A classic yet unusual case: the full spectrum of bicuspid aortic valve disease

Sushil A. Luis1,2*, Matthew Anderson2, Homayoun Jalali3, and Darryl J. Burstow1,2

1Department of Cardiology, The Prince Charles Hospital, Chermside, Queensland 4032, Australia; 2School of Medicine, University of Queensland, Herston, Queensland, Australia; and 3Department of Cardiothoracic Surgery, The Prince Charles Hospital, Chermside, Queensland, Australia

* Corresponding author. Tel: +61 731394000; Fax: +61 731394819, Email: sushil_luis@health.qld.gov.au

A 22-year-old male with no significant past medical history presented to the emergency department with sudden onset, sharp, severe central chest pain while drinking at the bar and smoking marijuana. He was markedly hypertensive with a blood pressure of 190/110. Admission chest X-ray demonstrated an absence of mediastinal widening, hypoplastic aortic knuckle, rib notching (arrowheads) and clear lung fields (Panel A). He was investigated with a CT pulmonary angiogram which showed no evidence of pulmonary embolism but demonstrated a large 7 cm ascending aortic aneurysm with intramural haematoma and hypoplastic descending aorta (Panel B). Transthoracic echocardiography demonstrated a large ascending aortic aneurysm measuring 74 mm at the aortic root with a dissection flap (white arrow) and significant thrombus in the false lumen (black arrow) (Panel C, Supplementary data online, Video 1). This was associated with a bicuspid aortic valve (Panel D), severe aortic regurgitation, and aortic coarctation (Panel E, Supplementary data online, Video 3). Pre-operative transoesophageal echocardiography confirmed these findings and demonstrated the presence of a severe discrete coarctation involving the descending thoracic aorta just distal to the left subclavian artery with a pinhole orifice (luminal diameter 1 mm) (Panel F, Supplementary data online, Video 3) and hypoplastic descending aorta distal to this measuring just 12 mm. He was taken emergently to theatre for an aortic valve and root replacement with bovine pericardial patch augmentation of the aortic arch and proximal descending aorta.

While unusual to find such concomitant pathologies in a single patient, the imaging illustrates the classical-associated features in patients with bicuspid aortic valves, including aortic coarctation and aortopathy with thoracic aortic aneurysm and dissection.

Supplementary data are available at European Heart Journal — Cardiovascular Imaging online.