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Rare disease

Intracardiac bronchogenic cyst in a 2-year-old Nigerian boy

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Summary

Primary cardiac tumours are rare in the paediatric age group. Bronchogenic cysts, although relatively rare, represent the most common cystic lesion of the mediastinum. Intracardiac bronchogenic cysts however, are extremely rare. The authors are unaware of any case previously reported in a Nigerian child and hence report the case of a 2-year-old boy for its rarity and interest. The boy was referred for evaluation of a cardiac murmur. Clinical, radiological and electrographic findings were suggestive of mild pulmonary stenosis or an atrial septal defect (ASD). 2-dimensional echocardiography however, revealed in addition to a small ASD, an intracardiac mass attached to the tricuspid valve. The mass was surgically removed and found on histology to be a bronchogenic cyst. Our experience highlights the importance of echocardiography in the evaluation of asymptomatic patients with cardiac murmurs, in whom a rare lesion might have otherwise been missed.

BACKGROUND

Primary cardiac tumours are rare in the paediatric age group, with a prevalence of 0.0017 to 0.28 in autopsy series.[1] Bronchogenic cysts, which result from anomalous development of the ventral foregut, although relatively rare, represent the most common cystic lesion of the mediastinum.[2] In children they make up 6–15% of primary mediastinal tumours.[3] These thin-walled, fluid or mucus-filled cysts are mostly encountered along the tracheo-oesophageal axis or within the lung parenchyma, with a predilection for the carinal region. They may also occur in remote sites such as the interatrial septum, neck, abdomen and retroperitoneal space.[4] They are often connected to the tracheobronchial tree, although they do not communicate with the lumen.

Bronchogenic cysts involving the heart are very rare, the intracardiac variety being extremely rare. The authors are aware of a few reports of intracardiac bronchogenic cysts occurring in adults,[5–13] involving the right atrium,[5] left atrium,[6 7] interatrial septum,[8–10] right ventricle[11 12] and left ventricle,[13] but could find only one previous report of an intracardiac bronchogenic cyst occurring in a child.[14] The only other reports of bronchogenic cysts related to the heart in children we found[15–18] were mostly of extracardiac origin and at best, intrapericardial. One of these was reportedly in the left ventricle in a 2-year-old boy,[18] but was subepicardial, not within the cardiac cavity. The authors are unaware of any case previously reported in a Nigerian child. We therefore report the case of a 2-year-old Nigerian boy with a right ventricular bronchogenic cyst for its rarity and interest.

CASE PRESENTATION

A 2-year-old boy was referred to the Paediatric Cardiology Unit of the Lagos State University Teaching Hospital for evaluation of a cardiac murmur detected during a febrile illness. Cardiac-wise, he was completely asymptomatic. His medical history was unremarkable.

Physical examination revealed a healthy-looking, well nourished, afebrile boy weighing 14 kg. Chest examination was normal. Auscultation of the heart yielded a single, soft S_2 with a grade $^3/_6$ ejection systolic murmur loudest at the left upper sternal edge and another Grade $^3/_6$ pan systolic murmur loudest at the left lower sternal edge, leading to a clinical diagnosis of ventricular septal defect (VSD) with right ventricular outflow tract obstruction (RVOTO).

INVESTIGATIONS

Chest x-ray (figure 1) showed situs solitus, laevocardia and a bulky cardiac silhouette with normal pulmonary vascular markings. ECG (figure 2) showed right atrial enlargement, right ventricular hypertrophy and right bundle branch block, raising the possibility of the presence of an atrial septal defect (ASD).

2-dimensional (2-D) echocardiography (figure 3) revealed normal intracardiac connections and a small secundum ASD. In addition, there was a 13×14 mm mass moving

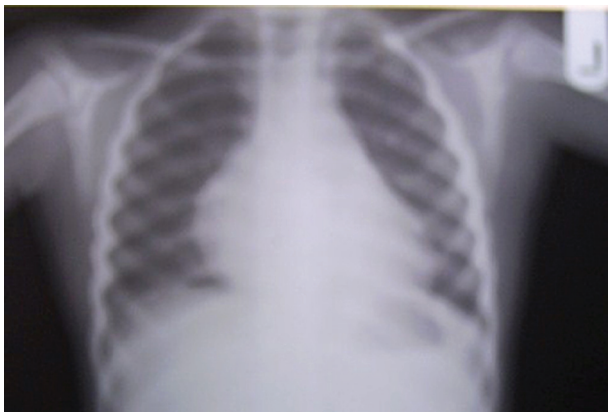

Figure 1 Chest radiograph, showing a normal shaped, though bulky cardiac silhouette.

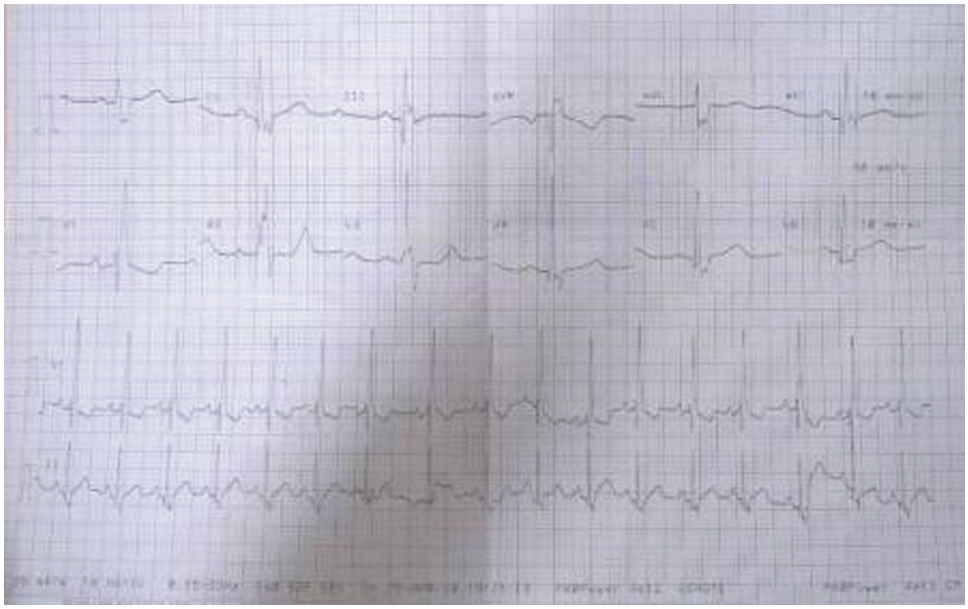

Figure 2 Electrocardiograph, showing right atrial enlargement, right bundle branch block and right ventricular hypertrophy.

with the tricuspid valve, flipping between the right atrium and right ventricle (figure 3). This was thought to be either a myxoma or a vegetation. There was also moderate tricuspid regurgitation.

DIFFERENTIAL DIAGNOSIS

- Myxoma
- Vegetation.

TREATMENT

As the options for surgical intervention in Nigeria were limited, the patient was referred to India for further evaluation and treatment. He subsequently underwent open-heart surgery at the Fortis Escorts Heart Institute, India, at which a 'large, broad-based mass attached to the interventricular septum, adherent to the septal leaflet of the tricuspid valve' was found. The mass 'caused distortion of the tricuspid valve apparatus and obstruction to the right ventricle inflow and outflow tracts'. The tumour was removed and the secundum ASD closed.

Histology of the tumour (figure 4) showed 'cystic areas lined by pseudo-stratified ciliated mucous secreting columnar lining along the surrounding muscular wall. Also seen were focal areas of cartilage, small glandular luminae, squamous metaplasia of lining. There were also areas of ulceration with surrounding connective tissue fragments with interspersed vessels. There were no ectodermal components or 3rd germ layer components seen. There were no immature or malignant components'. These findings led to a diagnosis of bronchogenic cyst.

OUTCOME AND FOLLOW-UP

He made an uneventful postoperative recovery and returned to Nigeria 8 days after the surgery. He has remained well and symptom free 1 year following the surgery, with normal parameters of cardiac function, though still with significant tricuspid regurgitation.

DISCUSSION

Bronchogenic cysts are often asymptomatic, being discovered incidentally, as was the case in our patient. They usually draw attention to themselves on account of compression on adjacent structures such as the tracheobronchial tree,[19] or the heart,[15 16] a situation which, particularly in childhood could be life-threatening. They may also be a cause of recurrent chest infections or chest pain.

In our patient, attention was only drawn to the heart by an incidental murmur discovered during routine evaluation of a febrile illness. In the only other report of an intracardiac bronchogenic cyst that we are aware of, the cyst was discovered incidentally in a 5-year-old girl during surgery for an aneurysm of the pars membranacea septi,[14] which itself was thought to be the cause of premature ventricular contractions with which the patient presented. In the case reported by Somwaru *et al*,[17] in a 3-year-old girl, the intrapericardial cyst, arising from pulmonary artery, was also discovered incidentally during surgery for a sinus venosus ASD.

In other reports we found of bronchogenic cysts related to the heart in children, the patients were mostly symptomatic. In Mosquera *et al's*[15] case, a 10-month-old girl presented with intractable heart failure. The cases of Göksel *et al*[16] and Wei *et al*[18] both presented with chest pain.

In retrospect, the auscultatory findings suggestive of RVOTO in the patient are likely to have been produced by the mass as it obstructed the outflow during systole, while the murmur suggestive of VSD was most probably due to tricuspid regurgitation consequent on the mass being adherent to it. In the similar case of a 48-year-old woman with a right ventricle bronchogenic cyst, reported by Prates *et al*,[11] the mass similarly caused RVOTO, but was not adherent to the tricuspid valve, and there was therefore no regurgitation.

The importance of echocardiography in the evaluation of children suspected to have cardiac lesions cannot be gainsaid. The discovery of the lesion in this child was unexpected, particularly as he was asymptomatic. A

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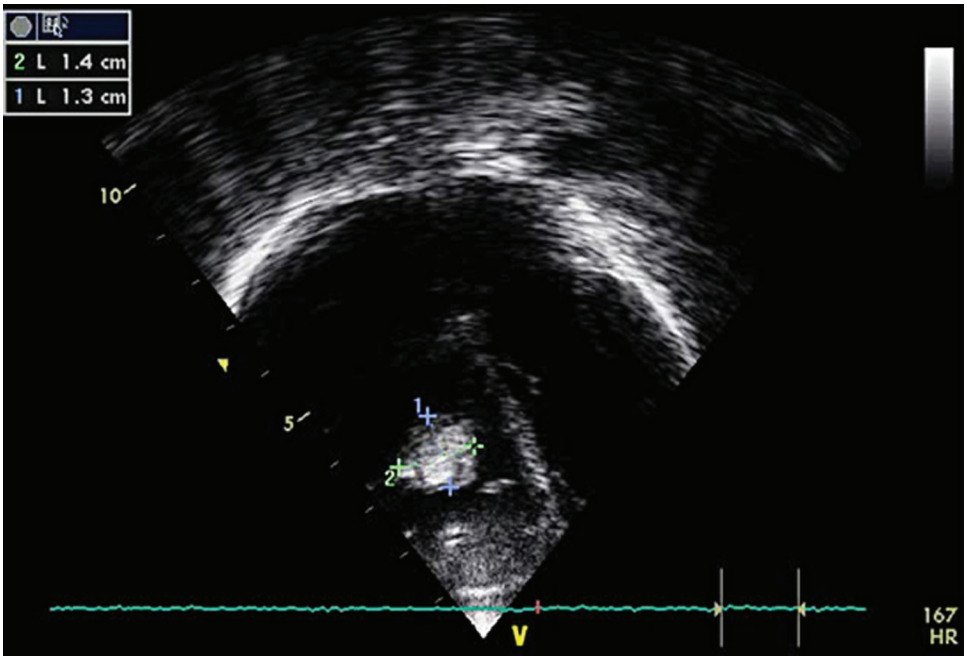

Figure 3 2-dimensional echocardiograph showing the tumour in the right ventricle.

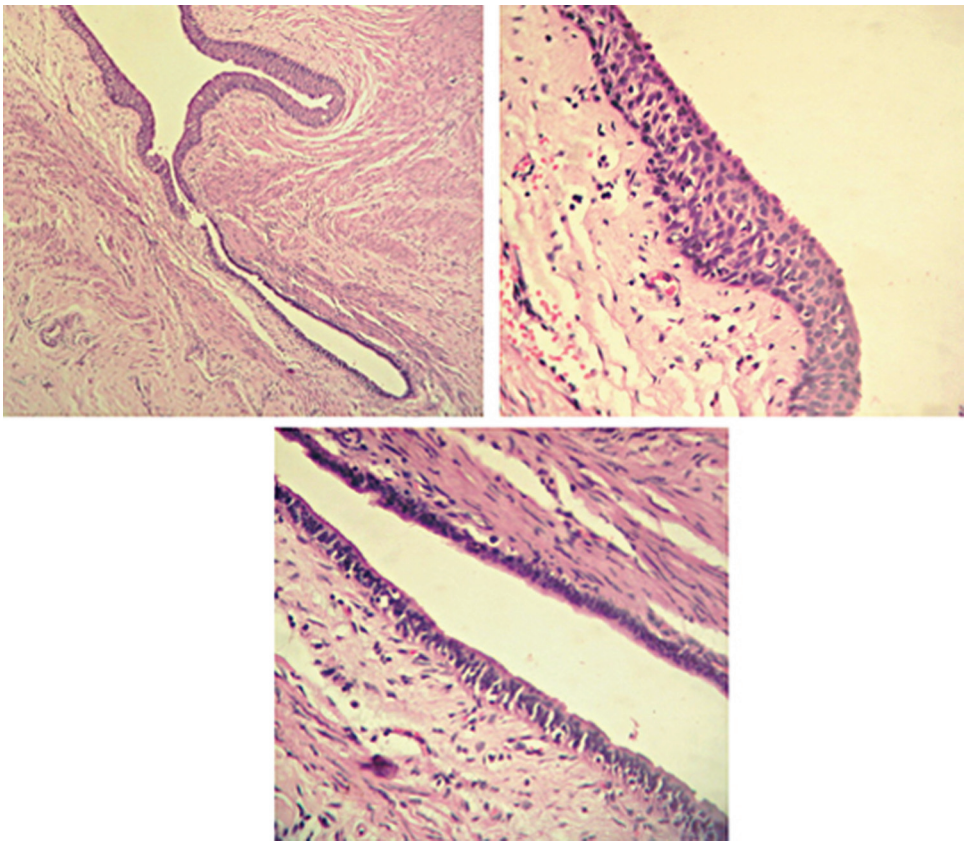

Figure 4 Histological section of the tumour.

similar experience of unexpected discovery on echocardiography, of an intracardiac tumour in a Nigerian neonate was reported in 1992,[20] which turned out to be a rhabdomyoma, the most common cardiac tumour encountered in children.[2] In Nigeria, particularly when patients are asymptomatic, the pressure to perform echocardiography is often downplayed because of lack of the equipment or the expertise to perform the procedure. Diagnosis of most common lesions is often therefore based on clinical features, supported by chest radiography and electrocardiography alone. Thus, a number of asymptomatic patients with a murmur, who might have significant pathology,

might be misdiagnosed, as would have been the case in our patient.

From a review of the previously reported adult cases of intracardiac bronchogenic cyst, it would appear that these cysts occur most commonly in the interatrial septum, (three cases),[8–10] followed by the left atrium,[6 7] (two cases), right ventricle,,[11 12] (two cases), with one case each in the right atrium,[5] and left ventricle.[13] Associated lesions reported in these cases include ASD,[10] as is the case with our patient, and persistent left superior vena cava (SVC) (two cases).[6 9]

Even when asymptomatic, surgery is the preferred option of treatment for all cases of intracardiac bronchogenic cyst, the main reason being that there is no other way of ascertaining the histology of any such tumour. Even though it is a benign tumour, it may pose dangers by reason of its site or size, and there is always a possibility, though remote, of future malignant change.

In conclusion, we wish to submit that bronchogenic cyst should be considered as a rare differential of intracardiac masses.

Learning points

- This is the first report of intracardiac bronchogenic cyst in an African child.
- There is a need for echocardiography in the evaluation of all asymptomatic children with cardiac murmurs, even in resource-poor settings. This would facilitate the identification of seemingly rare, asymptomatic cardiac lesions.
- Bronchogenic cyst should be considered as a differential diagnosis of a mass on the cardiac valve seen on echocardiography in the absence of fever.
- Intracardiac bronchogenic cyst, though rare, is a potential cause of RVOTO in childhood.

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Competing interests None.

Patient consent Obtained.

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