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Rare complicated presentation of a large invading cardiac rhabdomyoma in a 29 week of gestation fetus

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Introduction: Rhabdomyoma is the most common cardiac tumor in children. Depending upon the location and size of the tumor, patients can present with a broad spectrum of clinical signs and symptoms, however the clinical course is generally benign, and it has been well documented that these tumors tend to regress over time.

Case: We are reporting an unusual case of a preterm baby diagnosed at the age of 29 weeks of gestation to have a very large pericardial effusion with bradycardia and evidence of anhydramnios. The mother underwent an emergency caesarian section a week later. The baby born with an APGAR score of 2 at 1 minute and 7 at 5 minutes required cardiorespiratory resuscitation and intubation. His birth weight was 1.5 kilogram and he did not have dysmorphic features. Chest radiography showed wall to wall heart and a 12 leads electrocardiogram showed generalized low voltages, bradycardia with heart rate of 80 - 90 beats per minute with an abnormal P wave axis. Echocardiogram showed a very large circumferential pericardial effusion with a significantly bright thickening of the right and left atrial walls including both atrial appendages with findings of secundum atrial septal defect and a small patent ductus arteriousus. The findings were assessed also with cardiac Magnetic Resonant Imaging (MRI) and cardiac Computing Tomography (CT) scan. Five days post delivery, the baby underwent a median sternotomy with operative findings of large clear pericardial effusion with unresectable lobulated tumor mass almost involving the right atrium and left atrium extending to both atrial appendages. The histopathology result with immunostains showed a several large vacuolated cells seen in large cords of small clusters. These cells show a bland, mostly uniform medium sized nucleus with no mitotic activity and are seen intimately admixed with the normal muscles tissue throughout the biopsy with finding consistent with rhabdomyoma. The baby developed episodic paroxysmal ectopic supraventricular tachycardia and managed medically. The tumor demonstrated progressive regression in size and the patient developed clinically features of proteus syndrome.

Conclusion: We report an unusual presentation of a 29 week of gestation fetus with large intramural cardiac rhabdomyoma involving the supraventricular area of both atria and atrial appendages without intracavity extension affecting the cardiac conduction system and causing supraventricualr tachycardia with large pericardial effusion. The patient developed features of proteus syndrome with a report of a rare association with cardiac rhabdomyoma.

Biography

Khalfan S Al Senaidi is working in the Division of Pediatric Cardiology since July 2006 at Sultan Qaboos University Hospital, Oman. His main research interest is pediatric cardiology.

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