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Regression of cardiac rhabdomyomas producing severe aortic stenosis. Case Report

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INTRODUCTION

Fetal cardiac rabdomayomas (R). even large and multiple, are known to regress spontaneously after birth.
We report an unusual case followed-up since the prenatal age till the first decade of life.

MATERIAL AND METHODS

A 30 years old healthy women at her 2nd pregnancy, was sent at 32 weeks gestation (wg) for echocardiography for a finding at obstetrical scan of a mass in the heart. No familiar or obstetrical scan problems were reported.

RESULTS

A voluminous echodense mass was evident in the inlet left ventricle (LV) close to the mitral valve and a smaller mass protruding into the aortic outflow with a small free space around the valve and accelerated aortic flow, with pulsed Doppler velocity at upper limits. Mitral valve showed slightly reduced flow. No other nodes were visualized. The LV contractility was normal.

Diagnosis of multiple R was done, without abnormal findings in other fetal organs.
CNS MRI was postponed. At 36 w.g. a cesarean section was done fearing a progression of the masses.

A girl baby was born in good general conditions, birth weight 3.150 kg, Apgar 8-9.
The appearance of the mass in the LV inflow was unchanged, while a smaller mass in the aortic area obstructed aortic flow with maximum gradient 50 mmHg that remained unchanged thereafter. There were no rhythm problems. Neurological evaluation was normal, CNS MRI did not show specific lesions and tuberous sclerosis was excluded by negative genetics. No immediate surgery was considered necessary and the baby was followed up closely, the aortic gradient remaining stable.

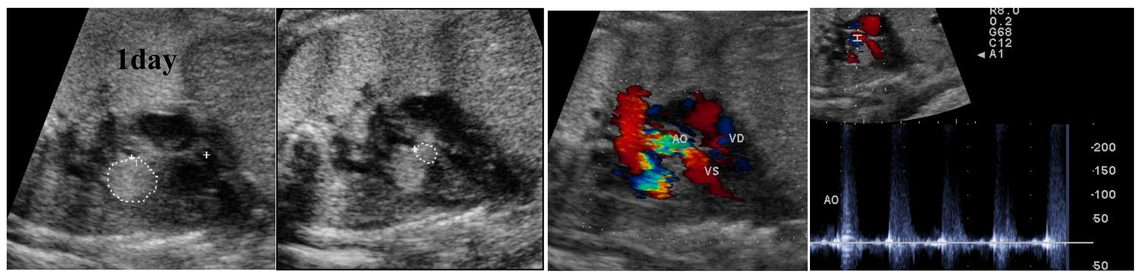
FOLLOW-UP **The large cardiac mass progressively regressed throughout the years, as well as the aortic mass, but with apparently damaged aortic valve.**

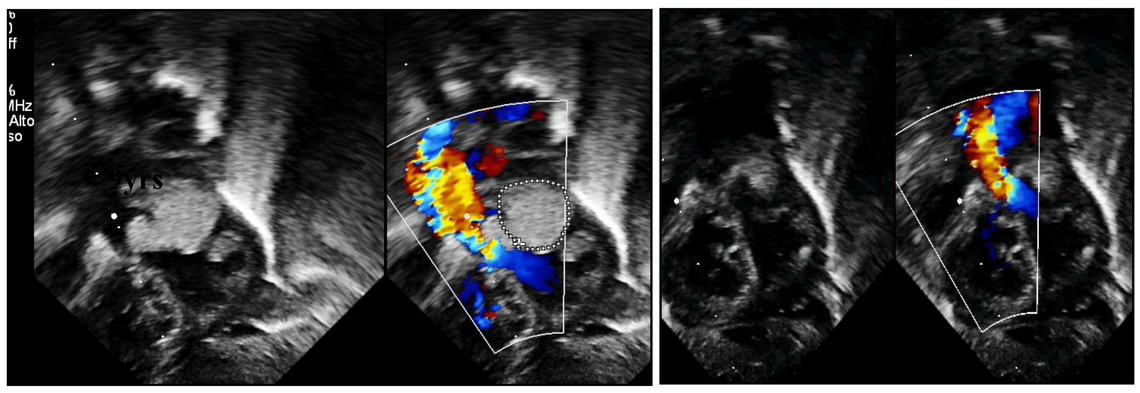
At 9 years the LV mass almost disappeared but the aortic valve looked more severely damaged, with increased aortic gradient reaching 87-100 mmHg.

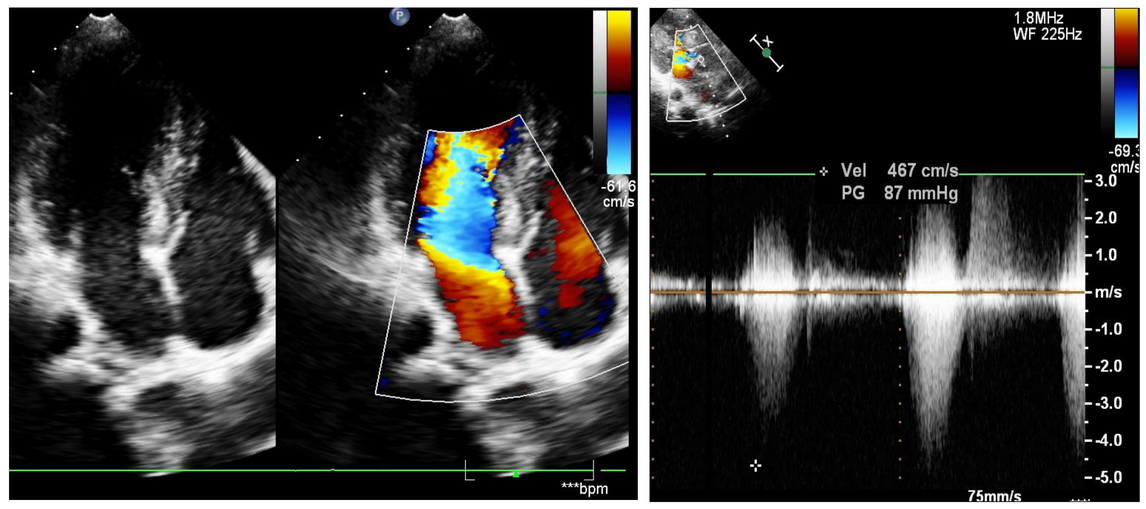
Therefore it was decided to plan a correction. It was possible to perform a plasty of the valve resecting a fibrous circumferential rim and removing the residual mass.

Afterthen only mild aortic regurgitation and mild systolic gradient of 20 mmHg with good LV function remained. The girl is now 12 years old and well.

ECHOCARDIOGRAPHY







COMMENT

The voluminous mass in the inflow and mainly in the aortic tract was very warrying as for the obstructive pattern in aorta, however the postnatal aortic gradient in aorta was not apparently creating hemodynamic problems, which allowed us to proceed only with a straight follow-up. The mass both in the inflow and outflow tracts progressively diminished in the years, as is known from the natural history of R. In fact, at 9 yrs. only a small residual echogenic mass was present in the iv septum, and around the aortic valve, that became however more dysplastic and damaged, with increased gradient. The LV function and cardiac compensation were reasonable, however we decided to proceed to a correction of the aortic problem.
The surgeons were prepared to perform the Ross procedure, however at the operative table it was possible to perform a plasty of the aortic valve, removing the small remnant mass around.

CONCLUSIONS

Our case is unusual showing a progressive spontaneous regression of R masses but with a subsequent progressive development of severe aortic valve stenosis.-.