Acute Heart Failure in an Adult Patient With 2:1 Atrial Flutter: Zebras or Horses?

To the Editor,

We present here the case of a 47-year-old patient with no past medical history of interest who went to the Accident and Emergency Department suffering from palpitations, progressive dyspnoea and orthopnoea. Examination revealed a II/IV systolic murmur; bibasal crackles; preserved and symmetrical pulses, albeit weak; blood pressure of 100/70 mmHg and cardiomegaly on chest X-ray. An electrocardiogram revealed 2:1 atrial flutter with a ventricular rate of 150 bpm. The emergency room, based on an initial diagnosis of suspected ‘tachycardiomyopathy’, administered digoxin, beta blockers, diuretics, and supplemental oxygen, thereby controlling the ventricular rate (75–100 bpm) with rapid clinical improvement. The next day, an echocardiography was performed that revealed left ventricular dysfunction with ejection fraction of 35%, dilated aortic root, moderate aortic insufficiency, and an abnormal mitral subvalvular apparatus with very elongated chords with a myxoid appearance with moderate mitral regurgitation (Fig. 1).

Given the new data, a multislice CT was performed, which showed a 49 mm aortic root, severe aortic coarctation (Fig. 1) and left atrial thrombus with no coronary abnormalities. Reviewing the basic diagnostic tests, the chest X-ray showed small costal notches that had gone unnoticed. The final diagnosis was left ventricular dysfunction by severe aortic coarctation with associated lesions of aortic and mitral insufficiencies. The rhythm became atrial fibrillation and cardioversion was not indicated by the left atrial thrombus. Cardiac catheterization was performed in an attempt to interrupt the aortic arch gradient of 40 mmHg. A radiofrequency catheter was used to get past the coarctation, allowing subsequent dilation and stenting (Fig. 2) with a disappearance of the gradient. At six months, the patient was asymptomatic, with an ejection fraction of 55% and the patient remained in sinus rhythm and asymptomatic with slight mitral regurgitation.

‘Zebras or horses?’ This case clearly illustrates one of the basic axioms of medicine: “when you hear hoofbeats behind you, it is almost certainly the sound of horses and not zebras.” This can be translated as “what is most frequent is most likely.” The presence of 2:1 atrial flutter with severe ventricular dysfunction in a young man without any past medical history made us think of the most obvious: dilated cardiomyopathy secondary to tachycardia. However, the echocardiogram revealed valvular disorders (pronounced myxoid degeneration of the mitral chordae and aortic annuloectasia). A subsequent multislice CT revealed another previously undetected disorder (severe aortic coarctation). Reviewing the medical history, blood pressure was normal (which can be due to heart failure), and femoral pulses were present and symmetrical in the arms (possibly due to collateral circulation). Only after a careful review of the X-rays were the small costal notches observed. Coarctation of the aorta constitutes 6% of congenital heart disease in childhood and 15% in adulthood.1 Its clinical manifestations depend on severity: in mild cases the manifestations do not appear until adulthood, usually with the discovery of hypertension. In our patient, however, there were no disorders until the onset of

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Figure 1. Left: echocardiography showing linear images within the left ventricular cavity corresponding to very elongated mitral chordae (1). Right: aortic CT with striking collateral circulation (2), an image of aortic interruption (3), and dilation of the aortic root (4).
acute heart failure. Percutaneous treatment with dilation of the coartation and stenting was decided upon, with good results.²

In our area, more than 90%–95% of heart failure cases are due to ischemic heart disease, hypertension, arrhythmias and valvular disease, but on occasion, as with this patient, the unexpected may happen. In this case, and contrary to all statistics and initial data, the sound of hoofbeats were those of a zebra and not of a horse.

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Transapical Technique as an Alternative Approach to Paravalvular Leak Closure

Vía transapical como alternativa para el cierre del leak paravalvular

To the Editor,

Paravalvular leaks occur in between 3% and 7% of cardiac valve prostheses. As many patients in whom closure is indicated are at high surgical risk, it is often necessary to resort to percutaneous techniques. Such approaches are complicated by the lack of specifically designed materials. Nevertheless, results continue to improve as a result of advances in imaging techniques and the development of new devices. Anterograde or retrograde approaches can be used according to the location of the prosthesis and the leak. Despite the use of stiff guidewires or looped guidewires to establish arteriovenous loops, advancing the introducer for the dispensing catheter through the leak is usually the most complicated step.

We present a case in which a transapical technique1,² was used when both anterograde and retrograde approaches failed. This new technique allowed rapid and straightforward access to the leak, as well as providing good support for advancement of the introducer.

The patient had aortic and mitral mechanical valve prostheses and had been admitted to hospital three times for acute pulmonary edema. A paravalvular leak estimated by transesophageal echocardiography (TEE) to be 8 mm high, 6 mm long, and 4 mm wide was observed in the posteroseptal region and was associated with severe mitral insufficiency and signs of hemolysis. As the patient had a logistic EuroSCORE of 32%, a decision was taken to attempt percutaneous closure. The first attempt employed an anterograde approach via transeptal puncture. Despite the use of stiff guide wires, no catheters could be introduced that would allow positioning of the device. In the same procedure, a retrograde approach was also attempted. When the catheter was advanced to the left ventricle through the aortic valve prosthesis, one of the discs of the prosthesis was repeatedly blocked, leading to severe hemodynamic deterioration. A decision was therefore taken to attempt a transapical procedure.

Forty-eight hours later, the procedure was done under general anesthesia with TEE guidance. After localization of the apex by transthoracic echocardiography, a left anterior minithoracotomy

Figure 2. Left: aortography showing aortic coarctation or interruption (5). Right: aortography after stenting, showing a normal aortic calibre (6).