Early Complete Regression of Multiple Cardiac Tumors Suggestive of Cardiac Rhabdomyomas

To the Editor:

Primary cardiac tumours are infrequently seen in paediatrics, with an established prevalence between 0.0017% and 0.28% in post-mortem studies. During the development of the foetus, a prevalence of 0.14%\(^1\) has been described. Rhabdomyoma is the most frequent cardiac tumour in infants and children, comprising more than 60% of all cases.\(^2\) It generally presents as small multiple tumours that are usually located in the ventricular myocardium, although cases affecting the atrium have also been described. It is intimately associated with tuberous sclerosis, with percentages that vary between 60% and 80% according to published studies.\(^3,4\) In a parallel way, presence of cardiac rhabdomyomas has been described in between 43% and 72% of all patients with a confirmed diagnosis of tuberous sclerosis. Physical examination of the patient can reveal a heart murmur, a reduction in peripheral pulse or cyanosis. The appearance of cardiac arrhythmias is not infrequent, and has been described with a higher incidence of Wolf-Parkinson-White syndrome. In more than half of the cases, it spontaneously remits after weaning.\(^5\) We present the case of a patient with an echocardiographic diagnosis of suspected multiple rhabdomyomas with spontaneous, complete early remission.

A 1-day-old newborn was referred to a paediatric cardiology consultation due to a heart murmur. Gestational development had been uneventful. The patient was asymptomatic from a cardiovascular point of view. Cardiac auscultation detected a systolic heart murmur in the LSE retaining a second tone. The rest of the physical examination was not significant. ECG was in sinus rhythm with no significant findings. The echocardiography revealed multiple cardiac tumours located in the apex of the right ventricle, the interventricular septum and the left ventricular outflow tract (Figure 1A) where the largest tumour (9×8 mm, Figure 1B) was located, producing a slight obstruction (maximum estimated systolic pressure gradient at 23 mm Hg). The systolic function of the left ventricle was conserved. The diagnosis of suspected multiple cardiac rhabdomyomas was given, and a conservative course of action with clinical follow-up was determined for the patient. Tuberous sclerosis screening was completed and the criteria for its diagnosis were met. Six weeks later, the echocardiographic study was repeated, and it showed the disappearance of all of the cardiac tumours described above (Figure 2A), including the largest one, which had been located in the left ventricular outflow tract (Figure 2B). The patient remained asymptomatic.
Letters to the Editor

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We were extremely interested in the recent article by Mingo et al., which analyses delays before reperfusion, its determining factors and its prognostic repercussions in 389 patients with acute ST-elevation myocardial infarction (STEMI) who received a primary percutaneous coronary intervention (primary PCI) in a Spanish hospital between 2005 and 2007. The purpose of this letter is to reflect on the different aspects of primary PCI in elevated ST segments.

Firstly, the distribution of coronary syndromes is not uniform throughout the day; rather, it undergoes rhythmic variations. It has been clearly demonstrated that the beginning of an acute myocardial infarction most frequently occurs in the early hours of the morning, which has raised our interest in finding out what causes explain this circadian pattern, and what their clinical and therapeutic implications are.

Secondly, various studies have provided data regarding the poor clinical results obtained by primary PCI performed outside normal working hours (between 17:00 and 8:00).

In particular, Henriques et al. demonstrated that a total of 1,702 patients with STEMI who came to the hospital between 18:00 y and 8:00 had a higher mortality index and a greater rate of primary PCI failure. Furthermore, in a cohort study of 102,086 patients from the National Registry of Myocardial Infarction (NRMI-USA), Magid et al. demonstrated that in patients who presented STEMI during the night, it took longer to perform a primary PCI than a fibrinolysis. The same authors concluded that 33,647 patients that were treated with primary PCI had a higher hospital mortality rate when symptoms began at night, in relation with the longer delay of reperfusion.

In a similar way to the above studies, our group recently published the results of STEMI patients who were treated with primary PCI between the hours of 8:00 and 18:00 and between 18:00 and 8:00. We studied 90 consecutive patients with STEMI who had been treated with primary PCI, and the results obtained were worse when the PCI was performed between 18:00 and 8:00. On the other hand, other studies have not shown the same effects.

REFERENCES


Figure 1. Two-dimensional echocardiography images in apical 4-chamber view (A) and short axis at the level of the left ventricular outflow tract (B) in which we observe the presence of multiple tumourous masses, with a higher echogenicity than that of the adjacent myocardium, compatible with multiple cardiac rhabdomyoma.

Figure 2. Echocardiographic examination 6 weeks later with views that are superimposable on Figure 1, showing total spontaneous regression of the tumourous masses.