful for large, low and high flow and deep lesions. In the present case, we selected the safer polidocanol over ethanol as the sclerosing agent, because we succeeded in reducing blood flow velocity in the AVM using a technique of coil embolization and balloon occlusion for the venous component of AVM.

In the present case, endovascular therapy for AVM by a transarterial approach was undertaken at three sessions before the transvenous approach was used. When we initially diagnosed arteriovenous fistulae based on angiography, the transvenous approach should have been attempted as soon as possible. We think that there are venous thrombosis and venous valve damage due to rough catheter manipulation as some possible complications related with this endovascular therapy for AVM by a transvenous approach.

Conclusion

AVM is a rare complication of Cowden syndrome. In the present case, we performed embolosclerotherapy with a transvenous approach using coil embolization and polidocanol as the sclerosing agent for AVM of the lower extremity complicated by Cowden syndrome. Thus, we recommend that the preferential treatment for type II arteriovenous fistulae should be endovascular therapy with a transvenous approach using a technique of coil embolization and liquid sclerotherapy with balloon occlusion of the venous component.

Discussion

AVMs and hemangiomas are not widely recognized features of Cowden syndrome, and only three previous articles have reported a relationship between Cowden syndrome and AVMs. In the present case, we also suspected a relationship between Cowden syndrome and AVMs.

In 1993, intracranial AVMs were classified into three types (arteriovenous, arteriolovenous, and arteriovenous) based on the form of the nidus. In 2006, an angiographic classification of AVMs in the body and extremities was reported. AVMs were classified into four types (Type I, II, IIIa, and IIIb) according to their angiographic morphologies. Generally, the transarterial approach has been preferentially used to embolize AVMs. However, this general approach should not be applied to all classification types, particularly to type II (arteriolovenous fistula). Endovascular therapies for type II AVMs by a transarterial approach were undertaken initially, but the therapeutic effect was minimal. In contrast, excellent results were achieved for type II AVMs using direct puncture or a transvenous approach with or without coil embolization for the venous component of the nidus. Combined embolosclerotherapy of pelvic AVMs with coils and ethanol was reported, and this therapeutic method was efficacious at achieving complete remission of AVMs.

Polidocanol is commonly used endovenously as a sclerosing agent for the treatment of small varicose veins. Ethanol is more effective than 1% polidocanol, but it is often associated with local complications compared with polidocanol. Polidocanol is useful for small, low-flow and superficial lesions. On the other hand, ethanol is useful for large, low and high flow and deep lesions. In the present case, we selected the safer polidocanol over ethanol as the sclerosing agent, because we succeeded in reducing blood flow velocity in the AVM using a technique of coil embolization and balloon occlusion for the venous component of AVM.

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Disclosure Statement

All authors declare that they have no conflicts of interest.

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