Combined left ventricular non-compaction and Ebstein’s anomaly

A 49-year-old woman was admitted with acute cholecystitis. Significant history included a diagnosis of Ebstein’s anomaly at age 18 and congestive heart failure during two previous pregnancies. Cardiac examination revealed a murmur of tricuspid regurgitation. Transthoracic echocardiogram showed Ebstein’s anomaly with non-compaction of the left ventricle. The patient underwent laparoscopic cholecystectomy and had an uneventful postoperative course.

The transthoracic echocardiogram showed a thickened left ventricular apex with prominent trabeculations with normal function (figure 1). The septal tricuspid valve leaflet was apically displaced with mild tricuspid regurgitation. These findings were confirmed on cardiac MRI (figure 2).

The coexistence of Ebstein’s anomaly and non-compaction is being increasingly identified with the use of improved imaging techniques such as cardiac MRI and contrast echocardiography. Ebstein’s anomaly is defined as >8 mm/m² displacement of septal tricuspid leaflet with evidence of tricuspid insufficiency.

There may be tethering of the tricuspid valve, atrialisation of part of the right ventricle and right atrial enlargement.

In left ventricular non-compaction there is smooth, hypertrabeculated left ventricle with recesses and wall thickening. These changes may lead to an underfilled left ventricle and consequently raised left atrial pressures and pulmonary oedema. Non-compaction is associated with asymptomatic left ventricular dysfunction, arrhythmias, thromboembolic events and chest pain.

Ongoing management of such patients involves regular clinical assessment and assessment of left ventricular and tricuspid valve function. Where a doubt exists, there should be a low index of suspicion to use cardiac MRI to clearly define left ventricular architecture.

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