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Cardiac denervation procedure to treat refractory angina in a patient with Churg–Strauss syndrome and non-obstructive coronary lesions

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Abstract

Cardiac involvement in Churg–Strauss syndrome is common and represents the main cause of mortality. We report the case of a patient with Churg–Strauss vasculitis, mitral regurgitation with left ventricular dysfunction, paroxysmal atrial fibrillation and refractory angina with non-significant coronary lesions. Cardiac denervation was proposed as an associated procedure to treat angina. The total removal of peri-adventitial and adventitial tissue around the superior vena cava, ascending aorta and main pulmonary trunk was performed. After 3 months of follow-up, the patient was angina-free and could resume his normal lifestyle.

Keywords: Angina • Autonomic nervous system

Churg and Strauss initially described their syndrome as a necrotizing vasculitis of medium-to-small-sized veins and arteries, associated with eosinophilic infiltration around the vessels and adjacent tissues [1]. Cardiac involvement is a leading cause of mortality and a common clinical manifestation [2]. Mitral regurgitation is associated with myocardial fibrosis and normal leaflets [3]. Angina pectoris is common and it is refractory to management with anti-ischaemic medical treatment, requiring a combination of immunomodulators and corticosteroids [4].

We report the case of a 38-year old male patient with Churg–Strauss syndrome whose chief complaint was severe chest pain refractory to medical treatment; subsequently surgical treatment by the use of cardiac denervation was proposed. The clinical manifestations of Churg–Strauss included bronchospasm, mononeuritis and autoimmune hepatitis. The patient was referred to our centre due to a 6-month history of minimal exertion atypical chest pain, progressive dyspnoea, actual NYHA III, and palpitations refractory to complete anti-ischaemic and heart failure treatment. The physical examination revealed a holosystolic 4/6 murmur in the apex. An electrocardiogram showed pathological Q-wave and T-inversion in DII, DIII and AvF. Holter (24 h) revealed paroxysmal atrial fibrillation. A chest X-ray was normal. Transthoracic echocardiogram (TTE) revealed a dilated left ventricle (end-diastolic diameter of 72 mm), diffuse hypokinesia with inferior and inferior-septal akinesia. Left ventricular ejection fraction (LVEF) was 30%. The size of the left atrium was 31 cm^2 (normal <19 cm^2). Severe mitral regurgitation with normal leaflets, annular diameter of 44 mm and posterior leaflet tenting with coaptation deficit was detected. Non-significant coronary lesions were evidenced at angiography (Fig. 1). Surgery was indicated with the following aims: the treatment of mitral regurgitation by mechanical prosthesis implantation with total subvalvular apparatus preservation; radiofrequency atrial ablation to treat atrial fibrillation and ventral cardiac denervation to reduce the patient's refractory chest pain.

The patient's chest was entered through a median sternotomy. Cardiac denervation consisted in the removal of a 4 cm segment of adventitial and peri-adventitial tissue around the superior vena cava, ascending aorta and main pulmonary trunk (Fig. 2). Pulmonary vein isolation was done using endocardial monopolar-irrigated radiofrequency. A 29-mm St Jude mechanical prosthesis was implanted with the total preservation of the subvalvular apparatus. The postoperative course had no complications. The patient was discharged from ICU after 72 h and discharged from hospital 7 days after surgery with the following medical treatment: warfarin, amiodarone, spironolactone, prednisone and azathioprine. TTE at the 1-month follow-up showed an important reduction in the left ventricular end-diastolic diameter (LVEDD) (52 mm), persistent akinesia, improvement in LVEF (40%) and normal functioning prosthesis. After 3 months of clinical follow-up, the patient is asymptomatic with no chest pain, dyspnoea nor palpitations. Twenty-four-hour Holter showed sinus rhythm.

COMMENT

Churg–Strauss syndrome is a medium–small vessel vasculitis. Diagnostic criteria defined by the American College of Rheumatology include: asthma, eosinophilia (>10% of leucocytes), mono- or polyneuropathy, pulmonary infiltrates, paranasal sinus abnormalities and extravascular eosinophilia [5]. Cardiac involvement is a common morbidity issue and represents the main cause of mortality in these patients [6]. Coronary vasospasm has been described as the main cause of chest pain in patients without significant coronary disease [4]. This kind of chest pain is usually refractory to management with

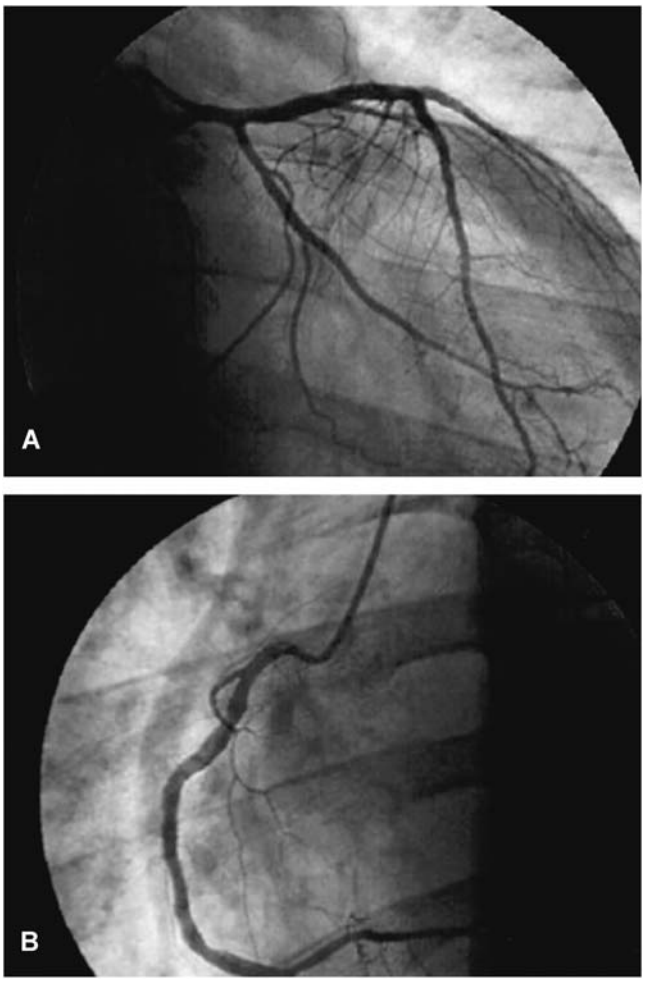

Figure 1: Right and left coronary angiography. The coronary arteries have a diseased appearance with no significant coronary obstruction. (**A**) Left and (**B**) coronary angiography.

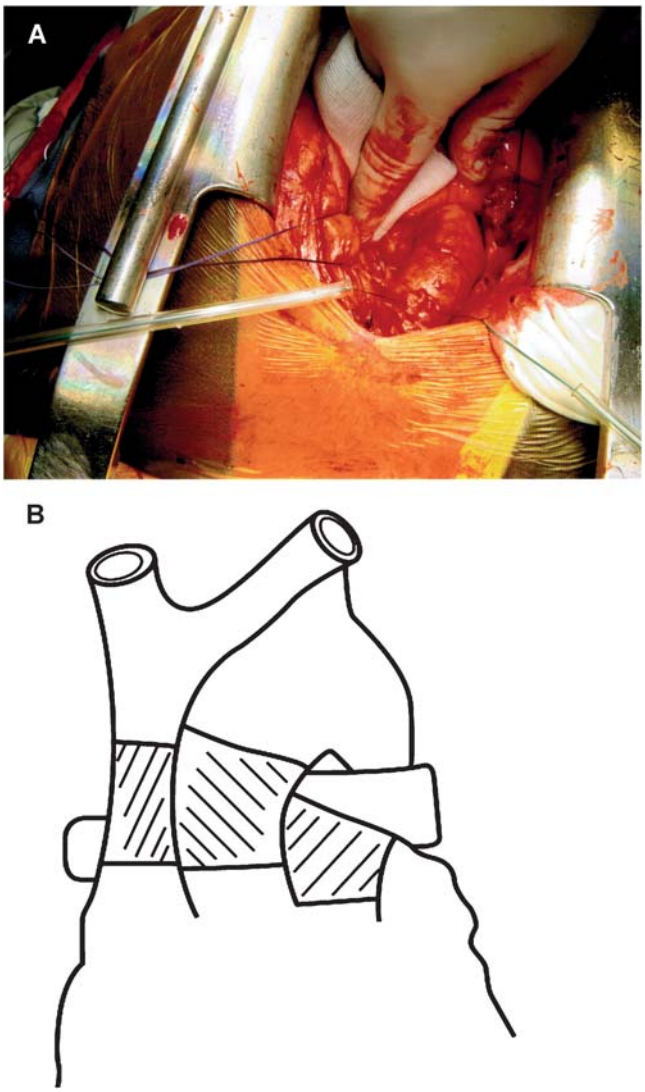

Figure 2: (**A**) The 4-cm cardiac denervation performed intraoperatively on the great vessels. (**B**) The denervation technique.

conventional medical treatment, requiring higher doses of corticosteroids in combination with immunomodulatory drugs [4]. In our patient, chest pain was the main issue of concern and, therefore, we associated ventral cardiac denervation with the treatment of mitral regurgitation and atrial fibrillation.

In 1939, Arnulf [7] proposed a surgical treatment for Prinzmental variant angina, which was based on ventral cardiac denervation. Later, this and similar procedures were used by several groups [8] when medical treatment was ineffective in treating coronary vasospasm. A minimally invasive approach using video-assisted thoracoscopic sympathectomy has been successfully used in patients without indication for surgery [9]. There is a little information on the use of cardiac denervation for the treatment of angina in patients with Churg–Strauss vasculitis. We performed a partial cardiac denervation by removing 4-cm peri-adventitial and adventitial tissue around the superior vena cava, ascending aorta and main pulmonary trunk, which included adipose tissue, autonomic nerve branches as well as the superficial and deep cardiac plexus. This procedure was easily done without cardiopulmonary support. Since the LVEDD exceeded 65 mm, we decided to replace the mitral valve instead of performing a restrictive annuloplasty and, therefore, to avoid late follow-up recurrence of mitral regurgitation, as shown by Braun *et al.* [10]. After 3 months of clinical follow-up, the patient was chest pain-free and enjoyed a normal lifestyle with excellent exercise tolerance. Although we were able to alleviate the chest pain in this patient, the precise mechanism is unknown. Two hypotheses could be brought up: the patient is painless due to the interruption in the afferent pain pathway, which could explain the absence of pain independently from its real cause (vasospasm, arteritis and pulmonary hypertension). The second possibility is that chest pain in this patient is due to coronary vasospasm, which was treated by cardiac denervation. This distinction could have been evaluated by doing pre and postoperative evaluations of coronary vasospasm. Such a procedure was not performed due to the patient's clinical situation.

In conclusion, we believe that the use of a cardiac denervation procedure in this patient with Churg–Strauss vasculitis and refractory angina with non-significant coronary lesions was successful in treating chest pain and improving the quality of the patient's lifestyle. The association of cardiac denervation to surgical indication procedures should be considered in patients with refractory chest pain and non-obstructive coronary lesions.

Conflict of interest: none declared.

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