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PRENATAL DIAGNOSIS AND CLINICAL MANAGEMENT OF FETAL INTRA-CARDIAC RHABDOMYOMA : ABOUT ONE CASE AND REVIEW OF LITTERATURE

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Introduction

Fetal intracardiac tumors are exceptionally rare, with an incidence estimated at 0.14% of pregnancies, and less than 0.03% in autopsy series. In ultrasound-based studies, the incidence is slightly higher, reported at approximately 0.3%. Among these tumors, rhabdomyoma is the most frequently observed benign cardiac tumor, representing nearly 80% of cases according to large series such as those from Toronto. These tumors are commonly diagnosed in the antenatal period due to advances in prenatal imaging, particularly ultrasonography. Although benign in nature, cardiac rhabdomyomas may lead to significant hemodynamic compromise depending on their size and location, and they are strongly associated with tuberous sclerosis complex (TSC). We present a case of prenatally diagnosed intracardiac rhabdomyoma, followed by a discussion of clinical management and review of the litterature.

Case presentation

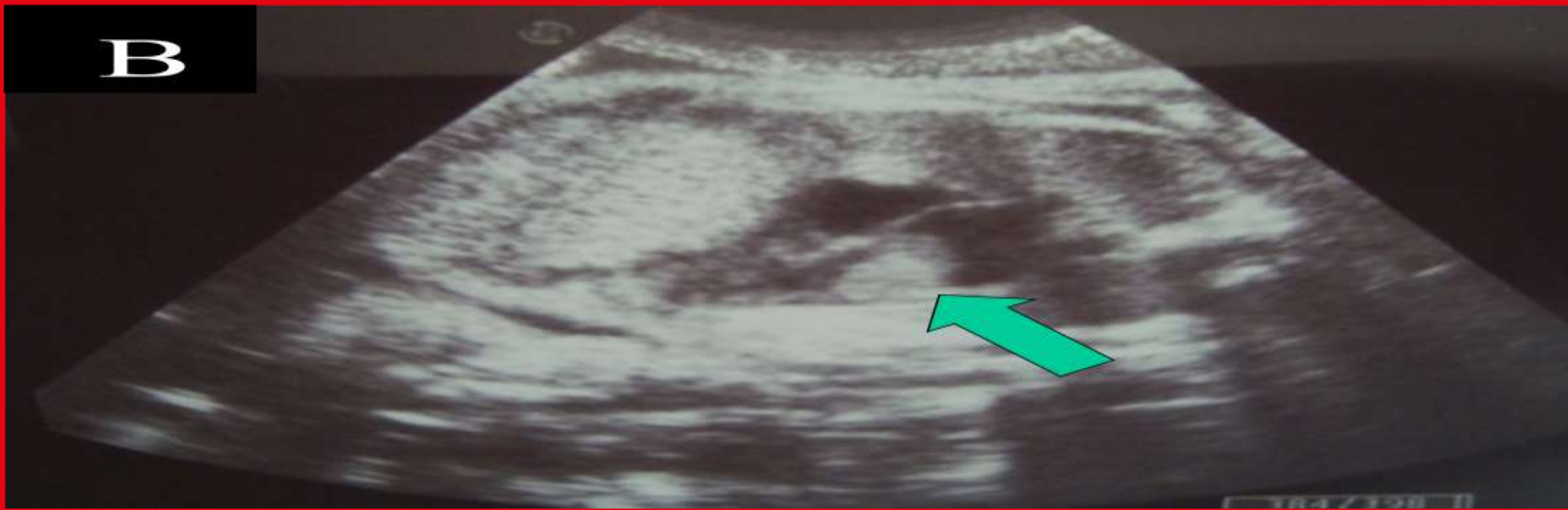
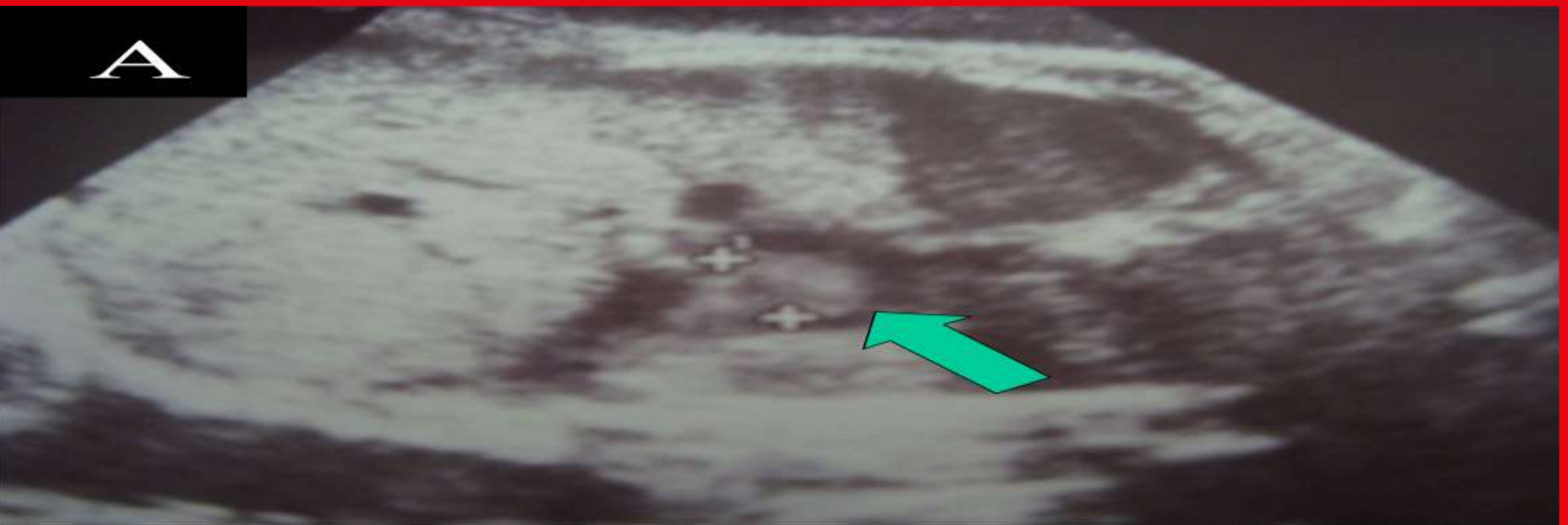
A 28-year-old primigravida was referred for routine fetal ultrasound at 32 weeks of gestation. Examination revealed multiple intracardiac masses, the largest of which was a multilobulated, mobile lesion arising from the lateral wall of the left ventricle and extending across the mitral valve. Despite the size and location of the mass, no signs of fetal cardiac failure or hydrops were observed. The diagnosis of rhabdomyoma was suspected based on the echogenic, well-circumscribed nature of the masses. Both parents underwent dermatological, neurological, and ophthalmological evaluations to assess for features of tuberous sclerosis complex, all of which were unremarkable. The pregnancy was monitored closely, and spontaneous preterm delivery occurred at 36 weeks of gestation. Postnatal echocardiography confirmed the presence of a 22 mm mass attached to the lateral wall of the left ventricle, significantly impeding mitral valve function. Surgical excision of the tumor was performed, and the postoperative course was favorable, with no recurrence or immediate complications.

Discussion

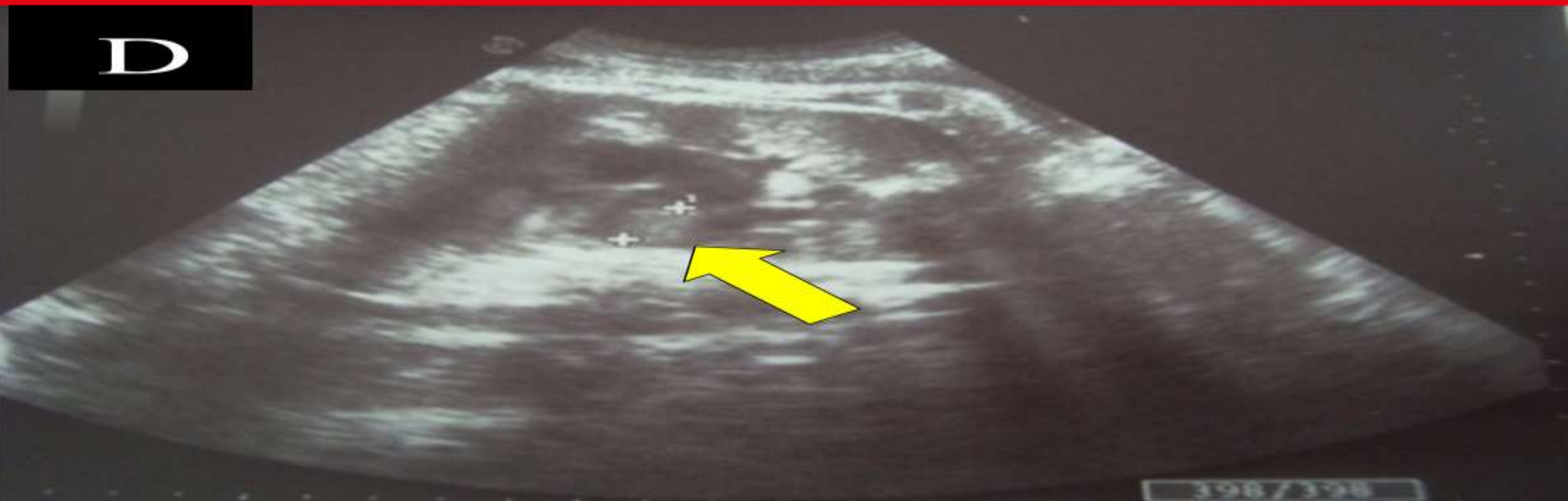
- Fetal cardiac rhabdomyomas account for the majority of intracardiac tumors identified during pregnancy and are considered benign hamartomas of striated muscle origin.
- Their natural history is variable, with some lesions regressing spontaneously after birth, while others, depending on size and anatomical location, may cause obstruction of blood flow, arrhythmias, or heart failure.
- Prenatal detection is essential for planning perinatal management. In this case, the tumor's large size and obstruction of the mitral valve highlighted the importance of close surveillance and delivery in a tertiary care setting with pediatric cardiology and cardiac surgery facilities.
- The strong association between rhabdomyomas and tuberous sclerosis complex (reported in up to 50–80% of cases) underlines the need for thorough parental evaluation and, when available, fetal MRI to detect associated cerebral lesions.
- Nevertheless, the absence of MRI findings does not exclude TSC, and postnatal follow-up remains crucial.
- From a management perspective, most asymptomatic cases are managed conservatively, as spontaneous regression is common. However, when hemodynamic compromise or obstruction occurs, as in our case, surgical intervention is indicated.
- Prognosis is generally favorable when prompt diagnosis and appropriate management are undertaken.

Conclusion

- Intracardiac rhabdomyoma is the most frequent fetal cardiac tumor and can be diagnosed antenatally by ultrasound. Although benign, these tumors may cause severe complications depending on their size and location.
- Prenatal diagnosis allows multidisciplinary planning, including genetic counseling for tuberous sclerosis and preparation for potential surgical intervention.
- Comprehensive management combining prenatal imaging, neonatal evaluation, and timely surgical treatment when indicated ensures the best outcomes for affected infants.



Figures A and B. Presence of a 20 mm tissue mass attached to the lateral wall of the left ventricle, prolapsing through the mitral valve (arrows).



Figures C and D. Presence of multiple masses: one located on the lateral wall of the right ventricle (white arrow) and another on the lateral wall of the left ventricle (yellow arrow).