Management of incidentally diagnosed pulmonary artery dissection in patients with pulmonary arterial hypertension

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Abstract

Pulmonary artery dissection is a rare entity, usually discovered post-mortem. In recent years, reported cases of living patients with a pulmonary artery dissection are growing and represent a challenging situation for clinicians.

Keywords: Pulmonary artery dissection • Pulmonary arterial hypertension • Computed tomography • Sudden cardiac death

INTRODUCTION

Pulmonary artery dissection (PAD) is a rare condition, usually diagnosed in post-mortem studies [1]. We describe 3 patients with pulmonary arterial hypertension (PAH), in whom PAD was incidentally diagnosed. All decisions were agreed upon by all the members of our PAH multidisciplinary unit including cardiologists, radiologists, anaesthesiologists and cardiac and thoracic surgeons.

CASES

Case 1: computed tomography in the setting of chest pain

A 70-year-old man presented to the emergency room with a complaint of chest pain. Computed tomography (CT) angiography showed a large pulmonary artery (PA) (66 mm) causing left main coronary artery compression. A PAD was also identified (Fig. 1A and B). Right heart catheterization confirmed PAH (the mean PA pressure was 58 mmHg). Coronariography with intravascular ultrasound revealed a significant narrowing of the left main coronary artery lumen, resulting from PA compression. A drug-eluting stent was implanted, and tadalafil and ambrisentan were started. During the last 8 years, he has not experienced PAD-related complications.

Case 2: computed tomography in the setting of worsening functional class

A 41-year-old woman with a previous atrial septal defect correction and diagnosed with PAH when she was 35 years old was admitted to the cardiology ward due to worsening functional class. She was being treated with sildenafil and had severe PAH with a Doppler-estimated systolic PA pressure of 113 mmHg. A chest-X-ray showed an enlarged PA. CT confirmed PA enlargement (46.6 mm). Furthermore, an intimal flap was identified at the left PA (Fig. 1C and D). Ambrisentan was added to her treatment regimen. As the body mass index was >35 kg/m², she was not considered suitable for lung transplantation. At the age of 51 years, 10 years after the diagnosis of the PAD, the patient died as a consequence of progressive heart failure. Complications secondary to the PAD were not reported.

Case 3: computed tomography prior to inclusion in the lung transplant waiting list

A 44-year-old woman, with PAH development after surgical correction of a ventricular septal defect at the age of 7, and under treatment with tadalafil, ambrisentan and teprostinil, started evaluation for inclusion in lung transplant programme due to signs of poor prognosis. Right heart catheterization showed the mean PA pressure of 86 mmHg. CT revealed an aneurysmatic dilatation of...
Figure 1: (A) Computed tomography in the axial view showing the pulmonary artery with an intimal flap (arrow) and thrombus in the RPA (arrowheads). (B) Computed tomography multiplanar reconstruction showing the left main coronary artery compression (arrow) and MAP dissection (arrowheads). (C) A chest X-ray showing the dilated MPA. (D) Computed tomography in the axial view showing the dilated RPA and LPA and LPA dissection (arrow). (E) Computed tomography in the axial view showing the dissection affecting the RPA (arrows) and the LPA (arrowheads). (F) Computed tomography—coronal multiplanar reformatted images showing the RPA (arrows) and LPA (arrowheads) dissection. Ao: aorta; LPA: left pulmonary artery; LV: left ventricle; MPA: main pulmonary artery; RPA: right pulmonary artery.
the right and left pulmonary arteries (41.9 mm and 31.2 mm, respectively). PAD was identified in both PA branches with extension up to the segmental arteries (Fig. 1E and F). The cardiothoracic surgery team decided to include the patient on the heart–lung transplant waiting list, instead of a bilateral lung transplant, due to the expected technical difficulties associated with the PAD. She remains stable without symptoms secondary to the PAD.

DISCUSSION

PAD is a rare entity among PAH patients [1]. However, in the past few years, diagnosis of PA dilatation and its complications are increasing [2], including potentially fatal conditions such as PAD, which could be responsible for the higher risk of sudden cardiac death among patients with a dilated PA [3]. PAD represents a challenging situation as the surgical risk in PAH patients is particularly high.

In patients with chest pain, conservative management with medical therapy has been shown to have variable results [1]. Several cases of successful surgical treatment with graft insertion have also been reported [1, 4], and some experts recommend surgery for all PAH patients with a PA aneurysm. Some other groups have recommend a conservative approach in the acute phase and deferred lung transplant [1].

In asymptomatic patients, clinicians must choose between a conservative strategy with an unpredictable risk of sudden cardiac death and a surgical approach with the well-known perioperative risk. A recent series examining the surgical approach for PA aneurysm in 38 patients with mild pulmonary hypertension (the mean systolic right ventricular pressure was 45.8 ± 21.2 mmHg) described excellent short and long-term outcomes [5]. However, none of these patients had a PAD.

Although a main PA diameter of >48 mm has been described as an independent risk factor for sudden cardiac death in PAH patients [3], some series have reported on patients with PAD with a PA diameter of >48 mm, who were treated medically and included on the transplant waiting list, while remaining stable for more than 3 years [2].

Similarly, we described 3 patients incidentally diagnosed with a PAD and who were conservatively managed with good long-term survival. All 3 patients had more severe PAH than that described in recent surgical series of uncomplicated PA aneurysm [5], which implies an extremely high surgical risk. Furthermore, 2 of the 3 patients had undergone previous cardiac surgery, which further increased their surgical risk. An additional surgery would discourage lung transplantation in the future if it was necessary.

Two patients were not included on the transplant waiting list and survived more than 8 years. In these patients, a transplant could have reduced their survival.

The third patient was included on the heart–lung transplant waiting list, but the indication was not based on the presence of a PAD. After inclusion on the waiting list, she remains asymptomatic and stable.

On the basis of our observations, we believe a conservative approach with close follow-up for PAH patients, with an asymptomatic PA dissection might be a safe option.

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REFERENCES