Abstract

Situs inversus totalis with dextrocardia is rare and presentation with coronary artery disease in the ninth decade of life is even rarer. Here we describe a case where a patient underwent coronary stenting and subsequently coronary bypass surgery for the same.

Keywords: Dextrocardia; Situs inversus totalis; Coronary revascularization; Angioplasty; Coronary stent; Off-pump coronary artery bypass grafting

1. Introduction

Hieronymus Fabricius is credited with describing situs inversus in the 1606, while Marco Severino described dextrocardia in 1643 [1]. Although the exact aetiology is unclear it is thought to be autosomal recessive. Situs inversus totalis with dextrocardia is rare but the incidence of atherosclerotic heart disease is similar in patients who survive long [2]. We are increasingly seeing a more ageing population coming for coronary artery surgery. Here we describe a patient who presented with coronary artery disease in the ninth decade of life with situs inversus totalis with dextrocardia.

2. Case report

An 82-year-old man who was known to have dextrocardia with situs inversus was admitted to hospital (Fig. 1), while on holiday in Spain with unstable angina for which he underwent right coronary artery angioplasty with stenting. His past medical history included 8 years history of chronic stable angina and a strong family history of ischaemic heart disease. He had undergone vagotomy and gastrojejunostomy in the past for a duodenal ulcer.

After his return from holiday, his symptoms recurred and he developed severe disabling angina, occasionally at rest and frequently associated with dyspnoea. This necessitated another coronary angiography; his dextrocardia with situs inversus did not present a problem for angiography and standard Judkins catheters were used. The aorta was right sided. Aortic valve was competent without any stenosis or gradient. Left ventricular function was moderately impaired. There was anterior hypokinesis with an estimated ejection fraction of 45%. Coronary vessels were an exact mirror image of coronary circulation. The morphological left main coronary artery was blocked at origin, showing a tubular atherosclerotic lesion in the proximal part with partial filling. The diagonal vessel was small and not graftable. The circumflex vessel was recessive. The morphological right coronary artery was large, dominant and patent; a stent was identified in the mid-third of this vessel and was satisfactorily positioned. There was a large posterior descending vessel. There was further obstructive disease at the crux (Fig. 2).

Following coronary angiography the patient proceeded to elective coronary artery bypass grafting. The chest was entered through a median sternotomy. Initial findings were a heart of good contractility and exactly the mirror image of a normally positioned heart. The procedure started as an off-pump operation and in this manner the left anterior descending artery was opened and grafted with the right internal mammary artery using the CTS (Genzyme Corp., Cambridge, MA, USA) stabilizer, with the surgeon standing on the left side of patient. While positioning the heart, in order to be able to reach the posterior descending artery (PDA), it became quite apparent that the heart would not tolerate any displacement of its inferior wall. In fact, as soon
as the inferior wall was displaced it went into atrial fibrillation with marked increase of the pulmonary artery pressure and decrease in the systemic pressure. The heart was then assisted with cardiopulmonary bypass after full heparinization. The PDA was then approached without stopping the heart, and using the stabilizer, it was grafted with a segment of vein. The proximal end of the vein was then anastomosed to the ascending aorta. Bypass lasted 38 min and was able to be discontinued with no problems. After accurate haemostasis and reversal of the heparin the chest was closed in routine fashion.

The patient’s postoperative course was uncomplicated and he was discharged from hospital on day 7. At the last follow-up, at 8 months, he remains angina-free with good exercise tolerance; after discharge he had a contrast enema, which revealed diverticular disease and a benign adenomatous rectal polyp, and this was excised endoscopically.

3. Discussion

There have been reports of patients presenting with coronary artery disease who had dextrocardia with situs inversus totalis [3–5]. Our patient lived a long and fruitful life to reach the ninth decade before presenting to us for coronary revascularization. It is usually considered that only patients who have dextrocardia with situs inversus totalis can survive long enough to develop atherosclerotic heart disease [2]; however, this is the first report which demonstrates that these patients can live up to the ninth decade and reflects the trend in a more elderly population presenting for heart surgery.

Our patient underwent coronary angiography twice, and both times there was no technical difficulty encountered, and also the deployment of a stent in the right coronary artery did not pose any difficulty. This is similar to earlier reports [6]. Unfortunately the stent did not alleviate the patient’s symptoms and he needed surgical revascularization. We anastomosed the right internal mammary artery to left anterior descending artery without using cardiopulmonary bypass, this technique having been used previously only once in dextrocardia with situs inversus totalis [7]. However, the posterior descending artery in our patient could not be grafted off-pump due to haemodynamic instability and hence, we decided to assist the heart with cardiopulmonary bypass without stopping the heart. This allowed us to avoid giving cardioplegia and allowed the completion of the procedure satisfactorily. The procedure was greatly facilitated by the surgeon standing on the left side of patient; otherwise we did not encounter any technical difficulty. To our knowledge, this is a first case of a patient with dextrocardia and situs inversus totalis who reached the ninth decade of life who needed coronary stenting and subsequently surgical revascularization.

In conclusion, Long-term survival is possible in patients who have situs inversus totalis. The mirror image anatomy does not pose an unusual technical challenge in myocardial revascularization, either surgically or non-surgically.

References


