CASE REPORT

Association of coronary to left ventricular microfistulae (vessels of Wearn) with atrial septal defect in an adult without cyanotic heart disease

Munem Hussain,¹ Elved Bryn Roberts²

¹Cardiology Department Care of Dr Elved Roberts, University Hospitals of Leicester, Leicester, UK

²Department of Cardiology, University Hospitals of Leicester, Leicester, UK

Correspondence to

Dr Munem Hussain,
munem.hussain@gmail.com

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SUMMARY

Vessels of Wearn are rare findings during coronary angiography in adults. They are known to be associated with forms of cyanotic congenital heart disease in infants but we are not aware of any published cases of association with non-cyanotic left to right shunts in adults. We present the case of a 69-year-old man with angiographically evident vessels of Wearn draining from the left and right coronary arteries into the left ventricle associated with an asymptomatic atrial septal defect. We postulate a developmental phase association between atrial septal maturation and closure of perfusing microchannels from the ventricular cavities to the epicardial coronary arteries on the same spectrum as that which leads to more widespread defects in infants. We also highlight a common medication side effect that might have been mistaken as a manifestation of the congenital anomalies.

BACKGROUND

In adults, vessels of Wearn are usually coincidental angiographic findings. There is limited understanding of the pathophysiological mechanisms underlying their development. The apparent association with an otherwise isolated atrial septal defect suggests a common disturbance in fetal cardiac development.

CASE PRESENTATION

A 69-year-old previously well Caucasian man was admitted to hospital suffering episodes of chest pain at rest and on minimal exertion. His prior health had been unremarkable and he had no cardiovascular disease risk factors or family history of relevance. He had a short soft systolic murmur that was not initially felt to be pathological. Investigations led to diagnosis of critical focal stenosis in the patient's circumflex coronary artery; hence appropriate treatment with a drug eluting coronary stent was given. However, the investigation series also revealed two interesting congenital anomalies that we postulate to be developmentally associated.

INVESTIGATIONS

An ECG revealed normal sinus rhythm and T wave inversion in leads V4 and V5. Serum troponin I level was 122 ng/L (normal <40 ng/L).

Coronary angiography demonstrated tight stenosis in left circumflex artery and a leash of fistulous connections between the distal portions of the left and right coronary arteries and the left ventricle

Figure 1 Left coronary angiogram with contrast in the left ventricle sufficient to cause a partial left ventriculogram.

(figures 1 and 2). Transthoracic echocardiography revealed a large atrial septal aneurysm (figure 3).

Subsequent transoesophageal echocardiography revealed the following notable features: severely dilated right atrium, mildly dilated right ventricle with preserved contractility, mildly dilated left atrium, a large aneurysmal septum primum bulging into the left atrium and a septum secundum defect with left to right shunting (figure 4). Left ventricular size and contractility were normal except for hypokinesia in the basal inferolateral wall. Pulmonary artery pressure was not raised.

TREATMENT

The circumflex artery was treated by deploying a XIENCE drug eluting stent.

Figure 2 Right coronary artery with contrast in the left ventricle.

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Figure 3 Transthoracic echocardiogram revealing atrial septal aneurysm and defect.

The patient made an uneventful recovery and was discharged on secondary prevention medication and dual antiplatelet therapy (aspirin and ticagrelor).

OUTCOME AND FOLLOW-UP

Our patient was readmitted within weeks with marked dyspnoea. This proved to be a side effect of ticagrelor rather than an effect of the ASD; the patient responded completely to switching to clopidogrel. A period of stability followed but he developed recurrence of angina several months later. Repeat angiography revealed severe in-stent restenosis and diffuse circumflex disease requiring further stenting with two Biomatrix drug eluting stents. Our patient has been well since. Cardiac surgical advice is that the septal defect should only be closed operatively if he has significant symptoms attributable to it. The microfistulous vessels of Wearn need no treatment.

DISCUSSION

Coronary to ventricular connections of the type described here are seen in 0.2% of coronary angiography studies.¹ The first

description of such arterioluminal vessels was in anatomical studies by Viessens in 1706. A clear distinction was made between these and the venular connecting vessels of Thebesius, which were first described in 1708.² Terms such as vessels of Wearn, Thebesians and coronary microfistulae are used interchangeably but erroneously, there being a defining anatomical nomenclature to which few adhere. Vessels of Wearn are known to be part of a cluster of cyanotic congenital heart defects surrounding pulmonary atresia with intact ventricular septum (PAIVS).³ In PAIVS, they can coexist with atrial septal defect, tricuspid valve anomalies, persistent ductus arteriosus and proximal coronary anomalies such as ectasia, anomalous origin, stenosis, fibromuscular dysplasia and atresia.⁴ Vessels of Wearn are almost always present when coronary anomalies and PAIVS coexist, although it is believed that the hypertensive right ventricle is a prerequisite for persistence of these myocardium perfusing channels. The coronary anomalies are thought to be due to sheer stress-induced alteration of fetal vascular gene expression, disturbing the development of proximal coronaries and their connection with the coronary orifices within the aortic sinuses.⁵ Microfistulae in keeping with vessels of

Figure 4 Transoesophageal echocardiogram confirming atrial septal defect.

Wearn have also been described in association with hypertrophic cardiomyopathy,⁶ although the causative mechanism is unknown.

In the case described, there is extraordinary atrial septal developmental anomaly as well as marked microfistulous connections between small coronary arteries and the left ventricular cavity; this combination raises a hypothesis of a common pathological developmental trigger responsible for failed microchannel collapse and also for failed atrial septation, probably on the same spectrum as that resulting in cyanotic congenital heart disease.

Restenosis following placement of drug eluting stents is unusual but not especially noteworthy. However, the extent of stenosis found on repeat angiography in this case is surprising.

Learning points

- ▶ Atrial septal defects and coronary microfistulae could plausibly be developmentally linked.
- ▶ Very acute changes in symptoms in the presence of an apparently stable chronic condition should prompt a search for an alternative cause as well as a deterioration in the chronic condition.
- ▶ Ticagrelor is an important cause of dyspnoea, which responds to switching to an alternative antiplatelet drug.

It does raise the possibility that our patient has an underlying non-atheromatous coronary abnormality on the fibromuscular dysplasia spectrum, as in PAIVS.

The fact that the patient presented with marked dyspnoea soon after initial diagnosis is worthy of attention. He had been started on ticagrelor as part of dual antiplatelet therapy following coronary stenting. Ticagrelor is well known to cause this side effect, probably mediated by ATP. This is going to be an important differential diagnosis in routine clinical practice in the coming months and years, as ticagrelor use expands.

Competing interests None declared.

Patient consent Obtained.

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