

Myocardial revascularization in dextrocardia with situs inversus totalis

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Abstract

Dextrocardia with situs inversus totalis occurs with an incidence of 1 in 10,000 and the pattern of susceptibility to coronary disease is similar to the general population. An adult male patient with situs inversus-dextrocardia who underwent percutaneous intervention for a discrete lesion in the right coronary artery presented two years later with rapid progression of lesions in the left coronary system. Off pump endarterectomy of the obtuse marginal branch and revascularization of the left anterior descending and obtuse marginal branch in this patient are discussed. In addition an outline of the natural history of normal appearing or mildly diseased coronary artery segments at follow-up angiography is presented. (*Ind J Thorac Cardiovasc Surg* 2008; 24: 171-173)

Key words: Endarterectomy, Off Pump, Angiography

Introduction

Situs inversus totalis (mirror-image dextrocardia) is a rare congenital disorder, known for more than 300 years, in which the major visceral organs are reversed from left to right in a mirror image of the normal condition. Its incidence is approximately 1:10,000 and its prevalence varies among different populations. Patients with situs inversus and dextrocardia have normal longevity and the incidence of coronary artery disease is similar to that of the general population.

Coronary revascularization in dextrocardia has been reported previously. This case is unique in that off pump endarterectomy was carried out for extensive atheromatous disease in the obtuse marginal artery in combination with coronary artery bypass grafting.

Case Report

A male patient aged 40 years was admitted with an acute coronary syndrome. Clinical examination revealed an anxious well built individual with a regular pulse rate of 100 beats per minute and a blood pressure of 170/90 mm Hg. A history of smoking, hypertension and a strong family history of coronary artery disease was forthcoming. Cardiovascular and respiratory system examination was normal. A serum cholesterol level of 250 mg/dl and high density lipoprotein level of 28 mg/dl were noted. Chest X-ray revealed dextrocardia and fundic gas shadow on the right side. An electrocardiogram revealed sinus rhythm, left ventricular hypertrophy and acute inferior wall ST elevation myocardial infarction. Echocardiography revealed situs inversus with dextrocardia. There were no associated cardiac malformations. Emergency coronary angiography revealed a significant obstructive lesion in the right coronary artery (Fig. 1a). The left anterior descending artery (LAD) and the circumflex artery were angiographically normal and a 25% stenosis at the origin of the first obtuse marginal branch (OMB) was noted. Primary angioplasty and stenting of the right coronary artery was done and flow re-established. He was started on statins, antiplatelet therapy and advised to stop smoking.

Two years later the patient presented with

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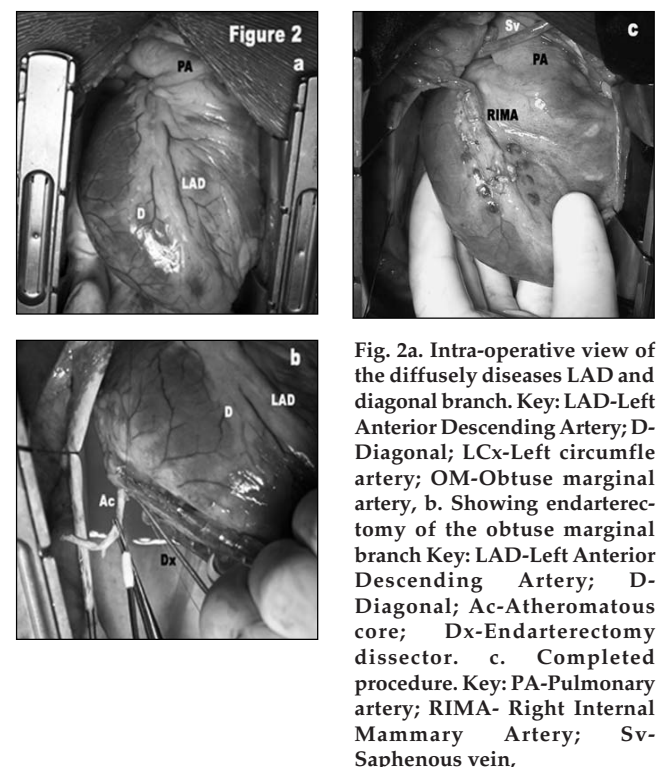
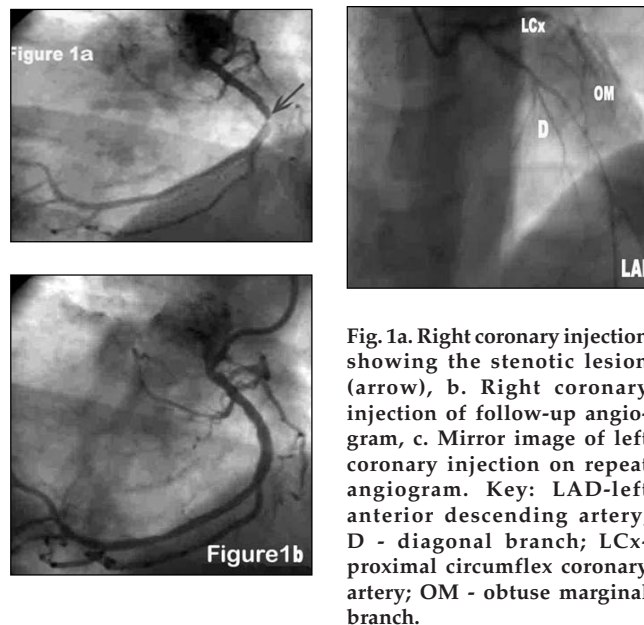
deterioration of the functional cardiovascular class. Although compliant with the medication, he continued to smoke 2 packs of cigarettes per day. The blood pressure was normal, serum cholesterol was 180 mg/dl and high density lipoprotein was 29 mg/dl. An echocardiogram revealed hypokinesia in the apex and antero-lateral wall. Stress test was positive for inducible ischemia. Repeat angiogram demonstrated good flow in the right coronary artery (Fig. 1b). Mirror image views of left coronary system revealed significant disease of the LAD and complete occlusion in the proximal portion of the OMB (Fig. 1c). Left ventricular ejection fraction was 48%.

The patient was scheduled for elective off pump coronary revascularization. An indwelling Swan-Ganz catheter was placed to monitor the pulmonary artery pressure. Intra-operatively the target coronaries were diffusely diseased (Fig. 2a). The completely occluded OMB was revascularized first. The lateral wall of the left ventricle was accessed by lifting the heart upwards with a gentle tilt towards the left pleural cavity through placement of laparotomy pads in the pericardial cavity. This maneuver resulted in torsion of the right ventricular outflow tract (Fig. 2b) leading to significant hypotension with a rise in the pulmonary artery pressure. These adverse hemodynamic effects were countered by placing the patient in the Trendelenburg position and volume loading to increase the filling pressures. In addition the degree of compression applied by the stabilizer device was optimized allowing the heart to slip back into the pericardial cavity. An arteriotomy in the obtuse marginal artery revealed

complete occlusion of the lumen by a calcific plaque. A closed endarterectomy was carried out using a round tipped dissector and an endarterectomy spatula to detach the atheromatous core. Constant gentle traction applied on the plaque by the surgeon and counter traction resulting from each heart beat aided in complete removal of the downstream segment of the plaque followed by removal of the proximal segment (Fig. 2b). Autologous saphenous vein was then used for distal anastomosis on the OMB. Meticulous attention was paid to maintenance of a stable rhythm and hemodynamic status. The pedicled right internal mammary artery was then anastomosed to the LAD (Fig. 2c). The procedure was carried out by the primary surgeon from the left side of the operating table.

Discussion

Dextrocardia with situs inversus totalis is a rare condition with an incidence of 1 in 10,000 and susceptibility to coronary artery disease is similar to that of the general population¹. Standard angiographic views in dextrocardia which may be difficult to interpret are simplified by the use of double inversion technique². Per-cutaneous intervention in dextrocardia necessitates image acquisition in mirror image angles and catheter rotation in the opposite direction¹. This patient



underwent successful percutaneous intervention on the right coronary artery during the initial presentation.

Off pump coronary revascularization has been performed for coronary artery disease in patients with dextrocardia by modifying the surgical approach^{3,4}. The limitations imposed by graft length and lie are offset by using the right internal mammary artery as the conduit for LAD and the anastomosis is conveniently performed from the left side of the patient.

Although endarterectomy is associated with a higher mortality and a poorer long term outcome in comparison to bypass grafting alone, in the presence of diffuse coronary artery disease, the procedure may be necessary to ensure a satisfactory anastomosis. In this context, good long term results have been reported⁵. Coronary revascularization under cardiopulmonary bypass for situs inversus with coronary artery disease has previously been reported. Of late off pump coronary revascularization has been carried out successfully in patients with dextrocardia⁴.

In our patient diffuse disease of the OMB necessitated an endarterectomy which was carried out without cardiopulmonary bypass. During off pump surgery the closed technique of endarterectomy is facilitated by the traction and counter-traction resulting from the rhythmic contraction and relaxation of the heart⁶.

An additional interesting feature in this case is the appearance of disease in previously normal coronary segments in this patient. Defined as a 20% or greater increase in pre-existing stenoses or formation of new stenoses with greater than 20% luminal narrowing⁷, progression of disease has a variable pattern and pace in each coronary artery and predominantly involves normal coronary segments⁸. Such progression suggests that patients with apparently normal coronary arteries may have intimal disease undetectable by angiography⁹. In mild to moderate disease, angiographic progression is slow, predominantly results in formation of new coronary stenoses and to a lesser extent in growth of pre-existing lesions with significant correlation to

cigarette smoking and serum cholesterol levels respectively⁷. Differential progression of lesions within the same coronary tree showing a higher annual rate of growth in complex lesions is reported¹⁰.

In conclusion application of the principles of off pump revascularization and endarterectomy ensured complete revascularization and a satisfactory outcome in this unusual presentation.

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