consequence of a blunt trauma has been reported [3, 4]. A rupture of the proximal RCA into the right atrium was also reported by Trotter and associates [5]. However, these reports refer neither to an ostium injury nor to fatal tamponade because of coronary artery rupture, as we have reported here. We report of a patient surviving traumatic free rupture of the coronary artery causing cardiac tamponade.

Because of other potentially fatal hemorrhagic lesions, including intracranial hemorrhage, systemic heparinization for CPB could be lethal in a multifocal blunt trauma patient. Thus, we applied buttressed suture hemostasis without identifying the exact bleeding site at the risk of acute myocardial infarction. Temporary cardiac arrest or ventricular fibrillation induced pharmacologically or electrically could have been applied for the identification of the bleeding origin. However, these tools could not provide sufficient time to repair the lesion definitively without CPB, even when RCA tear was identified. Thus, an attempt at using a buttressed suture involving the surrounding tissue was justified.

Although the initial hemostasis was successful without any infarction, a large pseudoaneurysm remained. This condition absolutely indicated the second surgery. Although Hwang and colleagues [6] reported that traumatic coronary aneurysm could conservatively be treated, the lesion they described differed from our patient in that it was not a pseudoaneurysm but a true aneurysm.

In summary, successful treatment of fatal cardiac tamponade due to RCA ostium rupture is reported, in which an initial buttressed suture hemostasis and a delayed definitive repair for a pseudoaneurysm formation under CPB were performed. Cardiac surgery necessitating systemic heparinization should be delayed until after treatment of other potentially life-threatening hemorrhagic lesions in a multifocal blunt trauma patient. Our treatment strategy was thus appropriate from the standpoint of initial damage control.

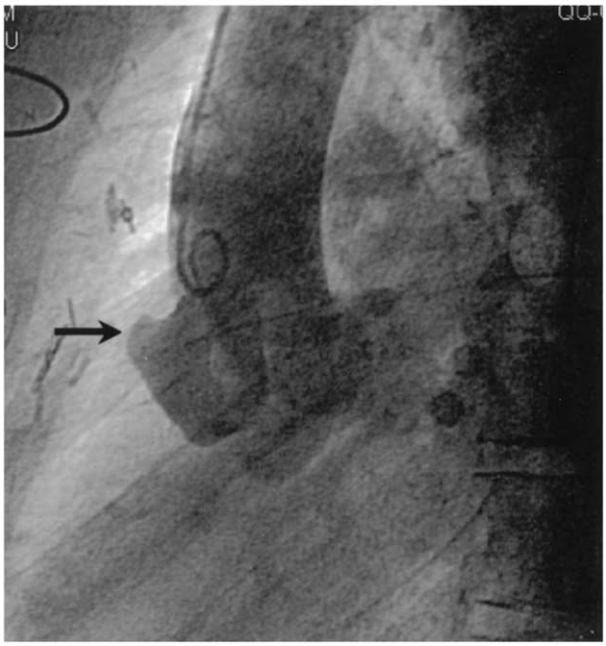

*Fig 3. Postoperative aortography shows the right coronary artery ostium (*arrow*) closed with patch.*

References

1. Fulda G, Brathwaite CEM, Rodriguez A, Turney SZ, Dunham CM, Cowley RA. Blunt traumatic rupture of the heart and pericardium: a ten-year experience (1979–1989). J Trauma 1991;31:167–73.
2. Brathwaite CEM, Rodriguez A, Turney SZ, Dunham CM, Cowley RA. Blunt traumatic cardiac rupture—A 5-year experience. Ann Surg 1990;212:701–4.
3. Reiss J, Razzouk AJ, Kiev J, Bansal R, Baily LL. Concomitant traumatic coronary artery and tricuspid valve injury: a heterogenous presentation. J Trauma 2001;50:942–4.
4. Pifarre R, Grieco J, Garibaldi A, Sullivan HJ, Montoya A, Bakhos M. Acute coronary artery occlusion secondary to blunt chest trauma. J Thorac Cardiovac Surg 1982;83:122–5.
5. Trotter TH, Knott-Kraig CJ, Ward KE. Blunt injury rupture of tricuspid valve and right coronary artery. Ann Thorac Surg 1998;66:1814–6.
6. Hwang SO, Park KS, Lee KH, Yoon J, Ha JW, Choe KH. Three-year follow-up of a posttraumatic right coronary aneurysm. J Trauma 1997;43:859–61.

CASE REPORTS

Aortic Valve Replacement in a Patient With Erythropoietic Protoporphyria

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Erythropoietic protoporphyria (EPP) is a disorder of heme synthesis that causes excessive accumulation of protoporphyrin. The predominant clinical feature is photosensitivity triggered by light at wavelengths near 400 nm. We describe a 52-year-old man with EPP who underwent aortic valve replacement due to severe regurgitation. To prevent burn injuries, astral lamps in the operating room were covered with yellow film filters. Preoperative autologous blood donation was not undertaken. Blood priming of the extracorporeal circuit was performed to maintain adequate hemoglobin concentrations, which resulted in reduction of heme synthesis. The patient was discharged in good health without any signs or symptoms of EPP.

(Ann Thorac Surg 2003;75:1003–5)

Accepted for publication Aug 24, 2002.

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Published by Elsevier Science Inc

0003-4975/03/$30.00
PII S0003-4975(02)04366-7

Ann Thorac Surg
2003;75:1003–5

Erythropoietic protoporphyria (EPP) is an autosomal dominant inherited disorder of heme metabolism caused by the partial deficiency of ferrochelatase, which is the terminal enzyme of heme synthesis [1]. Protoporphyrin (PP) accumulates in erythrocytes and plasma and is deposited in the skin. Its clinical expression results in a wide range of photocutaneous changes and occasionally liver dysfunction. The activating wavelengths of the light are around 400 nm (i.e., the blue spectrum), which correspond to the absorption spectra for porphyrins [1]. Light-excited porphyrins generate free radicals and single atoms of oxygen that lead to cell damage.

There is only one report of cardiac surgery in a patient with EPP [2]. We report here the case of a patient with EPP who underwent successful aortic valve replacement and discuss the perioperative considerations of this disease.

A 52-year-old male was admitted to our hospital for aortic valve surgery. He had a history of sensitivity to light since childhood, resulting in burning of the face and arms with occasional transient skin rash. His condition was diagnosed as EPP. The patient had undergone a hemorrhoidectomy under spinal anesthesia without complication 10 years previously.

Physical examination revealed a regular pulse of 64 bpm, a blood pressure of 138/58 mm Hg, and an early diastolic grade II/VI murmur in the aortic area. Neither hepatosplenomegaly nor skin lesions were noted. Electrocardiogram exhibited sinus rhythm with left ventricular hypertrophy. Chest Roentgenogram revealed cardiac enlargement with a cardiothoracic ratio of 60%.

Echocardiography revealed severe aortic regurgitation. There was no dilatation of the ascending aorta, sinus of Valsalva, or the aortic annulus. Left ventricular end-diastolic dimension was 68 mm. Fractional shortening was 32% and ejection fraction was 58%. Aortography revealed grade IV/IV aortic regurgitation. Cardiac index was 2.84 $L \cdot min^{-1} \cdot m^{-2}$. Left ventricular systolic pressure was 160 mm Hg and end-diastolic pressure was 33 mm Hg.

Laboratory data were unremarkable except for a red blood cell PP value of 2544 μg/dL (normal: 30 to 86) and a normal value for urinary coproporphyrin of less than 1 μg/dL.

On the day of surgery, premedication consisted of intravenous 10-mg morphine. To prevent burn injuries, all the lights in the operating room were switched off except for the astral lamps, which were covered with yellow film filters. This filter cuts off light waves at a wavelength of 500 nm and has a very high transmission for wavelengths above 550 nm. Anesthesia was induced and maintained with propofol and fentanyl infusion. Muscle relaxation was achieved with vecuronium.

Following median sternotomy and heparinization, cardiopulmonary bypass was initiated with blood pump prime, and the hemoglobin level was maintained at greater than 9 g/dL to reduce heme synthesis and, thereby, the PP level. For the same reason, preoperative autologous donation of blood was not undertaken. Eight units of allogeneic red blood cell concentrate were infused during the operation resulting in a postoperative hemoglobin level of 11 g/dL. The patient was cooled to 32°C and the mean arterial pressure was maintained at 60 mm Hg. Myocardial protection was provided by crystalloid cardioplegia and maintained by intermittent blood cardioplegia given in an antegrade fashion.

The aortic valve leaflets were relatively thin and their movement was not restricted; however, the free edge of the right coronary cusp was redundant and prolapsed. The valve was replaced by a 25-mm St. Jude Medical mechanical heart valve (St. Jude Medical, Inc., St. Paul, MN). Histologic study of the excised valve disclosed myxoid degeneration.

Discontinuation of cardiopulmonary bypass was uneventful, and heparin was reversed by protamine. Recovery was conducted in a darkened room in the intensive care unit. The patient regained consciousness 3-hours after completion of surgery and was extubated uneventfully 1-hour later.

The patient's postoperative course was unremarkable except for an episode of abnormal liver tests. His total bilirubin level increased to 6.0 mg/dL with a direct bilirubin of 2.5 mg/dL on the second postoperative day and thereafter approached normal. At the same time, serum alanine aminotransferase was elevated to a maximum of 153 IU/L, serum aspartate aminotransferase to 219 IU/L, and serum lactic dehydrogenase to 1025 IU/L. Hemoglobin level dropped from 11 g/dL to 9.6 g/dL, indicating possible hemolysis. The red blood cell PP level fell from 1450 μg/dL before cardiopulmonary bypass to 1057 μg/dL at the end of surgery, and then increased to 2430 μg/dL 1-week later. The patient was discharged on the 14th postoperative day in good health with no clinical signs or symptoms of EPP.

Comment

Major surgery in a patient with EPP requires careful perioperative management to prevent further induction of porphyrin production and exacerbation of the disease. Because maintenance of adequate hemoglobin concentrations is important to reduce heme synthesis and minimize PP levels [2], preoperative autologous blood donation was not undertaken. Blood priming of the extracorporeal circuit was performed to avoid anemia.

Artificial lights, including those in an operating theater, can cause severe photosensitivity. Eleven cases of EPP, reviewed by Torres and associates [3], were treated by liver transplantation. Extreme photosensitivity developed in 6 patients, resulting in severe burn injuries to the abdominal wall, intestinal ulceration and hemolytic anemia, and death in 2 patients. In these cases, the hazardous effects of operating room lights on the exposed splanchnic bed were not well recognized, and no preventive measures were taken. Meerman and coworkers [4] reported two successful liver transplantations in patients

with EPP using acrylate yellow filters to filter operating room lights, and stressed that treatment of EPP patients with cholestatic liver disease should involve the use of yellow filters because these patients have the highest tissue levels of PP. Recently, Asokumar and colleagues [2] reported on the successful closure of a ventricular septal defect in which yellow acrylate filters were used to cover the operating room lights. In our report, the patient might not have suffered any burns during surgery without the use of the filters because his preoperative PP level was not extremely high and hemorrhoidectomy had been performed previously under unfiltered operating lamps without complication. However, when considering such catastrophic possibilities as cardiac rupture or coronary stenosis caused by burn injury, we thought it an appropriate precautionary measure to cover the astral lamps with yellow filters to eliminate blue light. In the absence of blue light, blue objects are generally perceived as dark or black, and other colors have a yellow cast. This altered color perception; however, it did not significantly impede the surgeons, anesthesiologists, or the nurses.

Erythropoietic porphyrias, unlike hepatic porphyrias, lack neurologic involvement and are not fundamentally associated with drug-precipitated crises [1]. A few patients with end-stage EPP liver disease, however, are reported to have developed a syndrome that resembled the neurologic crises of the acute porphyrias [5, 6]. This syndrome is characterized by abdominal pain, hypertension, tachycardia, extremity pain and weakness, constipation, and nausea and vomiting. PP may infiltrate neural tissue when serum levels are markedly increased, causing neurotoxicity. However, most EPP patients with low PP levels, including our patient, do not require special consideration regarding drugs that trigger acute porphyric crisis. Nevertheless, we chose to use morphine, fentanyl, propofol, and vecuronium, which have not been associated with initiation of porphyric crisis. Neurologic symptoms were not seen in our patient.

References

1. Gawkrodger DJ, Porphyrias. In Champion RH, Burton JL, Burns DA, Breathnach SM, eds. Textbook of dermatology, 6th ed. Oxford: Blackwell Science, 1998:2577-2600.
2. Asokumar B, Kierney C, James TW, Amato J, Tuman KJ. Anesthetic management of a patient with erythropoietic protoporphyria for ventricular septal defect closure. Paediatr Anaesth 1999;9:356–358.
3. de Torres I, Demetris AJ, Randhawa PS. Recurrent hepatic allograft injury in erythropoietic protoporphyria. Transplantation 1996;61:1412–1413.
4. Meerman L, Verwer R, Slooff MJH. Perioperative measures during liver transplantation for erythropoietic protoporphyria. Transplantation 1994;57:155–158.
5. Herbert A, Corbin D, Williams A, Thompson D, Buckels J, Elias E. Erythropoietic protoporphyria: unusual skin and neurological problems after liver transplantation. Gastroenterology 1991;100:1753–1757.
6. Rank JM, Carithers R, Bloomer J. Evidence for neurological dysfunction in end-stage protoporphyric liver disease. Hepatology 1993;18:1404–1409.

Mechanical Ventricular Support Lowers Pulmonary Vascular Resistance in a Patient With Congential Heart Disease

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CASE REPORTS

Severely elevated pulmonary vascular resistance (PVR) is a relative contraindication to orthotopic heart transplantation. A potential novel strategy to reverse elevated PVR may be implantation of a chronic left ventricular assist device with subsequent left ventricular unloading. We present a patient with elevated PVR secondary to congenital heart disease who was listed for heart-lung transplant. The patient underwent placement of biventricular assist devices and subsequently experienced marked reduction of PVR, ultimately enabling successful heart transplantation.

(Ann Thorac Surg 2003;75:1005–7)

Severely elevated pulmonary vascular resistance (PVR) is a relative contraindication to orthotopic heart transplantation. Studies have reported an increase in operative mortality with perioperative donor organ failure in patients transplanted with severely elevated PVR [1]. Therefore, evaluation of potential heart transplant candidates includes the determination of PVR. If PVR is elevated, its response to acute interventions, such as nitric oxide, oxygen, or prostacyclin, are used to define reversibility. A patient whose PVR is markedly elevated ($>$ 7.0 Wood units) and unresponsive to pulmonary vasodilators ($<$ 20% change) generally is not considered for heart transplant and instead may be evaluated for heart-lung transplant. When PVR remains refractory to pulmonary vasodilators, a potential novel strategy to reverse elevated PVR may be implantation of a chronic left ventricular assist device (LVAD) with subsequent left ventricular unloading [2].

Patients with congenital heart disease and left-to-right shunts generally develop increased PVR associated with anatomic pulmonary artery changes. They may be less likely to respond to vasodilator therapy or mechanical support relative to patients with acquired left ventricular

Accepted for publication Aug 27, 2002.

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Published by Elsevier Science Inc

0003-4975/03/$30.00
PII S0003-4975(02)04372-2