Journal of College of Medical Sciences-Nepal, 2012, Vol-8, No-4, 51-54 *Case Report*

Angiofibroma, a rare cardiac tumour in children

G Gayen,[1] A Pan,[2] K Dasbakshi,[3] P Mukherjee[4]

[1]RMO Cum Clinical Tutor, Department of pediatrics, National Medical College, Kolkata. [2]Lecturer Department Of Paediatrics, college of Medical Sciences,Bharatpur, Nepal . [3]Assistant Professor, [4]Professor, Department of cardiothoracic surgery, National Medical College, Kolkata

Abstract

Angiofibromas, located in any other sites than nasopharynx are unusual. Cardiac angiofibromas are a very rare cardiac tumours in comparison to rhabdomyomas which are the commonest in the children. We report a right ventricular tumour in a10 year old girl which was excised under cardiopulmonary bypass successfully and diagnosed as angiofibroma on histopathology.

Key words: Angiofibroma, cardiac, children.

Introduction

Angiofibromas are benign but locally aggressive vascular tumours that occurs in nasopharynx, neck and face. These are rare soft tissue hamartomas which contain mainly blood vessels and fibrous tissues often found in association with tuberous sclerosis (TS). Angiofibromas located in other sites than head and neck regions are rare and as cardiac tumours in children these are even rarer in comparison to rhabdomyomas which are the commonest in children. We report a case of cardiac angiofibroma in a 10 years old girl who presented with frequent chest pain and fatigue for the last two years. Echocardiography with Doppler study showed a space occupying lesion in the right ventricular outflow tract. It was operated and found to be angiofibroma on histopathology. The child is now well on follow up.

Case report

A 10 year old girl was admitted in our department with the symptoms of frequent chest pain and fatigue and shortness of breath for the last two years. There was no history of cyanosis, or easy fatigability after birth. There was no such family history in the parents and siblings. There was no history of mental retardation, seizure disorder, any skin lesion suggestive of TS. She was normotensive, with a pulse rate of 80 per minute. There was right ventricular heave and an ejection systolic murmur. Electrocardiogram showed right ventricular strain pattern. Chest X ray showed normal cardio thoracic ratio. Haemogram and blood biochemistry were within normal limits. Echocardiography with Doppler study showed a space occupying lesion in the right ventricular outflow tract moving in and out of the RVOT in systole and diastole

Correspondence: Dr. Gargi Gayen
E-mail: dr.gargi07@gmail.com

and was diagnosed as a pedunculated right ventricular (RV) tumour. Gradient across the pulmonary valve was 80 mmHg (systolic). Pediatric cardiologist and cardiothoracic surgeon were consulted and the tumour was excised. Median sternotomy was done, aortic and bicaval cannulation made to put the patient on cardiopulmonary bypass. Aorta was cross clamped, cardioplegic arrest of heart achieved and pulmonary arteriotomy was made. The solid tough pedunculated 3cm X 2 cm rounded mass was excised from the subvalvular area below the pulmonary artery. It was attached to the inter ventricular septum near the right ventricular outflow tract with a base which was about 0.5 cm wide. It was completely excised and there were no residual tumour anywhere else. The clamp time was 15 minutes. Weaning off bypass, decannulation and closure and recovery was uneventful. The histopathology was reported as angiofibroma.

The patient is doing well after a one year follow up. Echocardiography done 8 months after operation was normal without any right ventricular obstruction.

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Figure 1.Echocardiography showing the mass

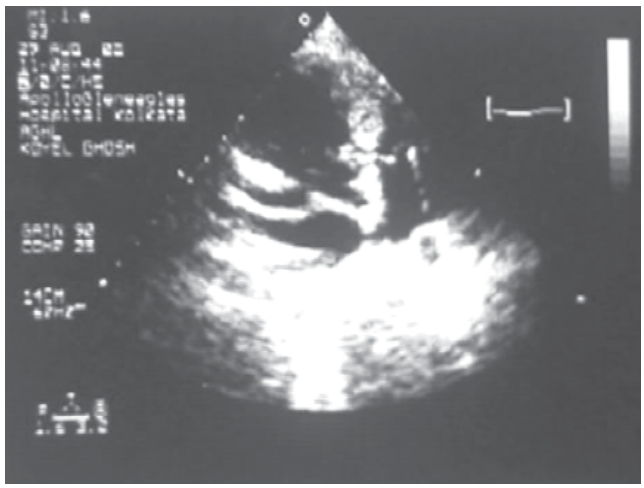


Figure 2: Echocardiography with colour doppler showing the mass

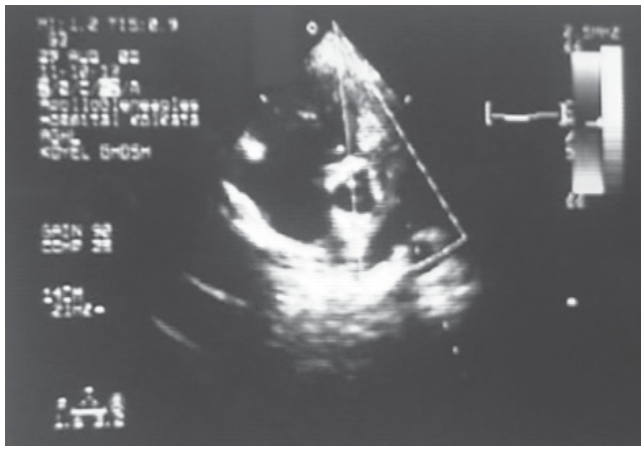
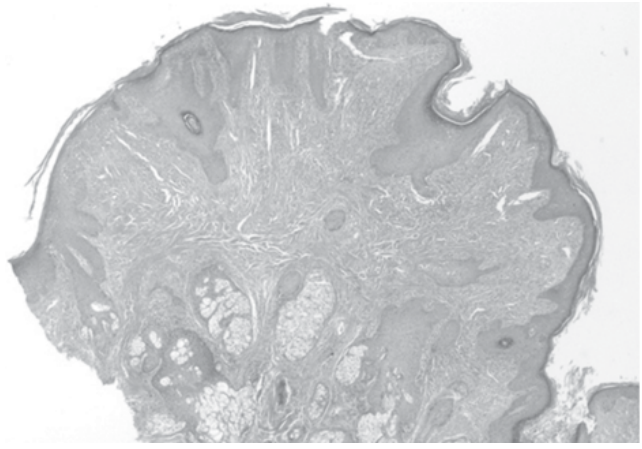


Figure 3: Excised tumour mass

Figure 4: Histopathology of the excised mass



Discussion

Primary cardiac tumors are rare in children. The majority are benign and noninvasive, although they may have significant hemodynamic consequences depending

on their location. By far, the most common type of tumors reported in children and adolescents is rhabdomyoma,and in 50 % case it is associated with tuberous sclerosis complex.[1]

Other benign tumours are fibroma, myxoma, and teratoma.[2] Sarcoma is the largest group of primary cardiac malignant neoplasms.[3] Angiofibroma is rare to present as cardiac tumour. Its usual location is mainly the nasopharynx and head and neck region where it presents with nasal obstruction and nose bleeds. It is most common in adolescents male(peak age15 yrs) and thus is sometimes referred to as a *juvenile nasopharyngeal angiofibroma*.[4] Under the microscope, these tumors are well-circumscribed but unencapsulated and composed of collagenized fibrous stroma with vessels ranging from small slit-like spaces to dilated lumens. No significant cytologic atypia is observed. Occasional multinucleated stromal cells are present.[5] The vascular nature is highlighted by angiograms showing a rich supply. Cardiac angiofibroma is very rare, Previously only two cases were reported as epicardial angiofibroma in a fetus with Beckwith-Wiedemann syndrome[6] and angiofibroma of the tricuspid valve as a presentation of the tuberous sclerosis complex[7] TS is usually associated with facial angiofibroma or adenoma sebaceum.[8] In our case the tumour was located in the RVOT. There was no associated features suggesting tuberous sclerosis, it was a sporadic case. The clinical presentation of a patient with a cardiac tumor is determined more by the tumor's location than by its histologic type, varying widely from asymptomatic presentations to life-threatening cardiac events by critical obstruction of a valve or outflow tract and causing arrhythmia. Because of the rapid growth of tumor mass in small-sized cavities, congestive heart failure has been frequently reported in childhood and surgical treatment is lifesaving. Right-sided tumors may present with congestive heart failure (CHF) manifested by fatigue, edema, jugular venous distention, and ascites. Other symptoms include shortness of breath, syncope, and night sweats. Pericardial effusions may occur. Vena cava syndrome, pulmonary embolism, and restrictive cardiomyopathy are some of the complications. Left atrial and left ventricular tumors can present with fever, chills, dizziness, dyspnea on exertion, cold sweats during exercise or at night, and nonproductive cough. Because tumors may embolize, they also can lead to seizures, transient ischemic attacks, and cerebrovascular and peripheral-vascular accidents. Based on their size and position, they may induce arrhythmias and interfere with ventricular compliance. Our child presented with the features of right sided outflow tract obstruction. Echocardiography with doppler is an excellent tool to diagnose the tumour and gradient if any obstruction. However, precise information on the tumor histotype is not attainable by echocardiography.[9] A correct therapeutic plan requires an accurate histopathologic diagnosis of the resected mass to rule out the rare case of a malignancy, CT scan, MRI , ECG can add to the diagnosis. In our child ECG, CXR were done and there was no rhythm abnormality.The mode of treatment varies and cannot be easily simplified because the kind of tumor (benign/ malignant, infiltrative/ localized) dictates therapy. Location and extent of the tumor, as well as symptoms, are clinical variables that direct treatment. Observation is sufficient when the mass is small and does not interfere with vascular hemodynamics. However, when the tumor is causing hemodynamic problems by either obstructing the outflow tract, valve or causing rhythm

abnormalities, aggressive management by surgical intervention should be done. In occasional cases, implantation of a pacemaker may be needed if atrioventricular block occurs. In our child it was excised successfully under cardiopulmonary bye pass, On follow up the child was normal and repeat echocardiography after 8 months show no further obstruction of RVOT.

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