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CASE REPORT

Rare Intracardiac Aneurysms seen in Jos, Nigeria: A Report of two Cases

Anevrysmes Intracardiaques Rares Vus à Jos, Nigeria: A Propos de 2 Cas

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ABSTRACT

BACKGROUND: **Some congenital heart diseases are compatible with early life presenting their peculiar challenges in adulthood. Some of them are particularly rare, and are misdiagnosed in the absence of modern imaging facilities. Intra – cardiac aneurysms fall into this group.**

OBJECTIVE: **The clinical presentation of the cases up to point of sudden death were documented and echocardiography done.**

RESULTS: **Two such cases which were never encountered in over 20 years of echocardiography in our unit are reported. One was a ruptured sinus of Valsalva aneurysm dissecting the interventricular septum and the other ventricular septal defect aneurysm involving the tricuspid valve.**

CONCLUSION: **Some congenital heart diseases present late. Increased availability of echocardiography and skilled personnel should lead to early diagnosis preventing fatality that follows late diagnosis as occurred here. WAJM 2013; 32(1): 76–78.**

Keywords: **Aneurysm, Rare, Congenital, Heart, Sudden death**

RÉSUMÉ

CONTEXTE: LaSéquence de perfusion artérielle inversée chez **WAJM 2013; 32(1): 76–78.**

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INTRODUCTION

Some congenital cardiac abnormalities can be survived into adulthood. They however present diagnostic challenges in the absence of cardiac imaging facilities or else are picked up at necropsy. Apart from congestive cardiac failure, such patients are at high risk for infective endocarditis[1] and rupture. We hereby report two cases of intra-cardiac aneurysms encountered in 2009, the types of which we never encountered in our 20 years of echo-cardiography. They are as interesting as they are rare; and continue to highlight the need for modern cardiac imaging facilities in all cardiology services in our environment. Early diagnosis would give time to mobilize resources for surgery overseas since they are not available locally. Late diagnosis by which time they are already in heart failure and close to catastrophic rupture seems to be the norm for now and should be prevented.

CASE 1:

This was a 30-year-old man who was refered from the staff clinic of a College of Education in Kaduna State. He belonged to the Eggon ethnic group of Nassarawa State. He was said to have presented five months earlier with upper abdominal pain and breathlessness. The finding of a murmur necessitated his being refered to our service. He gave a history of insidious onset of dyspnoea on exertion which progressed to dyspnoea at rest. There were associated orthopnoea and paroxysmal nocturnal dyspnoea. Other symptoms were palpitations, cough with frothy blood tinged sputum, body swelling, right hypochondrial pain and early satiety. There was no history of chest pain, syncope or pre-syncope. He had never smoked nor drank alcohol and had no history suggestive of childhood heart disease or rheumatic fever. There was however a family history of hypertension in his father and two uncles. A relation had aortic incompetence secondary to aortic aneurysm.

On examination he was mildly orthopnoeic, not pale, cyanosed, jaundiced or oedematous. He however had grade two finger clubbing. Central nervous system was normal and there was no abnormal respiratory sign.

He had a bounding pulse with wide pulse pressure of 80 mmHg as blood pressure was 130/50 mmHg. Corrigan's sign was positive. The apex beat was mildly displaced at the 5th left intercostal space about 4 cm lateral to the mid clavicular line. It was heaving and no thrills were palpable. Heart sounds were normal but there was a 3/6 aortic regurgitation murmur with 2/6 mitral regurgitant murmur. Abdominal exam revealed a 6cm hepatomegaly with mild to moderate ascites.

Investigation included viral screen (HBsag, anti HCV and HIV) which all returned non reactive. Haematology and blood biochemistry results were normal. Electrocardiography showed left axis deviation, left atrial enlargement, and left ventricular hypertrophy with ischaemic changes. Chest X ray showed cardio-megaly with bi-ventricular configuration, very prominent aortic shadow, upper lobe vessel diversion and patchy opacities of both lower zones. At echocardiography, the aortic root was wide (36mm) with connection between the right coronary sinus and a space in the interventricular septum (Fig. 1).

Colour flow showed regurgitation into the left ventricle and a space in the interventricular septum during diastole (Fig. 2). The diagnosis was therefore made as aortic regurgitation following ruptured sinus of Valsalva dissecting the interventricular septum. He was counseled on the need for surgery and put on anti-failure regimen. He defaulted thereafter apparently due to financial difficulties. He died suddenly a few months later according to his relations.

CASE 2:

She was a lady in her early twenties who had a rather uneventful childhood. However, in her first pregnancy she started to have difficulty with breathing. She did not avail herself of adequate medical care till around delivery time when the managing obstetricians refered her to a medical unit as a case of rheumatic mitral valve disease (mitral incompetence) in congestive cardiac failure.

Physical examination was said to be consisted with mitral incompetence in heart failure; for which she was sent to us for echocardiography.

At echocardiography a large ventricular septal defect was seen with an aneurysm the lower border of which consisted of the tricuspid valve (Fig. 3). With this finding the managing medical team refered her to us for further management. She was said to have died suddenly while arrangements were being made to transfer her to our cardiology service.

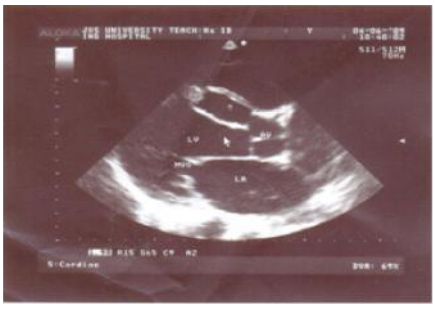

Fig. 1: Long axis parasternal view of the heart showing a connection between the right sinus of valsalva and a space in the interventricular septum (?).

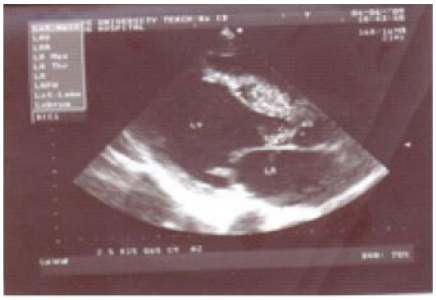

Fig. 2: Colour flow at the long axis parasternal view showing turbulence into the space created by dissection of the interventricular septum and the left ventricular out flow tract. The latter produced the aortic incompetence murmur.

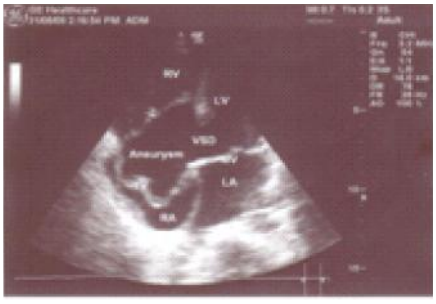

Fig. 3: A four chamber apical view of the heart showing a large VSD and aneurysm the lower border of which is formed by the tricuspid valve.

DISCUSSION

Both cases were misdiagnosed clinically until availed of echocardiography. That they all presented in congestive cardiac failure shows their late presentation. Also the sudden mode of their deaths suggest that once detected, surgical repair becomes an urgency. We lost both of them suddenly as we have no local functional cardiothoracic surgical support service; and traveling overseas for such is very demanding to our poor patients in financial terms.

Aneurysm of sinus of Valaslva is rare and no case report was encountered in medical literature from Nigeria, although cases have been reported from neighboring Cote d' Ivoire.[2] Case 1 had an uncle with aortic aneurysm suggesting a family history of cardiac malformation. His sinus of Valsalva aneurysm (Figures 1 and 2) had ruptured and was dissecting the interventricular septum as at the time of echocardiography. He died suddenly as we were informed later. This could have been the result of a malignant arrhythmia which they are prone to.[3] The involvement of the aortic valve with regurgitation which was the initial clinical diagnosis is one of the complications encountered with it.[4]

Case 2 presented even a rarer aneurysm, where the tricuspid valve formed part of an aneurysm of the ventricular septal defect. This is one of the pulmonary flow limiting cardiac morphologic alterations[5] that can prolong survival. She had been misdiagnosed in the obstetric and gynecology service as a case of rheumatic mitral incompetence. Tricuspid valve involvement is due to the anatomical relationship of the aneurysm of the membranous ventricular septum to the tricuspid septal leaflet. Specifically as in this case, the tricuspid septal leaflet formed part of the wall of the aneurysm. The tricuspid valve was adjacent to the ventricular septal defect and billowed into the right, a feature consistent typically with tricuspid tissue tags associated with perimembraneous ventricular septal defect. This used to be called ventricular septal aneurysm (Figure 3).[6] Her pregnancy and delivery may have contributed to unmasking the condition. Her sudden death may have been from spontaneous rupture of the aneurysm.

These cases are documented to further highlight the need for echocardiography in all cases of cardiac murmur.

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