Case report

An unusual case of anomalous origin of the right coronary artery and hepatic focal nodular hyperplasia

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Learning Point for Clinicians

Anomalous origin of the right coronary artery (ARCA) and focal nodular hyperplasia (FNH) are frequently reported in association with congenital heart abnormalities but not with each other. We propose that both conditions may share common origins in a maladative hyperplastic response to differential vascular flow due to developmental arterial malformations or aberrant Notch signalling during simultaneous gut and cardiac vascularigenesis.

Case report

Coronary artery anomalies are reportedly associated with congenital heart abnormalities which may in turn be associated with FNH.1,2 However, there are no antecedent reports of coronary artery anomalies in direct association with FNH.

We present the case of an anomalous right coronary artery (RCA) originating from the left main coronary artery (LMCA) with an interarterial course coincident with FNH in a 40-year-old asymptomatic man, who presented to our institution following an abnormal treadmill electrocardiogram in a routine screening service. Apart from moderate dyslipidaemia, the history, physical examination and initial investigations were unremarkable. A gated computed tomography coronary angiogram (CT-A) with three-dimensional (3D) reconstruction revealed the anomalous RCA being compressed between the aorta (Ao) and the right ventricular outflow tract (RVOT) before eventually giving rise to a small posterior descending artery in a co-dominant system (Figure 1a). There was a non-obstructive calcified plaque in the distal LMCA with no other calcification or plaques in the other coronary arteries. CT-A also revealed an individual solitary hepatic (segment 8) lesion. Subsequent magnetic resonance imaging (MRI) showed the diagnostic features of FNH including a stable centrally sited iso-intense hepatic lesion on T1- and T2-weighted images, which was hypervascular in the early phase and more hyperintense in the later phases at 20 min and 1 h. A tiny central scar was faintly enhanced on later phases (Figure 1b).

The ARCA from the left coronary sinus coursing between the Ao and the pulmonary artery (PA) is a rare but malignant congenital anomaly often incidentally documented on cardiac imaging. Most individuals with ARCA are young and asymptomatic but at risk of acute coronary syndromes and sudden death at rest or with exercise and in the absence of coronary artery stenosis, with a significantly higher prevalence of major adverse cardiac events in ARCA with an interarterial course between the Ao and the PA than between the Ao and the RVOT.3

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This patient was commenced on statins and remains event-free to date with regular medical surveillance and advice against strenuous activities.

FNH is a benign hypervascular tumour-like lesion of well-circumscribed hyperplastic liver parenchyma often characterized with fibrous septae radiating from a central stellate scar. Patients are mostly asymptomatic with normal liver function, although portal hypertension can rarely develop with time. FNH is purportedly derived from a maladaptive hyperplastic parenchymal response to differential blood flow due to an underlying developmental arterial malformation. FNH is also associated with congenital heart abnormalities, which are frequently coincident with coronary artery anomalies.\(^1\) This may be attributed to the intimate relationship between vitelline veins and the heart during embryogenesis and may explain the association between cardiovascular malformations and vascular malformations within the hepatic sinusoidal microcirculation, in turn implicated in the pathogenesis of FNH.\(^1\) FNH and cardiovascular anomalies also share common aberrant molecular origins during development as Notch critically influences the vascular patterning of the primitive coronary network\(^4\) during cardiogenesis, as well as vascular remodelling and homoeostasis of the hepatic sinusoidal microcirculation.\(^5\) Any embryological insult during simultaneous gut and cardiac development may lead to congenital heart defects, anomalous coronary development and the development of focal hepatic lesions secondary to maladaptive hepatic vasculorigenesis. The frequent associations of congenital heart malformations with coronary artery anomalies and FNH, respectively, suggest that the association of ARCA and FNH in this patient may not be entirely incidental.

Conflict of interest: None declared.

References


Figure 1. (a) 3D heart reconstruction showing the origin of the anomalous RCA arising from the left coronary ostium. LAD, left anterior descending artery; LCx, left circumflex coronary artery; LMCA, Left main coronary artery, LA, left atrium. (b) MRI of the centrally sited hypervascular hepatic lesion measuring 4.9 × 4.7 × 5.7 cm with a tiny central scar, consistent with FNH.