Heart Failure in a Patient With Wolff-Parkinson-White Syndrome

Insuficiencia cardíaca en un paciente con Wolff-Parkinson-White

To the Editor,

A 41-year-old patient was referred to our center for ablation of accessory pathways (AP) in the context of Wolff-Parkinson-White (WPW) syndrome. The patient, who was still asymptomatic, had been diagnosed with WPW 9 years earlier in a routine examination. Three years before the present admission she began to refer to short episodes (<5 min) of sudden-onset, sudden-offset palpitations, which were never recorded by electrocardiogram (ECG). In the past 9 months, she began to show signs of heart failure in the form of New York Heart Association functional class II effort dyspnea. Physical examination was normal, but during exercise testing she only achieved 6 METs. The ECG showed sinus rhythm with pre-excitation manifested by a right antero-septal AP suggesting a classic pattern of left bundle branch block (LBBB) (Fig. 1A). Both echocardiography and magnetic resonance imaging (MRI) showed a marked intraventricular dyssynchrony, with early activation of the septum and stent obstruction due to tumor growth. Repeat angioplasty was not technically feasible. Following considerable discussion by the medical team and in accordance with the patient’s wishes, surgical reduction of the tumor mass was undertaken as the last therapeutic option to relieve the progressive symptoms. The patient died 3 days later, due to multiple postoperative complications.

Primary angiosarcomas of the pulmonary artery are rare, and only a few cases have been described. The similarity of acute and chronic PTE at onset and the inability of imaging techniques to yield an accurate diagnosis (although they may be useful in the differential diagnosis) lead to delays in the definitive diagnosis, which can only be obtained by tissue sampling or autopsy (60% of cases). Percutaneous endomyocardial biopsy is a safe, easily performed procedure with low morbidity and mortality. To enhance the diagnostic benefit, the biopsy can be taken under fluoroscopic guidance, as in the case described, or under transesophageal echocardiographic guidance.

Angiosarcoma presents in middle age (40–65 years) and tends to affect only one of the pulmonary branches, unlike PTE which tends to affect both. The tumor grows rapidly and produces local invasion and distant metastasis. Vascular occlusion is the main cause of death. The prognosis is poor: the mean survival is 1.5 months if surgical resection is not possible and about 12 months if the patient undergoes surgery, which is usually partial.

Angioplasty of the pulmonary trunk is a minimally invasive, palliative procedure that has only been reported in a few cases. The procedure could be useful to improve symptoms due to obstruction and can be combined with chemotherapy or radiotherapy to reduce the tumor size and improve the quality of life in patients excluded from surgical treatment.

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Figure 1. A: electrocardiogram showed a pattern of pre-excitation by a right antero-septal pathway suggesting a classic pattern of left bundle branch block. B: normalization of the electrocardiogram after ablation.
compared to the free wall and a mildly depressed ejection fraction (48%).

The patient remained asymptomatic after ablation. The ECG after ablation was strictly normal (Fig. 1B). At 3 months after ablation, functional class was normal. An MRI 4 months after ablation showed normalization of the ejection fraction (63%) and absence of asynchrony (Fig. 2).

It has been reported that intraventricular dysynchrony and subsequent systolic dysfunction due to early septal activation in patients with a right septal AP occur relatively frequently (in up to 50% of patients). In addition, some prospective studies have emphasized that dysfunction in the left ventricle can lead to its expansion over the long term. The incidence of asynchrony and its progression to ventricular dysfunction and myocardial dilatation may be underestimated in these patients as they generally undergo early ablation due to the presence of palpitations secondary to paroxysmal tachycardia, and these can occur before heart failure symptoms appear. For that reason, it has been suggested that right septal AP ablation should be carried out even before palpitations occur.

It seems reasonable to rule out tachycardiomyopathy in this patient, as imaging techniques did not show ventricular growth and short episodes of tachycardia make this diagnosis unlikely.

Several studies have shown abnormalities in ventricular wall motion in patients with WPW syndrome. Left-sided pathways can produce premature anterior motion in the rear wall, while right septal APs can show motion abnormalities similar to those described in patients with LBBB.

In a recent study, a significant reduction in systolic function was observed in patients with right septal AP compared to patients with right or left free wall AP. Other series have confirmed that systolic function normalizes after ablation or spontaneous disappearance of preexcitation through the right septal pathways.

This case illustrates how ventricular preexcitation through a right antero-septal AP can cause heart failure secondary to left ventricular dysfunction and intraventricular asynchrony, similar to the effect of LBBB. Ablation of the AP subsequently improved heart failure symptoms and normalized myocardial contractility.

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