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Severe mitral regurgitation due to mitral valve prolapse associated with Bland-White-Garland syndrome

Bland-White-Garland 綜合症與二尖瓣下垂引致嚴重的血液回流

Bland-White-Garland syndrome refers to the rare congenital cardiac abnormality whereby the left coronary artery arises from the pulmonary artery. The natural history of this condition is highly variable, ranging from death in early infancy to asymptomatic adult survival. It is sometimes diagnosed in adults with mitral regurgitation thought to be of ischaemic origin. We report a case of a 29-year-old man with Bland-White-Garland syndrome and concomitant mitral valve prolapse, and review the literature on the appropriate investigations and management of this abnormality. Recognition and diagnosis of this condition is important because of the potentially life-threatening complications, which may be prevented by surgical intervention.

Bland-White-Garland綜合症是一種罕見的先天性心臟異常病，患者的左冠狀動脈從肺動脈而出。這種病例的自然發展情況差異很大：有些患者在嬰兒時已死亡；有些患者則可以生存至成人而期間並沒有任何徵狀。部份患者出現血液回流而被診斷為因局部缺血而引起。本文報告了一名29歲男性患者，他患有Bland-White-Garland綜合症並出現二尖瓣下垂。本文總覽了有關這種心臟異常病的研究及處理方法的文獻。辨別和診斷這種心臟異常病非常重要，因為這種病可以利用外科手術來預防患者產生其他致命的併發症。

Key words:
Coronary vessel anomalies;
Mitral valve;
Mitral valve insufficiency;
Pulmonary artery

關鍵詞：
冠脈管異常；
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Introduction

Anomalous origin of the left coronary artery from the pulmonary trunk, also known as Bland-White-Garland (BWG) syndrome, is rare, occurring in about 1 in 300 000 live births. Only 10% to 15% of patients with this anomaly reach adulthood and may present with heart failure, angina pectoris, effort syncope, ventricular arrhythmias, or sudden death.[1,2] We present an adult case with severe mitral regurgitation due to mitral valve prolapse, who was found to have this rare congenital abnormality after mitral valve replacement. Ligation of the anomalous left coronary artery with aorto-coronary saphenous venous grafting was carried out as a second operation.

Case report

A 29-year-old man presented to his general practitioner with dyspnoea and was referred to Grantham Hospital in August 1997. This patient was known to have had a heart murmur since the age of 9 years. Clinical

examination revealed an irregular pulse of 80 beats per minute, blood pressure of 120/80 mm Hg, and a pansystolic murmur at the apex. An electrocardiogram showed atrial fibrillation, left-axis deviation, and poor R-wave progression. A chest X-ray showed cardiomegaly, with a cardiothoracic ratio of 62%. An echocardiogram revealed a dilated left ventricle with satisfactory contraction and severe mitral regurgitation due to a redundant anterior mitral valve leaflet with prolapse. The right ventricular systolic pressure was estimated to be 75 mm Hg and the patient was referred to a cardiothoracic surgeon for mitral valve repair or replacement.

At operation, the mitral valve was found to be thin and flimsy. Repairing it was not feasible and thus it was excised and replaced with a prosthetic valve. The right coronary and left anterior descending arteries were dilated, with tapering of the distal branches. No thrills suggestive of coronary artery fistula could be detected. The excised valve showed myxomatous degeneration pathology, compatible with a diagnosis of floppy mitral valve. Following the operation, a repeat transthoracic echocardiogram was performed. This showed a dilated right coronary artery origin in the parasternal short-axis view at the aortic root level. Colour-flow Doppler at the parasternal short-axis view at the mid-ventricular level showed turbulence in a dilated intercoronary septal collateral within the septum. Diagnostic cardiac catheterization was performed. An aortogram showed a dilated right coronary artery (diameter measuring 10 mm) with retrograde perfusion of the left coronary system through intercoronary anastomoses. The left coronary artery system was also markedly dilated (the diameter of the anterior descending artery measured 15 mm) and was found to arise from the main pulmonary artery (Figs 1 and 2). An exercise thallium-201 single-photon emission tomography (SPET) scan revealed no evidence of ischaemia. The diagnosis of BWG syndrome was made.

A second operation was performed via a left thoracotomy approach. The anomalous origin of the left coronary artery was ligated and saphenous vein aorto-coronary bypass grafting to the distal left main coronary artery was carried out. Two years after the operation, the patient remained well, with a reduction in the size of the left ventricle and an improvement in exercise tolerance.

Discussion

Bland-White-Garland syndrome is a very rare congenital condition in adults, since most patients who are affected die in early childhood because of myocardial ischaemia or infarction in the left ventricular wall supplied by the anomalous left coronary artery. In adults who survive without undergoing an operation, left-to-right shunting occurs as a result of collateral filling from the enlarged right coronary artery supplying the left coronary artery in a retrograde manner. The dilated collateral vessels are often found in the myocardium of the interventricular septum. Pulmonary hypertension gradually develops because of combined left-to-right shunting, left ventricular dysfunction, and mitral regurgitation. Mitral regurgitation is thought to result from the dysfunctioning of ischaemic papillary muscles and adjacent myocardium,[3] and may improve after correction of the ischaemia. Rarely, mitral regurgitation is not due to ischaemia, but to an associated lesion such as cleft mitral valve or short chordae.[3] Mitral valve prolapse in association with BWG syndrome, however, has not been reported previously. In this case, severe mitral

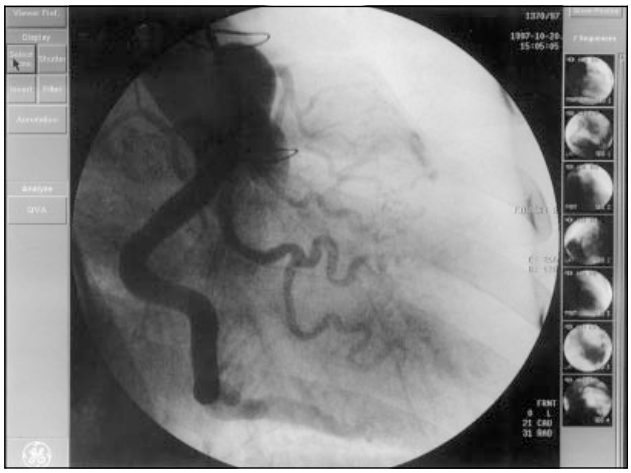

Fig 1. Aortogram in the right oblique view showing a grossly dilated right coronary artery arising from the right coronary sinus

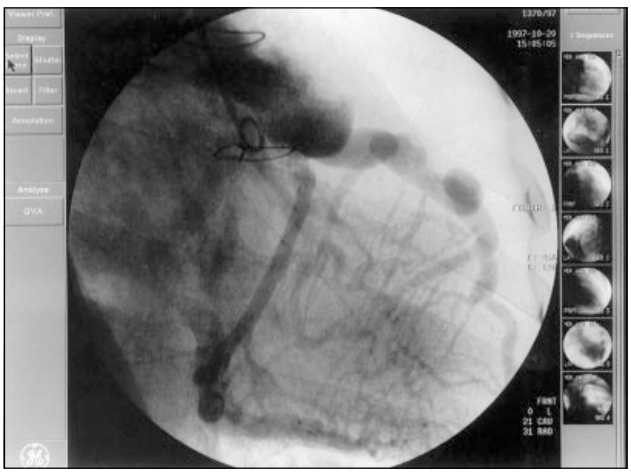

Fig 2. Delayed image of the same aortogram showing retrograde collateral filling of the dilated left coronary system, which drains into the pulmonary artery

regurgitation was due more to myxomatous degeneration of the mitral valve than ischaemic mitral regurgitation, as indicated by the pathological findings of the excised mitral valve and the absence of ischaemia on exercise thallium myocardial imaging.

Although a right coronary angiogram gives the definite diagnosis of this congenital abnormality, echocardiography is useful in identifying whether the dilated right coronary artery arises from the aorta. In addition, colour and pulsed Doppler can be used to demonstrate continuous blood flow from the left coronary artery into the pulmonary artery.[4] Visualisation of the dilated intercoronary collaterals within the interventricular septum using colour Doppler has been achieved previously in infants with BWG syndrome,[5] and we find that this is also possible in adults with this syndrome. Although not specific for diagnosing the condition, the finding of dilated intercoronary collaterals, together with other echocardiographic abnormalities, should prompt the use of a coronary angiogram to look for coronary artery abnormalities. Due to the rarity of this syndrome and the severity of the mitral regurgitation, it is perhaps not surprising that the subtle echocardiographic abnormalities were missed on the initial preoperative echocardiogram. A preoperative coronary angiogram is not indicated routinely in young adults with mitral regurgitation and no risk factors for ischaemic heart disease, although any discrepancy between the severity of mitral regurgitation, degree of left ventricular dilatation or dysfunction, and unexplained pulmonary hypertension, should raise suspicion about the presence of concomitant congenital coronary anomalies.

Electrocardiographic abnormalities of BWG syndrome include left-axis deviation, abnormal Q-waves in leads 1 and AVL, poor R-wave progression resembling lateral myocardial infarction, and left ventricular hypertrophy (with or without ST-segment depression). An abnormal tracing, however, may improve with age, and may be normal or near-normal in children or adults. Evidence of ischaemia in the anterior wall may be commonly demonstrated on stress echocardiography or thallium myocardial imaging, but inferoposterior wall perfusion abnormality has also been reported.[6] In theory, the more extensive the coronary collaterals, the better preserved the myocardial function due to lack of ischaemia, although the degree of coronary steal may then become significant.

Thallium-201 SPET is useful preoperatively for assessing the reversibility of ischaemia as well as postoperatively for evaluating the improvement in ischaemia. It is interesting to note that in Moodie et al's study,[7] all eight patients studied postoperatively with stress thallium scanning did not have ischaemia, irrespective of the type of surgery performed or the patency of the saphenous venous graft. The absence of ischaemia in the preoperative stress thallium scan did not deter us from performing coronary artery surgery in this case. This is because the stress thallium scan may not be able to detect ischaemia in patients with severe subendocardial scarring and calcification, and also because of overwhelming evidence in the published medical literature demonstrating the benefit of surgery in eliminating the coronary steal phenomenon and restoring a dual antegrade coronary flow. Recently, magnetic resonance imaging has been shown to diagnose BWG syndrome accurately and also to assess myocardial viability.[8] Whether this modality will provide better information than a stress thallium scan on myocardial viability and the degree of myocardial damage in this condition, remains to be seen.

Surgery is recommended in patients with BWG syndrome, even in the absence of symptoms or a significant left-to-right shunt syndrome, given the risk of ventricular arrhythmias and sudden death. The pulmonary:systemic flow ratio was not calculated in this case but elsewhere has usually been reported to be small (1.3:1 to 1.6:1).[4,9] The surgical options include reimplantation of the anomalous left coronary artery onto the aorta, ligation alone, or ligation with saphenous vein bypass grafting. Reimplantation is technically difficult in adult patients because of stretching on the coronary repair and the friable nature of the dilated coronary artery.[7] In children with BWG syndrome, re-establishment of a two-coronary artery system is preferable to simple ligation of the anomalous left coronary artery because of the high mortality associated with the latter approach.[10] In adults, simple ligation of the anomalous left coronary artery is sometimes enough by eliminating the coronary steal due to the left-to-right shunting.[7,9,11] In a similar reported case of BWG syndrome with severe mitral regurgitation, however, the patient underwent mitral valve replacement and ligation of the anomalous left coronary artery.[12] After the initial operation, the patient remained in a low-output state, as the presence of severe pulmonary hypertension with raised right ventricular end-diastolic pressure was thought to compromise collateral flow from the right coronary artery to the left coronary artery. A second operation with saphenous venous aorto-coronary grafting to the left coronary system was thus required.[12] For the patient in this study, we took the option of ligation with saphenous vein bypass grafting of the left coronary artery, since the

operation can restore a dual antegrade coronary flow and can be done via a left thoractomy approach, thus avoiding reoperation via a previous sternotomy wound.

Ischaemic mitral regurgitation may improve spontaneously after revascularisation in BWG syndrome, and so initial surgery on the mitral valve is not indicated. In this study, it was difficult to say whether the patient's clinical improvement was due to the mitral valve replacement or correction of the coronary artery abnormality, or both. In patients in whom revascularisation has been delayed, or whose mitral valve is structurally abnormal, however, it is not uncommon to find that mitral regurgitation may be severe enough to warrant mitral valvuloplasty[3] or mitral valve replacement.[9,11,12]

Conclusion

Although BWG syndrome is a rare condition presenting in adulthood, awareness of this congenital abnormality is important, since early diagnosis and treatment may prevent irreversible damage to the myocardium and subsequent complications including myocardial infarction, heart failure, mitral regurgitation, and sudden death. Ischaemic indications, such as previous anterolateral myocardial ischaemia on resting electrocardiogram or abnormal stress thallium myocardial imaging, may not always be present. An anomalous origin of the left coronary artery from the pulmonary trunk should be included in the differential diagnosis of mitral regurgitation in adults and careful echocardiographic interrogation should be performed to prevent a misdiagnosis. Certain echocardiographic findings are highly suggestive of the syndrome, including the demonstration of a dilated right coronary artery arising from the aorta and, using colour and pulsed Doppler, the demonstration of continuous blood flow from the left coronary artery into the pulmonary artery and visualisation of dilated interventricular septal collaterals. A definitive diagnosis can be made using a right coronary angiogram or aortogram. The potentially life-threatening complications in untreated BWG syndrome may be prevented by surgical correction of the coronary artery anomaly, with or without mitral valve surgery.

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