Bioprosthetic Pulmonary and Tricuspid Valve Replacement in Carcinoid Heart Disease From Ovarian Primary Cancer

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Carcinoid tumors usually originate in the gastrointestinal tract, but in rare instances they may arise in other organs. A patient with severe tricuspid and pulmonary regurgitation because of carcinoid syndrome successfully underwent double valve replacement using bioprostheses. The patient was finally diagnosed with carcinoid heart disease from an isolated ovarian carcinoid cancer. The diagnosis of carcinoid syndrome should be recognized as an etiology in patients with organic tricuspid and pulmonary regurgitation without left valvular disease. (Circ J 2009; 73: 1554–1556)

Key Words: Fibrosis; Plaque; Prosthesis; Tumor; Valvular diseases

Case Report

A 52-year old woman was hospitalized because of right heart failure. On admission she complained of increasing fatigue and dyspnea. Physical examination showed peripheral edema and hepatomegaly. Two-dimensional and Doppler echocardiography revealed severe tricuspid and pulmonary regurgitation. The pulmonary and tricuspid valves were thickened and immobile. The right ventricle was enlarged because of volume overload. There was no abnormal finding in the left heart. Cardiac catheterization showed a right atrial pressure of 18 mmHg, a systolic right ventricular pressure of 39 mmHg, mean pulmonary pressure of 18 mmHg, and a pulmonary capillary wedge pressure of 9 mmHg. Complete blood count, serum electrolyte levels, and liver enzymes were normal. Because of the severity of the cardiac symptoms, she was referred to surgery.

She had been involved in a car accident 1 year ago and had sustained trauma to the anterior chest. Echocardiographic examination performed 2 years ago showed no evidence of valvular disease, and she had noticed the clinical symptoms after the traffic accident. Therefore, preoperatively we thought the pulmonary and tricuspid regurgitation might be traumatic in origin.

Cardiopulmonary bypass was initiated through a standard median sternotomy and arterial and bivacal cannulation. The pulmonary valve was inspected through a horizontal incision into the main pulmonary artery. The cusps were thickened and retracted, so they were excised followed by pulmonary valve replacement with a stentless bioprosthesis (Prima plus, Edwards Lifesciences, Irvine, CA, USA) as the root. The tricuspid valve was exposed through a right atriotomy. Its leaflets were visibly fibrotic, thickened, and retracted. After failed tricuspid annuloplasty, the valve was replaced by a size 27 mitral porcine heart valve (Mosaic, Medtronic Inc, Minneapolis, MN, USA) leaving the leaflet tissue (Figures 1A, B). The patient was successfully weaned from bypass, and her postoperative course was uneventful. Transthoracic echocardiography at

Figure 1. (A) Intraoperative view of the pulmonary valve shows it to be remarkably thickened. (B) The tricuspid valve is also markedly thickened and retracted.
discharge showed normal function of both bioprostheses and the mean gradient over the tricuspid and pulmonary valve prostheses was 12 mmHg and 4 mmHg, respectively. Histopathologic evaluation of the excised pulmonary valves revealed a clear demarcation between the normal valvular tissue and the upper carcinoid-related fibrous plaques deposition (Figure 2A).

After surgery, she was diagnosed as having carcinoid heart disease because the urinary 5-HIAA level was 117 mg/24 h (normal, 0–5 mg/24 h). Abdominal computed tomography showed the primary tumor in the right ovary, and no metastasis to the liver. The patient was finally diagnosed with carcinoid heart disease from an isolated ovarian carcinoid tumor without hepatic metastases.

Six months after cardiac operation, a laparotomy confirmed the right ovarian mass and total hysterectomy and bilateral salpingo-oophorectomy was performed. The pathological examination revealed that the right ovarian mass was a carcinoid tumor, insular type, measuring 10 cm (Figure 2B). Her urinary 5-HIAA level fell to 1.4 mg/24 h after the laparotomy.

Discussion

Carcinoid heart disease associated with a primary ovarian carcinoid tumor is extremely rare. Although carcinoid syndrome prevalence is approximately 2–5% among patients with carcinoid tumors, which has a reported incidence of 1.5 per 100,000 population, the incidence of carcinoid syndrome rises to 30% in primary ovarian carcinoid tumors.1-2 This difference is explained by direct release of serotonin-like substances into the systemic circulation via the ovarian venous system, bypassing deactivation by hepatic metabolism. Carcinoid heart disease is reported to be detected clinically in 25% of patients with carcinoid syndrome.1-3

After double valve replacement, she was diagnosed as having carcinoid heart disease from an isolated ovarian carcinoid tumor without hepatic metastases. The duration from onset of symptoms to the diagnosis of carcinoid heart disease is usually 2 years, but longer intervals of up to 5 years have been reported. The diagnosis may be delayed because symptoms, such as facial flushing, are often absent or subtle in the early stage.

Furthermore, the presence or absence of symptoms of carcinoid syndrome is also dependent on the number of secreting tumor cells. There is a good correlation between the size of the tumor and the presence of clinically detectable carcinoid syndrome; in particular, all reported functioning ovarian carcinoid tumors have measured approximately 10 cm in diameter, whereas intestinal carcinoids are usually smaller.5

A recent surgical pathology series from the Mayo Clinic described 139 valves excised from 75 patients with carcinoid heart.8 The carcinoid plaque consisted of a cellular component (myofibroblasts) and an extracellular component (collagen, myxoid matrix, and elastin). Other features included neovascularization, chronic inflammation, and mast cells. Severe fibrosis, with severe collagenization, primarily affected tricuspid valve plaque, whereas severe thickening of the pulmonary valves was principally caused by proliferation of myofibroblasts and deposition of myxoid matrix.

In the present case, pulmonary and tricuspid valve replacement was performed. Connolly et al reported 22 patients with pulmonary and tricuspid valve regurgitation and suggested that tricuspid and pulmonary valve replacement should be considered when both valves are involved, because the double valve replacement has a beneficial effect on right ventricular size compared with isolated tricuspid valve replacement.7

As for the selection of the prostheses, we used xenografts for both right-sided valves. There are 2 reports of secondary fibrous carcinoid involvement of tissue valves that were used in the tricuspid or pulmonary position in patients with carcinoid syndrome.8-9 Early reports recommended using a mechanical prosthesis, based on the assumption that circulating vasoactive tumor substances may damage bioprosthetic valves.8 The potential danger of involvement of the bioprosthetic cusps in the carcinoid process may be prevented by surgical therapy against the primary carcinoid tumor. Mechanical prostheses are not ideal because of the life-long anticoagulation, and the risk of thrombosis in mechanical tricuspid prostheses is 4% per year? Furthermore, a stentless bioprosthesis is preferred in the pulmonary position because its excellent hemodynamic characteristics and flexibility. However, follow-up including echocardiography and computed tomography is mandatory.

We report a patient with severe tricuspid and pulmonary regurgitation because of carcinoid syndrome who successfully underwent double bioprosthetic valve replacement. The diagnosis of carcinoid syndrome should be recognized as an etiology in patients with organic tricuspid and pulmonary regurgitation without left valvular disease.
References