CASE REPORT

Treatment of severe Parkes Weber syndrome with flexion contracture of the lower limb

Chi-Yuan Liu a, Kuei-Ton Tsai b, Shuo-Suei Hung a,c,*

a Department of Orthopedics, Taipei Tzu Chi Hospital, Buddhist Tzu Chi Medical Foundation, Taipei, Taiwan
b Department of Cardiovascular Surgery, Taipei Tzu Chi Hospital, Buddhist Tzu Chi Medical Foundation, Taipei, Taiwan
c College of Medicine, Tzu Chi University, Hualien, Taiwan

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Summary Vascular malformations are difficult to treat, especially in cases with fast-flow arteriovenous fistulae. We herein present a case of severe Parkes Weber syndrome of the right lower limb, complicated by progressive heart failure and limb ischemia, eventually necessitating hip disarticulation. Vascular control was achieved with suture ligation of the right common iliac artery at its bifurcation, and ligation of the femoral artery from the inguinal site, but the patient still required massive transfusion due to a large amount of blood loss. Unfortunately, because of pulmonary edema, she died on the 5th postoperative day.

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1. Introduction

Vascular malformations are rare congenital disorders that occur in approximately 0.3–0.5% of the population, and have been systematically described by the Hamburg classification. These lesions can further be classified according to their flow rates, and be called, for example, slow-flow Klippel–Trénaunay syndrome (KTS), which may show great severity leading to additional limb deformity, or the fast-flow Parkes Weber syndrome (PWS) that may eventually cause cardiopulmonary failure. Treatment for majority of patients with slow-flow lesions includes graded compressive stockings, percutaneous sclerotherapy, or surgical resection. In patients with high-flow
malformations, radical surgery or limb amputation may be needed if embolization fails. Here, we describe a rare case of extensive congenital vascular anomaly, which led to cardiopulmonary dysfunction. The laboratory data and image studies, as well as the patient’s clinical course, are presented.

2. Case report

A 17-year-old female patient was referred to the Taipei Tzu Chi Hospital for treatment of progressive painful ischemia of the deformed right foot. It was reported that a soft mass had been found in her right dorsal foot since early childhood, and she had a tissue biopsy at the age of 3. Results of a pathological examination revealed a vascular lesion, and her parents refused any recommendation of treatment at that time. Therefore, the patient never received further treatment thereafter.

As she grew up, gradual swelling with flexion contracture of the right knee developed, and the patient (after attending junior high school) became chair-ridden because of functional disability of the limb.

Figure 1  Gross appearance of the patient with deformity of the right lower limb.

Figure 2  Images of the patient. (A) Whole-body bony structure with scoliotic spine. (B) Reconstructed computed tomographic angiography. (C) Angiography during embolization, showing poor result after placement of multiple coils.
Three months before her first visit to our orthopedic clinic, she experienced orthopnea, and the condition worsened soon in association with gangrenous changes of the right foot (Fig. 1), and therefore, a series of studies were arranged subsequently. A whole-body X-ray showed a scoliotic spine with dysplastic bony structure of the right lower limb (Fig. 2A). A lung perfusion scan excluded pulmonary embolism. Cardiomegaly was found, and pulmonary hypertension leading to right heart failure was shown by echocardiography, with a left ventricular ejection fraction of 77%. Angiography revealed a large systemic arteriovenous malformation with a $Qp/Qs$ ratio of 5.3. Vascular reconstructed images also showed extensive structural anomaly starting from the right common iliac artery to the entire right lower limb (Fig. 2B).

Embolization was attempted before right hip disarticulation, but the procedure failed due to excessive aneurismal dilatation of the vessels (Fig. 2C). The patient subsequently received open ligation of the right common iliac artery at its bifurcation (Fig. 3A), and ligation of the femoral vessels from the inguinal area (Fig. 3B). Despite adequate arterial control, the entire procedure still showed the presence of numerous intramuscular vascular malformations at the hip, and the total blood loss was estimated to be 11,000 mL.

The patient received 24 units of packed red blood cells, 4 units of whole blood, and 14 units of fresh frozen plasma to maintain her hematological stability during the operation, and her vital signs were stable in the early postoperative days. However, a fever up to 38.9°C developed on the 3rd postoperative day, and $pO_2$ as low as 39 mmHg was noted on the 4th postoperative day. A chest film showed hilar dilatation and repeated echocardiography showed poor left ventricular function with an ejection fraction of 44%. An impression of pulmonary edema with pneumonia was made, and despite aggressive resuscitation, the patient died on the 5th postoperative day.

### 3. Discussion

The PWS is differentiated from the KTS by the presence of arteriovenous fistula, and has a poorer prognosis and a higher rate of surgical complications. Few articles in the literature have reported on the treatment of PWS, especially on cases with such extensive involvement as in our case. Sclerotherapy and embolization are usually the initial choice before surgery in high-flow lesions. Hip disarticulation was decided for this patient after taking into consideration her functionless right lower limb with flexion contracture of the knee joint, and a progressive gangrenous change of the forefoot. However, due to high-flow shunting in the thigh, an adequate vascular control was mandatory during the surgery. Sclerotherapy was not considered in this patient because of massive involvement of the entire lower limb, and therefore embolization was attempted by a radiologist. Unfortunately, the coils failed to stay in place in the arterial system, and tended to enter the venous system as soon as they were released due to the strong flow and large caliber of the vessels. Because it was not possible to use a tourniquet during the surgery, vascular ligation proximal to the abnormal vessels or cardiopulmonary bypass was considered. After a review of the reconstructed images, the arterial blood supply was controlled for the right common iliac artery at its bifurcation site and for the right femoral artery from the inguinal area. The venous flow was ligated at the femoral vein, sparing the right common iliac vein to keep a better venous return from pelvic structures. The operation was complicated by the presence of dilated vessels in the right hip and thigh muscles, which might have

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**Figure 3**  Vascular controls. (A) Transection of the right common iliac artery with suture ligation of stump. (B) Looping of the engorged femoral artery before ligation.
emerged from the collateral circulations proximal to the common iliac artery, or from the back flow of the pelvic venous system, as the common iliac vein was not ligated. In such circumstances, perhaps ligation of the internal iliac vein and temporary control from the abdominal aorta would have been more adequate for this patient. Although hemostasis was finally controlled with the use of LigaSure (Covidien, Boulder, CO, USA), massive blood loss had already been encountered during the entire operation. Massive blood loss is defined as blood loss requiring transfusion of more than one blood volume in 24 hours, and massive transfusion is defined as the transfusion of a blood volume within 24 hours. Well-known complications of massive blood loss are hemostatic defects, electrolytes imbalance, and pulmonary insults. Transfusion-related acute lung injury (TRALI) is defined to occur within 6 hours of transfusion, and our case developed a fatal pulmonary condition as late as on the 5th postoperative day when her O2 saturation dropped abruptly, and therefore, it was not likely to be due to TRALI. The possible cause could be pulmonary edema following massive transfusion and fluid overload, because a butterfly pattern was found at the perihilar area on the chest plain film. The patient did not respond to cardiopulmonary resuscitation later in her clinical course. Had extracorporeal membrane oxygenation been available in our hospital, it could possibly have rescued this patient.

In conclusion, we described a case of severe PWS with complication of heart failure and limb ischemia. For such cases, adequate vascular control is mandatory during surgery, and the potential of massive bleeding from the collateral circulations and dilated vessels should not be underestimated. Although hemostatic devices such as LigaSure may be helpful in the control of venous bleeding, cardiopulmonary bypass ought to be considered in certain cases.

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References