A 52-year-old woman, with a previous history of hypertension, was admitted to our institution presenting exertional dyspnea. A comprehensive study was performed including a cardiology examination and echocardiography. Transesophageal echocardiography detected a quadricuspid aortic valve with a central severe aortic valve insufficiency. Moreover, a discrete dilation of the aortic root was seen. Left ventricular ejection fraction was in normal range. A computed tomography scan was performed especially to recognize any displacement of the coronary ostia and evaluate the aortic valve (Fig. A). No abnormality of coronary artery origin was confirmed. Surgical treatment was proposed, and the patient underwent open-heart surgery. Quadricuspid valve was confirmed (Fig. B). An aortoplasty to reduce the ascending aorta and a mechanical aortic valve replacement were performed. The postoperative course was favorable, and the patient had an uneventful recovery. Postoperative echocardiography assessment confirmed normal prosthetic aortic valve function and left ventricular ejection fraction. Quadricuspid aortic valve is a rare congenital abnormality, with an estimated incidence of 0.04%. In approximately 80-90% of cases, it is related to an aortic valve dysfunction. Quadricuspid aortic valve is usually associated with additional malformations, mainly related to the origin of coronary arteries. Due to the presence of an additional leaflet, absence or displacement of coronary ostia has been found in 10% of cases. It is of utmost importance to prevent coronary ostia obstruction with valve prosthesis implantation, and to plan myocardial protection during cardiopulmonary bypass. In the presence of prior surgery for congenital aortic valve abnormalities and regardless of patient age, we highlight the necessity to perform a careful evaluation of the coronary anatomy.