

Quadricuspid aortic valve with D (Rh₀) antigen negativity: a novel case report

To the Editor,

Here we report a rare case of a Chinese female patient presenting with aortic insufficiency due to a quadricuspid aortic valve and Rho negativity. The 64-year-old woman was referred with a 1-month history of dyspnea and cough (NYHA class II). A quadricuspid aortic valve was suspected, and grade 3 aortic regurgitation was identified by transthoracic echocardiography (TTE). After admission, the ABO blood type was surprisingly identified as AB positive, and results of the D (Rho) antigen test were negative. Considering her clear diagnosis and indications for aortic valve replacement, surgical intervention was the best choice to resolve the aortic insufficiency and relieve the symptoms. Due to her rare blood type, the surgery was postponed by a week. The patient successfully underwent elective aortic valve replacement with a mechanical prosthesis at her own will. In addition, no blood transfusion was arranged perioperatively. The symptoms vanished and the patient was discharged with a contented condition on the 7th postoperative day.

A quadricuspid aortic valve is a rare manifestation of congenital aortic valve abnormalities. The incidence significantly varies according to different reports. Hurwitz et al. (1) reported an incidence of only two cases in 6000 autopsies, while the Mayo Clinic noted an incidence of 1% in a review of 225 patients undergoing surgery for pure aortic regurgitation (2). The most common complication of a quadricuspid valve is pure insufficiency, while other common complications of

a quadricuspid valve are coronary anomalies and aortic root dilation (3). In this case, no anomalous origin of coronary arteries and aortic root dilation was found in both TEE and intraoperative findings.

The prevalence of Rho negativity is variable in different areas. The frequency of Rho negativity varies from 20% to 40% in Basques (4), while less than 0.3% population has been found to be D (Rho) negative in China (5). In addition, less than 10% of the entire Rho-negative population is AB positive (4). Therefore, the AB-positive and Rho-negative blood type is really rare in China. To the best of our knowledge, no case of a quadricuspid aortic valve with an AB-positive and Rho-negative blood type has been reported to date.

In summary, here we report, for the first time, a female presenting with a quadricuspid aortic valve with an AB-positive and Rho-negative blood type, who successfully underwent aortic valve replacement.

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