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.

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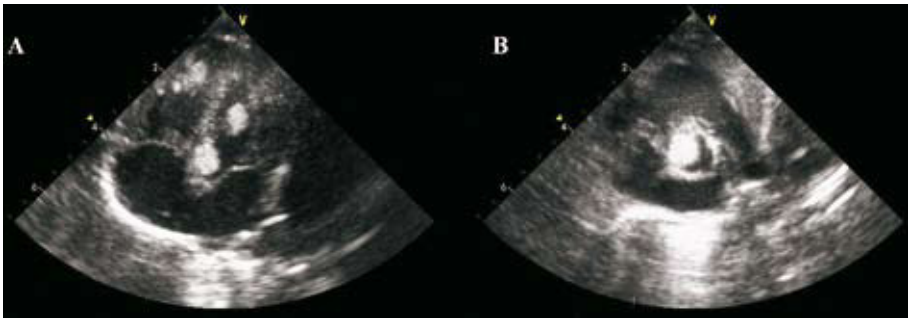

Figure 1. Two-dimensional echocardiography images in apical 4-chamber view (A) and short axis at the level of the left ventricular outflow tract (B) in which we observe the presence of multiple tumourous masses, with a higher echogenicity than that of the adjacent myocardium, compatible with multiple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

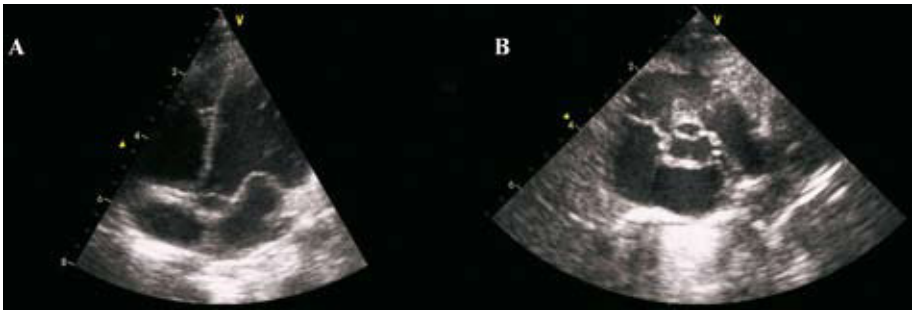

Figure 2. Echocardiographic examination 6 weeks later with views that are superimposable on Figure 1, showing total spontaneous regression of the tumourous masses.

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