Repeat pulmonary thromboendarterectomy after recurrence of chronic thromboembolic pulmonary hypertension*

Reoperação de tromboendarterectomia pulmonar em recidiva de tromboembolismo pulmonar crônico hipertensivo

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Abstract

Pulmonary thromboendarterectomy has been established as the standard method for the treatment of chronic thromboembolic pulmonary hypertension, with excellent results. However, repeat pulmonary thromboendarterectomy due to recurrence of pulmonary embolism has never been reported in the Brazilian literature. Its safety and effectiveness remain obscure. We report the case of a patient presenting recurrence of chronic thromboembolic pulmonary hypertension five years after the first pulmonary thromboendarterectomy and requiring a second operation for resolution of the symptoms.

Keywords: Hypertension, pulmonary; Pulmonary embolism; Endarterectomy/methods.

Introduction

Thromboendarterectomy has been established as the treatment of choice in the treatment of chronic thromboembolic pulmonary hypertension (CTEPH), providing, in the majority of cases, a consistent, definitive alternative for treating patients with this disease, with satisfactory short- and long-term results.1,2 Recurrence of CTEPH after the first surgical treatment is rare, seen in 1.5% of the patients evaluated in the only case series study of such patients in the international literature.3 However, reoperative thromboendarterectomy in patients presenting recurrence of CTEPH has not been reported in the national literature, its safety and effectiveness therefore remaining unclear.

Here, we report the case of a patient presenting CTEPH recurrence five years after the first pulmonary thromboendarterectomy and requiring reoperative thromboendarterectomy for the resolution of the symptoms.

Case report

A 28-year-old female patient presented with progressive dyspnea on exertion and chest pain for two months. She had been submitted to pulmonary thromboendarterectomy five years prior, thereafter receiving warfarin, which had been discontinued one year prior. An electro-
cardiogram showed signs of right ventricular overload, and a chest X-ray revealed a moderate increase in the cardiac silhouette, with prominence of the pulmonary artery.

An echocardiogram revealed right atrial enlargement, with a dilated, hypokinetic right ventricle, as well as mild to moderate tricuspid insufficiency. We observed two thrombi, one occluding the left pulmonary artery and the other in the right atrium near the junction with the superior vena cava. The pulmonary artery systolic pressure was estimated at 65 mmHg.

A CT scan of the chest revealed thrombosis and occlusion of the left branch of the pulmonary artery (Figure 1).

The patient was again submitted to median sternotomy involving extracorporeal circulation and total circulatory arrest, with systemic hypothermia at 18°C. Thromboendarterectomy of left branch of the pulmonary artery was performed, and exploration of the right branch revealed no thrombi (Figure 2). In addition, the thrombus was removed from the right atrium. A Greenfield inferior vena cava filter was also inserted. The duration of extracorporeal circulation was 109 min, and that of total circulatory arrest was 31 min. The patient presented favorable post-operative evolution, without complications, and was discharged from the hospital on post-operative day 5. At 59 months after the surgery, the condition of the patient was categorized as New York Heart Association functional class I, and she was under continuous treatment with warfarin. A control echocardiogram showed the pulmonary artery systolic pressure to be 40 mmHg, which remained stable over the following years, together with mild tricuspid insufficiency.

**Discussion**

It is currently estimated that only 3.8% of patients initially presenting acute pulmonary embolism develop symptomatic CTEPH within the following two years. Recurrence of the disease after thromboendarterectomy is even more rarely reported.

Among patients with a mean pulmonary artery pressure greater than 30 mmHg, the five-year survival rate is 30%. If the mean pulmonary artery pressure exceeds 50 mmHg, the five-year survival rate drops to only 10%. Other than lung transplantation, pulmonary thromboendarterectomy is the only effective treatment for this disease, although it carries considerable risk, the reported mortality ranging from 6.6% to 23%. The most common causes of post-operative mortality are reperfusion edema of the lungs, massive hemoptysis, right ventricular insufficiency, respiratory failure and multiple organ failure. In addition, pulmonary thromboendarterectomy does not always result in normalization of pulmonary vascular resistance, principally due to the concomitant peripheral lung injury.

Patients submitted to pulmonary thromboendarterectomy require lifelong anticoagulation...
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Inadequate levels or discontinuation of anticoagulation therapy puts patients at risk for recurrence of the disease, as in the case reported here.

In the only case series study of reoperative pulmonary thromboendarterectomy, the authors found that, for the second procedure, peri-operative morbidity was greater than and mortality was similar to that resulting from the first procedure. In addition, there was less long-term post-operative reduction in pulmonary artery pressure after the second procedure. However, the patients presented pronounced improvement of symptoms during the long-term follow-up treatment. Therefore, reoperative thromboendarterectomy is preferred over lung transplantation or the inexorable evolution to right heart failure and death. Despite being empirical and analogous to other cardiac procedures, in which a second operation carries an increased mortality risk, no such increase was observed in the previously cited case series study. A satisfactory post-operative evolution of our patient corroborates this observation.

Based on the angiographic, tomographic and anatomical findings, it was possible to recommend, plan and devise strategies for the surgery. In the approach to the pulmonary artery, despite the fact that it was a reoperative procedure, the plane of dissection between the thrombus and the arterial wall was clearly outlined, and it was possible to extract a significant part of thromboembolic material. For the sake of safety, the right branch of the pulmonary artery was also explored. However, there was no evidence of contralateral thrombus.

In conclusion, reoperative pulmonary thromboendarterectomy for the treatment of recurrence of chronic pulmonary thromboembolism can be performed with an acceptable surgical risk, providing long-term functional and symptomatic improvement.


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