Isolated Left Coronary Ostial Stenosis as a Result of Fibromuscular Dysplasia in a Young Man

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A 28-year-old man was admitted to hospital for investigation of a 2-week history of angina occurring on exertion. Coronary angiography showed an isolated left coronary ostial stenosis and left main trunk plasty was performed 2 weeks later. The pathological diagnosis of the left coronary ostial stenosis was fibromuscular dysplasia, which makes this a rare case. (Jpn Circ J 2000; 64: 988–989)

Key Words: Adolescent; Fibromuscular dysplasia; Isolated left coronary ostial stenosis

Isolated coronary ostial stenosis, in the absence of distal vessel stenosis, is a rare form of coronary artery disease that occurs predominantly in women, usually before menopause! It is uncertain whether such lesions are non-atheromatous or conventional atheroma. We report a case of isolated coronary ostial stenosis in a young man.

Case Report

A 28-year-old man was admitted to hospital for investigation of a 2-week history of angina occurring on exertion. Physical and laboratory examinations revealed that he was not suffering from syphilis, aortitis syndrome or any other inflammatory disease. He was mildly obese (body mass index = 29.8), but did not have hyperlipidemia or hypertension and he was not a smoker. He did not have any symptoms of renovascular hypertension or neurological abnormalities.

Chest X-ray was normal and there was no region of asyn-ergy of the left ventricle on echocardiography. However, although the resting ECG was normal, exercise (Master’s double steps test) induced marked ST segment depression in leads II, III, aVF, V2–6 with chest discomfort and general fatigue. We strongly suspected a severe coronary artery stenosis and performed coronary angiography as soon as possible, which revealed an isolated left coronary ostial stenosis (Fig 1). There were no stenotic segments in any other coronary artery. Two weeks later, left main trunk plasty using pericardium was performed and the symptoms of effort angina completely disappeared.

The pathological diagnosis of the left coronary ostial stenosis was medial hyperplasia, resulting from fibromuscular dysplasia (Fig 2).

Discussion

Fibromuscular dysplasia is not uncommon in middle-aged women! but is rare in young men. Thompson reported 5 patients (0.2%) with an isolated coronary ostial stenosis among 2,105 cases with angiographically defined coronary disease and Topaz et al documented 12 patients (0.06%) with an isolated coronary ostial stenosis among 21,545 consecutive patients who underwent coronary angiography. Two-thirds of those patients were female, so the present case of an isolated coronary ostial stenosis in a young male patient is extremely rare.

The etiology of isolated coronary ostial stenosis is unknown, although several factors have been reported in the literature. Hypoplasia or atresia of the coronary artery ostium or congenital membrane of the coronary artery are causes of ostial stenosis in children, but are extremely rare in adults. Inflammatory involvement of the wall of the aorta, such as syphilitic aortitis or aortitis syndrome, has been documented; but such cases are diagnosed by the serological findings and inflammatory signs. One other cause of this disease has been early atheroma on the coronary ostia; but such patients usually have multiple coronary risk factors. The present patient had neither coronary risk factors nor atheromatous plaque.

The most common cause of isolated coronary ostial stenosis is fibromuscular dysplasia! which is characterized by non-atherosclerotic segmental stenosis and tends to predominate in the young or middle-aged female patients. Although fibromuscular dysplasia is a well-known cause of renovascular hypertension, and is sometimes identified in the carotid or vertebral arteries, it is rarely found in the coronary arteries, especially at the coronary ostia. Because coronary ostial stenosis occurs predominantly in premenopausal women, humoral factors may accelerate the intimal thickening, but as the present case of isolated ostial coronary stenosis occurred in a young man, the cause of the fibromuscular dysplasia remains unclear.

Coronary artery bypass grafting! or either left main trunk plasty or endarterectomy! are the standard surgical treatments, all of which have satisfactory short- and long-term postoperative prognoses. Left main trunk plasty or endarterectomy are technically more difficult compared with bypass grafting, and the direct surgical approach to the ostial lesion may injure the intact intimal layer and accelerate the intimal hyperplasia postoperatively. In this case, we performed coronary angiography 1 month after surgery, which...
revealed that left main trunk plasty had been successful, but further follow-up is necessary to ensure the earliest diagnosis of restenosis, should it occur.

References