A rare combination of coronary anomalies

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Coronary anomalies are found in less than 1% of diagnostic coronary angiograms. The clinical relevance of these anomalies varies from insignificant to potentially lethal. The major role of coronary angiography in interventional cardiology and coronary surgery underscores the importance of having knowledge of the variations in coronary anatomy and their clinical relevance. We report a rare case of a patient with a combination of coronary anomalies: coronary fistulae, a double circumflex coronary artery and anomalous origin of a circumflex artery from the proximal right coronary artery. (*Neth Heart J* 2009;17:387-9.)

Keywords: coronary anomalies, double circumflex, coronary fistulae

During the fifth week of foetal development vascular plexuses of sinusoids are formed in the sub-epicardial space. Coronary buds on the aortic sinuses connect with these plexuses leading to the development of coronary arteries.¹ Malformation within one of these systems may cause coronary anomalies; persistence of sinusoids results in coronary fistulae. Coronary anomalies affect approximately 1% of the general population. The importance of coronary anomalies varies from minor to life-threatening.²⁻⁴

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Apart from the clinical relevance, angiographic recognition of these anomalies is important because of the consequences for coronary intervention and coronary bypass grafting in these patients. The most frequently found anomalies include a circumflex artery with a separate ostium from the left anterior descending artery (LAD) originating in the left coronary cusp, an origin of the circumflex artery taking off from the right coronary artery or arising separately from the right coronary cusp (table 1).

Less frequently patients are seen with an anomalous LAD embedded between the aortic root and the pulmonary artery. These patients are at increased risk of sudden cardiac death due to compression of the anomalous LAD when the aortic root expands during the increase in cardiac output during or after vigorous exercise. This can cause myocardial ischaemia and/or sudden cardiac death. Coronary anomalies are responsible for approximately 13% of sudden cardiac death in young athletes.^{5,6} Hypertrophic cardiomyopathy is the only cardiovascular cause more frequently found as cause of sudden cardiac death in young athletes. The understanding of the pathophysiological mechanisms leading to sudden cardiac death is still far from clear and so is the role of the anomalous coronary anatomy.

Another coronary anomaly is the occurrence of coronary artery fistulae. This diagnosis represents only 5 to 15% of all coronary anomalies.^{7,8} If a fistula is small it will not compromise myocardial blood flow and therefore will be without clinical significance. Large arterio-venous shunts, however, can steal blood from the coronary circulation: if the pressure in the terminating vessel or chamber is low, coronary blood flow will predominantly run into the low pressure area. This may cause transient myocardial ischaemia or even myocardial infarction at young age. If necessary, large fistulae can be coiled during cardiac catheterisation; sometimes surgery is a better option.^{9,10}

Clinical history

A 53-year old woman with a prior history of myocardial infarction presented with chest pain. A recurrent myocardial infarction was ruled out on the basis of ECG and serial cardiac enzyme analysis.

Table 1. Incidence of coronary anomalies and dominance patterns, as observed in a continuous series of 1950 angiograms.

Variable	Number	Percentage
Coronary anomalies (total)	110	5.64
- Split RCA	24	1.23
- Ectopic RCA (right cusp)	22	1.13
- Ectopic RCA (left cusp)	18	0.92
- Fistulae	17	0.87
- Absent left main coronary artery	13	0.67
- Cx arising from right cusp	13	0.67
- LCA arising from right cusp	3	0.15
- Low origination of RCA	2	0.10
- Other anomalies	3	0.27
Coronary dominance patterns		
- Dominant RCA	1641	89.1
- Dominant LCA (Cx)	164	8.4
- Co-dominant arteries (RCA, Cx)	48	2.5

Cx=circumflex artery, LCA=left coronary artery, RCA=right coronary artery, split RCA=duplication of the posterior descending branch of the right coronary artery. From Angelini P, et al.⁸ Reprinted with permission.

Figure 1. Left anterior oblique (LAO) straight projection. RCA=right coronary artery, CX2=anomalous circumflex coronary artery arising from the right coronary artery.

Coronary angiography was performed and demonstrated the three large coronary arteries without significant lesions (figures 1 to 3). However, a second circumflex artery, smaller than the one arising from the left main coronary artery, originated from the right coronary artery (figures 1 and 4).

A 90% stenosis of the luminal diameter was found at the ostium of this small aberrant circumflex coronary artery. Further opacification revealed several small coronary arterio-venous fistulae from a large diagonal branch (figure 2).

Figure 2. Left anterior oblique (LAO) cranial projection. LAD=left anterior descending coronary artery, CX=normal circumflex coronary artery, fistulae=coronary arterio-venous fistulae. The images of the fistulae are somewhat vague because of their small dimensions.

Figure 3. Left anterior oblique (LAO) caudal projection. LAD=left anterior descending coronary artery, CX1=normal circumflex coronary artery, D1=diagonal branch.

Figure 4. Schematic illustration of the double circumflex coronary system. RCA=right coronary artery, CX=circumflex, LAD=left anterior descending coronary artery, CX2=anomalous circumflex coronary artery arising from the right coronary artery.

Because of the ostial location of the stenosis of the small circumflex artery and the previous infarction in the area perfused by the small anomalous circumflex artery no coronary intervention was undertaken. The most likely aetiology of the proximal stenosis in the anomalous circumflex branch is atherosclerotic. An intravascular ultrasound could be performed to confirm this; however, the small size of the vessel makes this technically difficult, and the clinical benefit of a percutaneous coronary intervention would be limited. Because the fistulae were small, a conservative approach was preferred and the patient was discharged from the hospital on medical treatment.

Discussion

Coronary anomalies should be recognised during routine coronary angiography. Anomalous circumflex arteries occur most frequently. Especially in the absence of a posterior infarction, one should be aware of the possibility of an anomalous circumflex anatomy instead of assuming a 100% occlusion is present when the circumflex artery can not be displayed during left coronary angiography. If the circumflex artery is indeed proximally occluded, a coronary stump will usually be visible. In addition, wall motion of the left ventricle will be abnormal in the posterior and lateral region during left ventricular angiography. Otherwise, an aberrant origin of that vessel is likely. When a contrast injection into the right coronary artery does not show filling to the aberrant circumflex artery one should look for the 'aortic root sign' during left ventriculography: If the circumflex artery becomes visible in profile behind the aortic root during left ventriculography, the sign is positive and the circumflex artery should be found in the right sinus of valsalva.^{11,12}

Another angiographic sign highly suggestive for anomalous origin of the circumflex artery is known as the 'sign of non-perfused myocardium': An area in the posterior lateral left ventricular myocardium remains avascular during left coronary artery opacification while wall motion in that area is normal.^{11,12}

The anomalous origin of the circumflex artery from the proximal right coronary artery or from the right sinus of valsalva was first described by Antopol and Kugal in 1933. The circumflex artery and the right coronary artery can arise from a common ostium, as separate ostia, or the circumflex artery can arise as a proximal branch of the right coronary artery. These three options occur with approximately equal frequency; however, a combination of one of these with a normal developed circumflex artery has not been previously described.

Conclusion

In general, coronary artery anomalies and fistulae are usually without clinical relevance. However, both may cause ischaemia and can be life-threatening at young age. The occurrence of typical angina or syncope

during or after exercise in young athletes could indicate the existence of a compressed left anterior descending coronary artery or severe shunting due to a fistula and should always be investigated thoroughly.

Case reports describing double right coronary arteries and double left anterior descending coronary arteries originating from right and left coronary arteries have both been published.^{13,14} A circumflex artery arising from the main stem and an anomalous circumflex artery from a separate ostium in the right coronary sinus has been diagnosed as well.¹⁵ To our knowledge, double circumflex coronary arteries arising from the left and right coronary arteries have not been described before. The combination of a second circumflex coronary artery with anomalous origin from the right coronary artery and small coronary fistulae makes this rare angiographic case even more unique. ■

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