J Anesth (2010) 24:256–259
DOI 10.1007/s00540-010-0875-8

CLINICAL REPORT

General anesthesia in a patient with Parkes Weber syndrome with high-output cardiac failure due to multiple arteriovenous fistulas complicated by severe aortic regurgitation

Jun Ninagawa · Yoshitsugu Yamada

Received: 31 March 2009 / Accepted: 19 November 2009 / Published online: 6 February 2010

Abstract Parkes Weber syndrome is a rare disease characterized by overgrowth of an extremity linked to the presence of an arteriovenous malformation with multiple arteriovenous fistulas (AVFs). We report a patient with Parkes Weber syndrome with high-output cardiac failure due to multiple AVFs complicated by severe aortic regurgitation (AR) who required surgical treatment for AVFs. Division of the left deep femoral artery and banding of the left superficial femoral artery were performed. Such procedures can cause aggravation of AR and left ventricular failure due to the sudden increase in cardiac afterload. Pulmonary artery pressure, mixed venous oxygen saturation and cardiac index monitored by a thermodilution catheter, and a transesophageal echocardiography were useful in evaluating the effect of the surgical procedure and resultant acute increase in cardiac afterload on cardiac output and left ventricular function.

Keywords Aortic regurgitation · Parkes Weber syndrome

Introduction

Parkes Weber syndrome is a rare disease characterized by overgrowth of an extremity linked to the presence of an arteriovenous malformation (AVM) with multiple arteriovenous fistulas (AVFs) [1]. These AVFs can cause high-output cardiac failure [1, 2], requiring surgical treatment for AVFs. We present a patient with Parkes Weber syndrome with high-output cardiac failure complicated by severe aortic regurgitation (AR) who underwent surgical treatment for AVFs.

J. Ninagawa (✉) · Y. Yamada
Department of Anesthesiology, University of Tokyo Hospital, 7-3-1 Hongo, Bunkyo-ku, Tokyo, Japan
e-mail: ninagawa-tky@umin.ac.jp

Case report

A 68-year-old woman (48 kg, 156 cm) was scheduled for surgical treatment of AVFs. The patient had been diagnosed with Parkes Weber syndrome based on the asymmetrical enlargement of her left leg, with venous dilation and AVFs. She had had cardiac failure symptoms (NYHA classification II) for 2 years. An electrocardiogram showed atrial fibrillation and bigeminal pulse of premature ventricular contraction. An echocardiography revealed severe AR, mild mitral regurgitation, decreased left ventricular function (ejection fraction 47%), and dilatation of the left ventricle (end-diastolic dimension 70 mm, end-systolic dimension 53 mm). The AR jet was central with no aortic cusp prolapse. A three-dimensional computed tomography (CT) angiography showed an AVF located between the perforating branch of the left deep femoral artery (DFA) and its accompanying vein and some AVFs near the popliteal artery. The arteries from the left common iliac artery to the popliteal artery were tortuous and dilated to 47 mm in diameter (Fig. 1). The CT scan showed dilatation of the ascending aorta (diameter 57 mm), an infrarenal abdominal aortic aneurysm (diameter 47 mm), and dilatation of the main pulmonary artery (diameter 64 mm) (Fig. 2). Preoperative examination demonstrated a pulmonary artery pressure (PAP) of 43/15 mmHg (mean PAP 23 mmHg), a pulmonary capillary wedge pressure of 12 mmHg, and a Fick cardiac index (CI) of 5.1 l/min/m^2.

To minimize high-output cardiac failure, we scheduled division of the left DFA, which fed the largest AVF. This procedure, however, could potentially induce an increase in

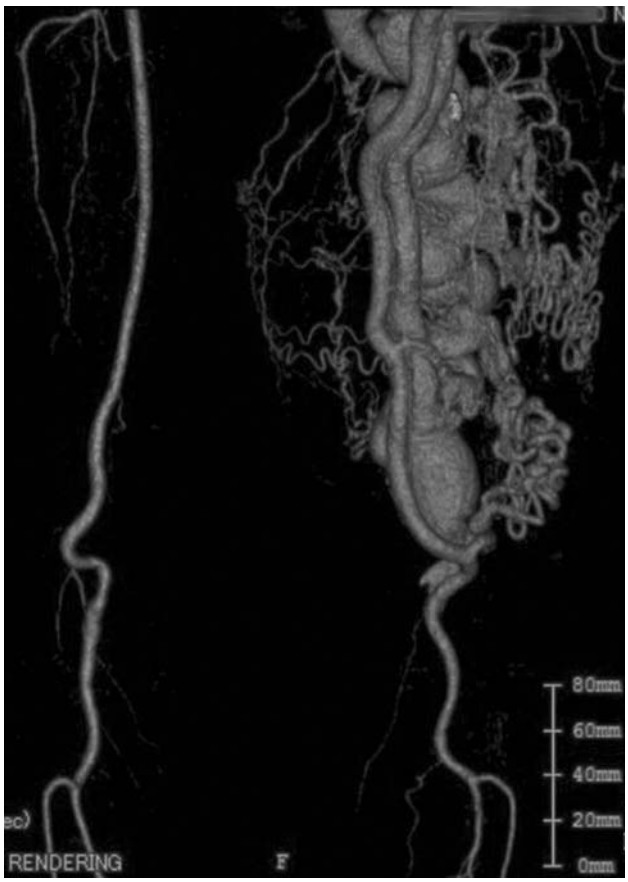

Fig. 1 Computed tomography angiography of the lower extremities. Multiple arteriovenous fistulas and varices are observable in the left thigh

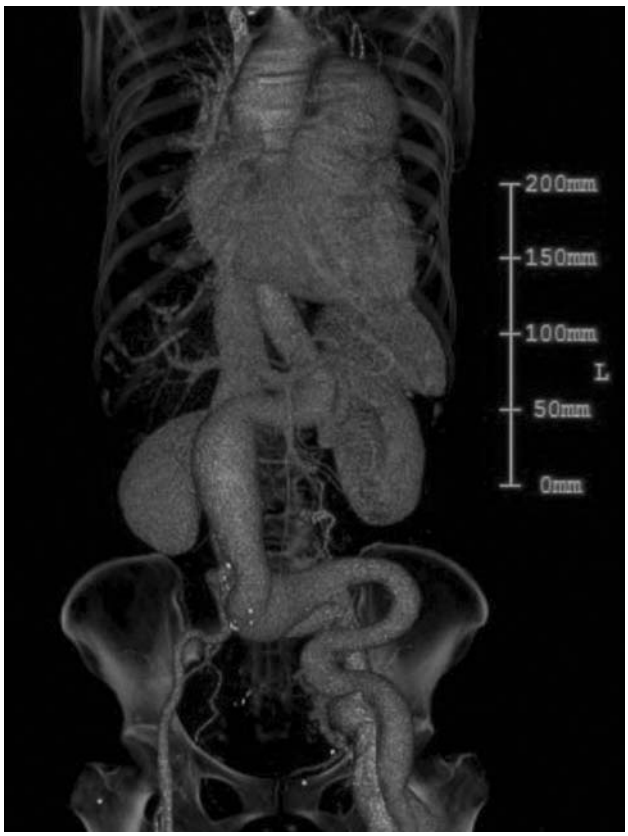

Fig. 2 Computed tomography angiography of the trunk. Dilatations of the ascending aorta, abdominal aorta, and pulmonary artery are present. The abdominal aorta is severely tortuous below the renal arteries. The arteries from the left common iliac artery to the left popliteal artery are also tortuous and dilated

systemic vascular resistance (SVR) resulting in aggravation of the AR and worsening cardiac failure. If this were to occur, the Bentall procedure was to be performed. Hence, during the surgery for dividing the left DFA, cardiac surgeons stood by in the operating room and cardiopulmonary bypass equipment was kept ready for use.

Anesthesia was induced with 4 mg midazolam and 200 μg fentanyl. Tracheal intubation was facilitated with 6 mg vecuronium. Anesthesia was maintained with 1–2% sevoflurane and fentanyl (total 300 μg) under 60% oxygen with air. In addition to monitoring the standard parameters (noninvasive blood pressure, electrocardiogram, and pulse oximetry), we also monitored arterial pressure, central venous pressure (CVP), PAP, and continuous cardiac output with a thermodilutional catheter. After the induction of general anesthesia, an arterial line was inserted into the left radial artery, a central venous catheter into the right internal jugular vein, and a pulmonary artery catheter via the right internal jugular vein. A transesophageal echocardiography (TEE) was performed to evaluate AR and cardiac function. The TEE after the induction of general anesthesia showed decreased global left ventricular function (ejection fraction 44%), as shown preoperatively, the AR jet extending to the head of the papillary muscle of the left ventricle, and holodiastolic flow reversal in the proximal abdominal aorta. The ratio of AR jet width to left ventricular outflow tract (LVOT) diameter was 71% (AR jet 17 mm, LVOT 24 mm), indicating severe AR.

Following isolation of the left femoral artery, trial clamping of the left femoral artery was performed. If clamping resulted in aggravation of the AR, as shown by the TEE, or in worsening of the left ventricular function so that catecholamine infusion was necessary, the Bentall procedure would also be performed. Dopamine and milrinone were readily available. After the test clamp of the left femoral artery was in place, radial arterial pressure changed from 86/49 (mean 65) to 102/53 (72) mmHg, the CVP from 10 to 8 mmHg, PAP from 36/15 (23) to 31/12 (19) mmHg, mixed venous oxygen saturation ($S\bar{v}O_2$) from 93 to 86%, and CI from 6.1 to 3.3 l/min/m^2. The systemic vascular resistance index (SVRI), which is calculated using the equation 80 × (mean arterial pressure − right atrial pressure)/CI, increased from 721 to 1552 dynes s m^2/cm^5. There was no remarkable change in the TEE in terms of the

degree of AR and left ventricular function, and no catecholamine infusion was needed. Hence, we were able to perform the left DFA division without the Bentall procedure. After the left DFA had been divided, the left femoral artery was declamped. After this declamping, radial arterial pressure decreased from 113/60 (81) to 81/50 (65) mmHg, the CVP remained unchanged at 8 mmHg, the PAP decreased from 31/14 (21) to 29/12 (19) mmHg, $S\bar{v}O_2$ increased from 85 to 90%, and CI from 3.3 to 4.2 l/min/m^2. In order to decrease the blood flow through the left superficial femoral artery (SFA), which fed the other AVFs near the popliteal artery, we banded the left SFA. Another arterial line was inserted into the left dorsalis pedis artery, and the pressure of the left dorsalis pedis artery was monitored as an index of the degree of banding. With banding of the left SFA, the left dorsalis pedis pressure changed from 116/40 (56) to 97/42 (58) mmHg, while the radial artery pressure, CVP, PAP, and $S\bar{v}O_2$ did not change appreciably. At the end of surgery, The CI was 4.7 l/min/m^2 and the SVRI was 1310 dynes s m^2/cm^5.

Anesthetic duration was 4 h and 49 min. Intraoperative blood loss was 140 ml, crystalloid infusion was 2780 ml, and urine volume was 1340 ml. No continuous cathecolamine infusion was needed intraoperatively. The postoperative course was uneventful, and the patient was discharged on the 13th postoperative day. Postoperative echocardiography showed normal left ventricular function (ejection fraction 58%) and moderate AR. The bigeminal pulse of premature ventricular contraction disappeared. She has shown no symptoms of worsening of cardiac failure after discharge.

Discussion

Parkes Weber syndrome is an extremely rare condition defined by overgrowth of an extremity linked to the presence of an AVM with multiple AVFs [1]. Revencu et al. [3] recently reported that Parkes Weber syndrome with multifocal capillary malformations, previously considered to be sporadic and nongenetic, is caused by de novo or inherited RASA1 mutations. To the best of our knowledge, there has been no report of the anesthetic management of patients with Parkes Weber syndrome.

Parkes Weber syndrome is known to cause high-output cardiac failure due to multiple AVFs [1]. Treatment of a large AVF by embolization and ligation results in a sudden increase in SVR. The arterial blood pressure can rise significantly and the CVP and PAP can decrease, signifying an improvement in the high-output cardiac failure. However, the sudden increase in afterload can also add to the left ventricular strain and failure, especially in patients with ventricular dysfunction. In those patients with high-output cardiac failure who undergo surgery for AVFs, a thermodilution catheter may be useful in the diagnosis and management of intraoperative left ventricular decompensation due to the sudden increase in afterload and in evaluating the effect of the surgical procedure on cardiac output [4, 5], as shown in the case presented here. In our patient, arterial blood pressure rose, and CVP and PAP decreased with no signs of worsening of cardiac failure. The most significant quantitative effects of the surgical procedure were a decrease in $S\bar{v}O_2$ and CI. Severe AR, as in this patient, can also be a cause of congestive cardiac failure after a sudden increase in afterload. In our case, a TEE evaluation was useful for differentiating the exact cause of cardiac decompensation. After division of the left DFA and banding of the left SFA, the arterial blood pressure gradually decreased, and the CVP, PAP, $S\bar{v}O_2$ and CI gradually increased. These findings indicated the possibility that blood flow through the residual AVFs had increased.

Parkes Weber syndrome can sometimes affect multiple limbs, the head, and the trunk [6]. In our patient, vascular malformations extended from the left lower limb to the aorta. The ascending aorta was dilated together with enlargement of the sinotubular junction (diameter 36 mm). This condition was considered to be the cause of her severe AR, allowing incomplete coadaptation of aortic leaflets. Severe AR can be an independent cause of congestive cardiac failure that requires surgical correction. In addition, treatment for AVF in these patients can suddenly increase SVR and worsen AR. CPB in patients with multiple AVFs generating a significant AV shunt is associated with the risk of hypotension during and after CPB, with this hypotension being refractory to vasoconstrictor therapy [7]. Therefore, the indications of surgery requiring CPB should be carefully determined in these patients. We decided that the Bentall procedure would be performed if the AR and cardiac failure worsened with division of the left DFA. Fortunately, the TEE showed no worsening of AR after division of the left DFA, and the Bentall procedure was not required.

In summary, our case of Parkes Weber syndrome with multiple AVFs that induced high-output cardiac failure and with severe AR required surgery for AVFs. The Bentall procedure was to be added if the AR worsened with surgical treatment of AVFs, with the procedure being scheduled with cardiopulmonary bypass and cardiac surgeons on stand-by. Division of the left DFA and banding of the left SFA improved cardiac failure without worsening AR. $S\bar{v}O_2$ and CI monitored by a thermodilution catheter combined with TEE evaluation were useful in evaluating the effect of the surgical procedure on cardiac output, changes in left ventricular function, and the degree of AR resulting from the sudden increase in afterload.

References

1. Garzon MC, Huang JT, Enjolras O, Frieden IJ. Vascular malformations Part II: associated syndromes. J Am Acad Dermatol. 2007;56:541–64.
2. Berger TM, Caduff JH. Hemodynamic observations in a newborn with Parkes–Weber syndrome. J Pediatr. 1999;134:513.
3. Revencu N, Boon LM, Mulliken JB, Enjolras O, Cordisco MR, Burrow PE, et al. Parkes Weber syndrome, vein of Galen aneurysmal malformation, and other fast-flow vascular anomalies are caused by RASA1 mutations. Hum Mutat. 2008;29:959–65.
4. Neema PK, Ramakrishnan S, Sinha PK, Rathod RC. Anesthetic implication of surgical repair of an aortocaval fistula. J Cardiothorac Vasc Anesth. 2003;17:236–9.
5. Sharma ML, George KA, Gamble JAS. Anaesthetic implications of endovascular repair of aortocaval fistula. Anaesthesia. 2000; 55:697.
6. Garcia FR, Gonzalez RS, Gonzalez EF, Cabrera de Paz R, Martin MR, Martin-Neda F, et al. Klippel Trenaunay–Weber syndrome: a long term study of a singular case. Pediatr Dermatol. 2004;21:397–8.
7. Radu C, Reich DL, Tamman R. Anesthetic considerations in a cardiac surgical patient with Osler–Weber–Rendu disease. J Cardiothorac Vasc Anesth. 1992;6:461–4.