



Thrombosis of a Long-Segment Aneurysm from the Iliac to Popliteal Artery Associated with Arteriovenous Malformation and Varicose Veins

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A 58-year-old male patient with severe claudication due to thrombosis of the left ilio-femoro-popliteal artery aneurysm. He also had a venous stasis ulcer with a history of multiple embolotherapy of arteriovenous malformation. Duplex sonography revealed reflux and varicose veins of the left great saphenous vein (GSV). A sequential bypass surgery was performed that consisted of excision of the left external iliac and common femoral artery aneurysm, external iliac to deep femoral interposition with an expanded polytetrafluoroethylene graft, and femoro-posterior tibial artery bypass with the reversed left GSV. Symptoms of claudication were alleviated and the chronic ulcer was healed in time. To our knowledge, this is the first report of successful bypass in a patient with arterial aneurysm, arteriovenous malformation, and venous insufficiency that can be diagnosed as an atypical case of Parkes Weber syndrome. Long-term follow-up is needed to define the fate of aneurysms and varicose vein graft.

Key Words: Arteriovenous malformations, Aneurysm, Varicose veins, Parkes Weber syndrome, Thrombosis

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INTRODUCTION

Reports of concomitant arterial aneurysm, arteriovenous malformation (AVM), and chronic venous insufficiency (CVI) are sparse in the literature. Here, we present the case of a patient with a leg ulcer complicated by arterial ischemia and CVI that was primarily caused by a thrombosed ilio-femoro-popliteal arterial aneurysm, AVM, and refluxed great saphenous vein (GSV).

Informed consent was obtained from the patient for the publication of this case report with images, not revealing personal informations.

CASE

A 58-year-old male patient visited the outpatient clinic complaining of left calf pain after walking more than 50 m and a 4-month history of an unhealed ulcer at the left medial malleolus (Fig. 1A). His left leg was swollen, and engorged varicose veins (VVs) as well as hyperpigmentation around the ulcer were noted, but no port wine stains were evident. The circumferences of the right and left mid-thighs were 37.5 cm and 48.5 cm, respectively, while those of the right and left mid-calves were 30.5 cm and 42.5 cm. He previously underwent excision of the vascular tissues in the left popliteal area and multiple embolosclerotherapy sessions to treat AVM of the lower left leg at another hos-

pital. Eight years prior, he underwent surgery to remove a pelvic mass that was pathologically confirmed as an organized hematoma. He denied any relevant family history.

Computed tomography angiography showed a diffuse aneurysmal change from the left common iliac artery (CIA) to the popliteal artery and thrombotic occlusion along the entire femoro-popliteal aneurysm (Fig. 2, 3). The maximum diameters of the popliteal artery aneurysm and external iliac artery (EIA) aneurysm were 58 mm and 27 mm, respectively (Fig. 3). Limb hypertrophy, subcutaneous swelling, a varicose GSV, and calcified lesions of the residual AVM in the lower left leg were also observed. The left leg was perfused by the collateral vessels from the patent deep femoral artery (DFA). On duplex ultrasonography, the diameters of the left GSV at the upper, middle, and lower thigh were 0.72, 0.63, and 0.79 cm, respectively; these were larger than the normal diameters on the right side (0.42, 0.39, and 0.45 cm, respectively). Deep vein thrombosis was not observed. The reflux durations of the left sapheno-femoral junction and posterior branches of the upper thigh were 4,886 ms and 4,866 ms, while the reflux durations of the mid-thigh and mid-calf were 1,034 ms and 1,339 ms, respectively.

Under the presumption that the patient had atypical Parkes Weber syndrome (PWS) lacking port wine stains,

we planned an elective open bypass surgery to reconstruct the arterial flow to the calf and remove the refluxed GSV. Using a curvilinear incision on the lower left quadrant of the abdomen, the left EIA was exposed and dissected. Due to severe periarterial adhesions, the CIA aneurysm was not actively exposed. Another longitudinal incision in the left inguinal area was used to expose the common, deep, and superficial femoral arteries. The thrombosed ilio-femoral aneurysm was excised and replaced by a ringed expanded polytetrafluoroethylene interposition graft (Advanta, Maquet, Wayne, NJ, USA) from the EIA to the DFA (Fig. 4A). A longitudinal incision along the thigh was used to ligate the ipsilateral autologous GSV at the sapheno-femoral junction and harvest its branches. Since there was no aneurysmal change along the GSV, we decided to use the GSV as a bypass conduit. We performed a bypass from the left femoral graft to the left posterior tibial artery (PTA) using this re-



Fig. 1. (A) The patient reported a 4-month history of a non-healing ulcer at the left medial malleolus. (B) At 1 week postoperative, the ulcer had noticeably healed.



Fig. 2. Computed tomography angiography image showing the aneurysmal change from the left common iliac artery to the left common femoral artery, occluded left superficial femoral artery, and popliteal artery aneurysm arteriovenous malformation with calcifications in the posterior subcutaneous layer of the proximal lower leg.

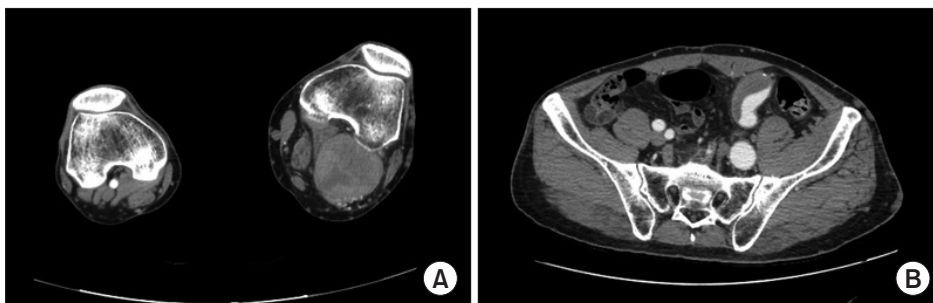


Fig. 3. Transverse view of the computed tomography angiograph showing (A) a thrombosed popliteal aneurysm and (B) an external iliac aneurysm with mural thrombus.

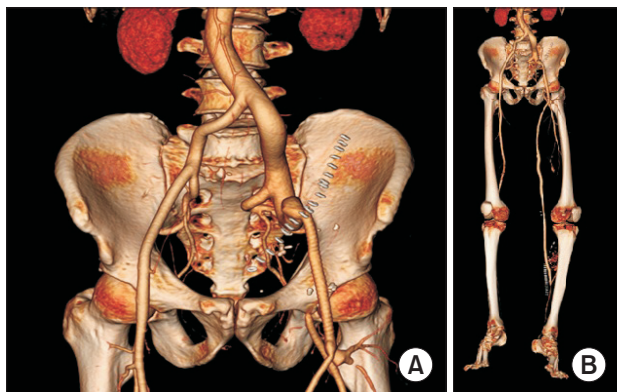


Fig. 4. Postoperative computed tomography angiography images showing (A) a left external iliac-deep femoral interposition graft with a ringed expanded polytetrafluoroethylene graft and (B) a left femoro-posterior tibial bypass with reversed left great saphenous vein.

versed autologous vein (Fig. 4B). At 1 week postoperative, the patient's symptoms were relieved and the left medial malleolar ulcer had healed remarkably (Fig. 1B).

DISCUSSION

To our knowledge, this is the first report of a successful bypass in a patient with congenital AVM combined with a long-segment ilio-femoro-popliteal artery aneurysm as well as CVI. We found no similar cases in the literature except those of Klippel-Trenaunay syndrome (KTS) with arterial aneurysms. KTS is an extremely rare disease characterized by limb hypertrophy, port wine stains representing capillary malformations, and superficial VVs [1]. Only six cases of KTS with arterial aneurysms have been reported in the literature; of these, two included popliteal aneurysms [2,3]. However, our patient had no capillary malformations such as port wine stains; instead, he had a history of being treated for AVM in the lower left leg at another hospital, which ruled out the diagnosis of KTS. Another rare entity, PWS, defined by the presence of capillary malformations such as port wine stains and limb hypertrophy with an underlying high-flow AVM can also be considered diagnostic for our case [4]. Roebuck stated that PWS has vascular malformation similar to those of KTS but differs due to the presence of any functionally significant arteriovenous fistulas [5]. Our case meets the definition of "atypical PWS" due to the lack of port wine stains. Diagnosis of PWS can be confirmed by *RASA1* gene mutations, which is significant because PWS patients are at a risk of cardiac overload, cardiac failure, and ischemia [6]. However, studies showed that *RASA1* gene mutations were not identified in PWS patients without capillary malformations or port wine stains [7]. We

concluded that genetic testing would be of no benefit since our patient had no relevant family history or port wine stains.

In our case, two main clinical dilemmas required attention: the severe claudication after walking 50 m and the non-healing venous ulcer. The patient had thrombosis involving a long segment of aneurysm from the external iliac to the popliteal artery with a maximum size of 5.8 cm that probably led to the chronic symptoms of severe claudication. In cases of popliteal arterial aneurysms, surgical repair is generally indicated for symptomatic patients, popliteal aneurysms greater than 2 cm, or cases with the presence of mural thrombus [8,9]. Therefore, to restore arterial inflow throughout the thrombus and aneurysm, we decided to perform elective open bypass surgery rather than endovascular surgery.

Once arterial flow has been restored, the next concern was that the venous ulcer could worsen due to venous hypertension caused by the increased arterial inflow and GSV insufficiency. We speculated that the congenital AVM increased the arterial inflow, causing the diffuse aneurysmal change over the affected limb from the CIA to the popliteal artery and that the increased outflow to the GSV caused venous dilatation and valvular reflux. Therefore, our patient's venous ulcer was most likely aggravated over time by the high inflow of the AVM and the venous pooling of the VVs. An AVM can affect the lower extremities in a wide variety of ways such as distal ischemia, venous ulcer or gangrene, and venous stasis dermatitis due to venous hypertension [10]. There are various approaches to treating AVMs such as sclerotherapy, transarterial embolization with onyx or ethanol and surgical resection of the AVM nidus [11,12]. Since AVM is very challenging to treat and features high recurrence rates, some studies have attempted to integrate sclerotherapy or embolization with surgery as a multidisciplinary approach [13]. Our patient had already undergone surgery and sclerotherapy in the past to treat AVMs. Because only a residual AVM was visible on the computed tomography scans, high ligation and stripping of the refluxed GSV was the next step in treating the CVI. Stripping is generally accepted as a safe and effective method with a low ulcer recurrence rate [14].

Another concern in this case was that a long bypass from the femoral to the PTA was required. Autologous vein conduits are considered better than prosthetic grafts in long bypasses due to their long-term patency and resistance to infection [15-17]. The use of a refluxed GSV as a conduit in arterial bypass is controversial because the increase in intimal hyperplasia and dilatation can eventually lead to aneurysm and thrombus formation [18,19]. Although weak in evidence, there was one case report of use of the vari-

cose GSV without an external mesh for a femoro-popliteal bypass [20]. In that case, the VV graft showed no dilatation or aneurysmal changes within a 15-month follow-up period. Thus, we decided to remove the refluxed GSV and use it as a bypass conduit in a reversed pattern after venorrhaphy of any area with aneurysmal change or weak valves. Fortunately, no focal dilatations of the GSV were noted after careful examination. As in this case, a full-length inspection and repair should be mandatory prior to the use of a varicose GSV as an arterial conduit since we do not know the long-term fate of refluxed GSV grafts. Our patient requires close long-term monitoring for any stenosis or aneurysmal change of the refluxed GSV graft.

In conclusion, in this complicated case of arterial ischemia and CVI due to thrombosed ilio-femoro-popliteal aneurysm, VV and AVM were treated successfully with aneurysm excision, an interposition graft from the EIA to DFA, and a bypass graft from the DFA to PTA with a reversed VV graft.

CONFLICTS OF INTEREST

The authors have nothing to disclose.

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