Myocardial ischaemia in a case of a solitary coronary ostium in the right aortic sinus with retroaortic course of the left coronary artery: documentation of the underlying pathophysiological mechanisms of ischaemia by intracoronary Doppler and pressure measurements

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Abstract

Only a few cases of a single coronary ostium and retroaortic course of the coronary artery have been described. Almost all cases reported so far had additional coronary artery or valvar disease. However, myocardial ischaemia may be caused by the coronary malformation alone. A 40 year old woman with severe myocardial ischaemia in the absence of clinically relevant coronary atherosclerosis is described. To clarify the origin and mechanisms of ischaemia, intracoronary Doppler, pressure and ultrasound studies were performed using microtransducers. In its outer portion along the course behind the ascending aorta, coronary blood flow velocities were increased, there was an external elliptical compression, and distal coronary flow reserve was reduced. Furthermore, an overshoot in diastolic pressure above aortic pressure was detectable within this portion. Dobutamine stimulation exaggerated the observed intracoronary haemodynamics and induced myocardial ischaemia. The intracoronary diagnostic procedures performed were helpful in clarifying the pathophysiological mechanisms of functional coronary obstruction and ischaemia in this malformation. Bypass surgery was successfully performed with symptomatic improvement.

Keywords: coronary anomaly; Doppler; intravascular ultrasound; single coronary ostium; congenital disorders

A solitary coronary ostium in or immediately cephalad to the right aortic sinus and the course of the left coronary artery dorsal to the ascending aorta is a very rare congenital anomaly. This coronary malformation has been classified as the type IV.A.2a according to the classification of Roberts. At present, only a few cases with this abnormality have been described in the literature. In most of these patients, the diagnosis was incidental either during angiography or necropsy. Most patients had significant concomitant coronary atherosclerosis causing symptoms of angina, or myocardial ischaemia and infarction. Even in the absence of severe coronary atherosclerosis, myocardial ischaemia can occur in cases of anomalous origin of the left coronary artery from the right sinus of Valsalva, if the left coronary artery passes posterior to the aorta. The pathophysiological mechanisms leading to myocardial ischaemia in the absence of coronary artery disease are incompletely understood. This is in contrast to other coronary anomalies where the left coronary artery arises from the right coronary artery and courses between the aorta and pulmonary artery (interarterial or intertruncal course). This has been associated with myocardial ischaemia and sudden death. The mechanism in this scenario is most likely a transient occlusion of the left coronary artery caused by an increase in blood flow through the aorta and the pulmonary artery, resulting in kinking or pinching of the artery. In contrast, in the abnormality presented here, coronary obstruction is usually absent on angiography.

Figure 1  (A) 12 lead ECG showing ST segment elevation in leads I, aVL, V2–V5 with ST segment depression in II and aVF directly after exercise testing, associated with severe symptoms of angina. (B) At rest, ST segment changes diminished and symptoms were alleviated.
We present the first report describing myocardial ischaemia, and its potential pathophysiological mechanisms, in a patient with a left coronary artery originating from the right sinus of Valsalva coursing posterior to the aorta in the absence of clinically relevant coronary atherosclerosis.

Case report

A 40 year old woman reported a four week history of exercise induced angina pectoris; ST segment elevation was documented during bicycle ergometry in the anterior electrocardiographic leads (V2–V5) (fig 1). Immediately after exercise testing, torsade de pointes occurred. There was no history of coronary artery disease or hypertension. The patient had a history of cigarette smoking for a few years before admission. Physical examination revealed an otherwise healthy woman. Coronary angiography revealed a solitary ostium in the right aortic sinus associated with an aberrant retroaortic course of the left coronary artery (figs 2 and 3). There was a 30% atherosclerotic stenosis in the mid-portion of the right coronary artery. Despite several additional angiographic projections, it was impossible to demonstrate a significant phasic luminal narrowing of the left main within its retroaortic course.

Intracoronary Doppler flow velocity and pressure measurements using miniaturised 0.014 inch guidewire transducers and intravascular ultrasound were performed at rest and during dobutamine stimulation. Intravenous Doppler showed only slightly increased blood flow velocities along the curved retroaortic segment of the left main stem (arrow, fig 3), which were enhanced during intravenous infusion of dobutamine (5–20 µg/kg) (fig 4A), compared with measurements at rest (fig 4B) and with measurements at the proximal or distal portions of the left coronary artery (not shown). Distal coronary flow reserve, measured as the ratio of maximal flow velocity after stimulation with 12 mg of intracoronary papaverin to baseline flow velocity, was significantly reduced at 1.7.17

Intracoronary pressure measurements demonstrated a local overshoot in diastolic pressure (fig 5) at the site of accelerated flow velocity within the course of the left descending coronary artery. In vitro validation of pressure measurements within squeezed coronary segments using the 0.014 inch pressure microtransducer has recently been demonstrated.18 Intravascular ultrasound demonstrated only a slight systolic and diastolic reduction of cross sectional area at this portion (fig 4A) compared with a normal distal portion (fig 4B), without signs of atherosclerosis. Measurements during intravenous infusion of dobutamine demonstrated augmentation of blood flow velocities and of intravascular pressure (fig 5), and a further reduction in cross sectional area. Moreover, angina symptoms with ST segment elevation in the anterior leads occurred similar to those during exercise testing. In view of the severity of symptoms and objective evidence of...
ischaemia and ischaemia induced malignant arrhythmias, the patient had aortocoronary bypass surgery with the left internal mammary artery to the left anterior descending coronary artery. Intraoperatively, the course of the left coronary artery behind the aorta was confirmed. No relevant stenosis of the left coronary artery along its course was evident. At eight weeks’ follow up the patient was free from angina, and arrhythmias did not occur during or after exercise testing.

**Discussion**

Congenital coronary malformations are rare causes of angina, myocardial infarction or malignant arrhythmias. Usually, the diagnosis is incidental and clinical symptoms are attributed to other associated disorders, most commonly coronary artery disease. Five possible anatomical anomalies with a single coronary orifice in the right sinus of Valsalva with different clinical implications are known.
Intravascular ultrasound demonstrated an external phasic diastolic vessel compression producing an elliptical shaped vessel lumen that was most evident during dobutamine stimulation. This phenomenon did not occur in any other portion of the coronary artery.

The intracoronary haemodynamic alterations were most likely caused by the diastolic extension of the aortic bulb and sinus of Val- salva compressing the left main coronary artery at the outer most portion of its retroaortic course. Most likely, the left coronary artery was unable to compensate for the expansion of the aortic bulb by its own motion as the vessel was more or less fixed at the aortic origin and during its distal course. Intraoperatively, it appeared that the left main stem was compressed at the above described location, while the other segments of the left coronary artery were completely normal. It remains unclear why the patient was asymptomatic until age 40 and why symptom onset was relatively sudden; reduced flexibility of the elastic properties of the vessel wall by age might in part serve as an explanation.

In summary, we demonstrate a case of severe myocardial ischaemia in a patient with a single coronary orifice in the right sinus of Valsalva and a retroaortic course of the left coronary artery. Intravascular diagnostic procedures at rest and during pharmacological stimulation demonstrated the mechanisms leading to myocardial ischaemia in this rare coronary anomaly. Intracoronary Doppler, pressure and ultrasound measurements were clearly of benefit in this patient in diagnosis and hence in guiding treatment.

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Cardiac valve invasion in chronic adult T cell leukaemia

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The cause of adult T cell leukaemia (ATL) is human T cell leukaemia virus type I (HTLV-I), which was discovered in 1980. This virus is endemic in southwestern Japan, the Caribbean, Africa, and South America. However, cardiac valve involvement by ATL cells is extremely rare and there is no previous echocardiographic evidence for this involvement. We present serial echocardiographic observations in a patient with chronic ATL in whom leukaemic involvement of the mitral valve showed pronounced improvement after chemotherapy.

A 58 year old woman was admitted because of two previous episodes of acute heart failure. On examination, generalised skin eruptions and lymphadenopathies were noted. Third and a fourth heart sounds (gallop rhythms) and a grade 3/6 pansystolic murmur were present at the apex. Transthoracic echocardiography showed extensively thickened mitral and tricuspid valves with moderate regurgitations. The left ventricle was mildly dilated with reduced wall motion. Transoesophageal echocardiography showed an echogenic endocardial lesion at the left atrial wall and appendage, which extended continuously to the bizarre mitral valve abnormality (fig 1). Gallium scintigraphy showed a significant cardiac accumulation.

Serological tests were positive for anti-HTLV-I antibodies by particle agglutination and immunofluorescence. ATL cell infiltration was found in lymph node and skin biopsy specimens. Leukaemic cells from the lymph node expressed T cell markers (CD2+, CD3+, CD4+, CD7+, CD8−, and HLA-DR+). Together with echocardiographic and gallium scintigraphic findings, valvar invasion of tumour cells was suspected, possibly with myocardial involvement.

Figure 1 Transoesophageal echocardiogram showing the bizarre mitral valve abnormality. (A) Echogenic endocardial lesion within the left atrium (arrows). (B) Pronounced improvement after chemotherapy.
Combination chemotherapy with cyclophosphamide, doxorubicin, vincristine, and prednisolone was started. After three chemotherapy sessions, the lesions in the mitral and tricuspid valves responded favourably with obvious reduction in thickness and improved flexibility in leaflet motion (fig 1). Repeated gallium scintigraphy demonstrated disappearance of the abnormal cardiac accumulation.

Despite these improvement the patient had another episode of acute heart failure 10 months later and underwent mitral and aortic valve replacements. At surgery, the mitral valve was extremely thickened. Similar whitish thickenings were found in the left atrial wall and in the basal left ventricular wall. The diagnosis of valvar ATL cell infiltration was confirmed by histopathological examination of a surgically excised mitral valve.

This is the first echocardiographic documentation of cardiac valve involvement in ATL. The serial morphological changes of valvar lesions in response to chemotherapy have not been reported previously. This case has two important messages: first, it may at times be difficult to differentiate echocardiographically leukaemic valvar invasion from myxomatous valvar changes seen in patients with mitral valve prolapse, not only because of the similarity of echocardiographic valvar features but also because of the rarity of leukemic valvar invasion; second, it is important to raise the possibility of involvement of the myocardium by ATL cells when the echocardiographic valvar abnormality is detected in the presence of positive HTLV-I serology, as myocardial involvement is probably more common as suggested by this case as well as by previous pathological reports.4 5 We propose ATL associated cardiac valvulopathy as one of the important signs of cardiac invasion by ATL cells that may be detectable echocardiographically.