Electrocardiographic Case: An Interesting Case of Cardiomegaly and Pulmonary Hypertension

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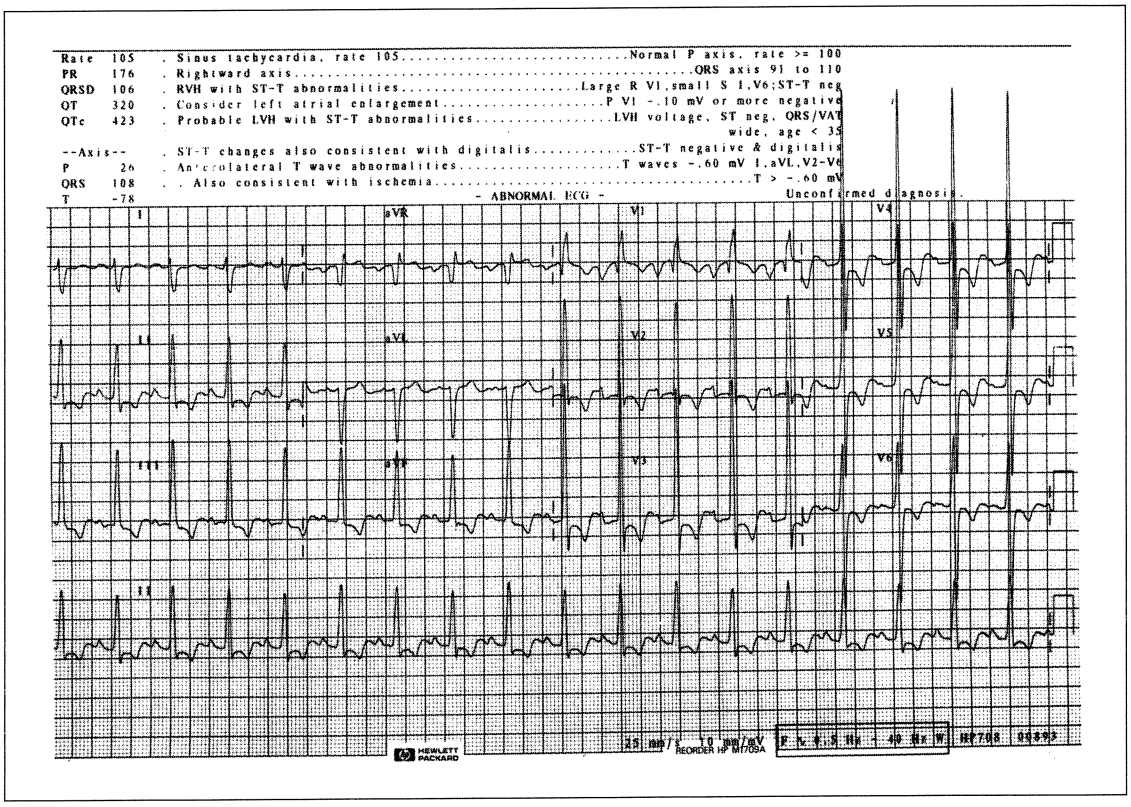

Fig 1 – 12-lead electrocardiogram

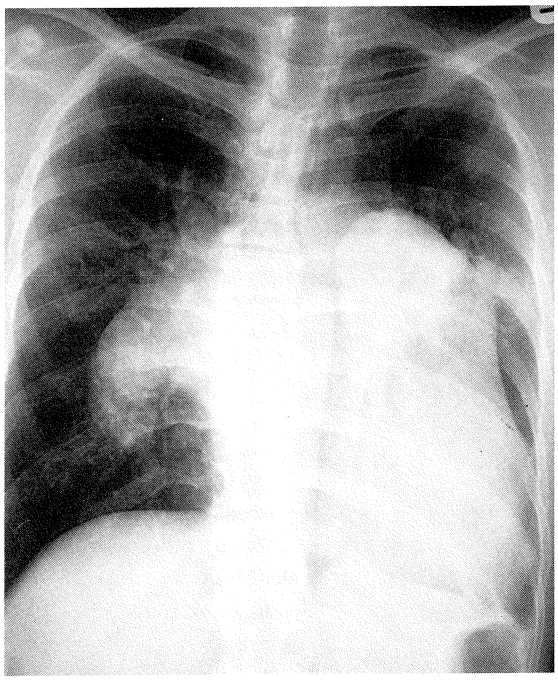

Fig 2 – Chest X-ray

CASE HISTORY

A 30-year-old Malay man presented at the Emergency Department with the complaint of progressive dyspnoea for the past 6 months. Clinical examination revealed that the patient was centrally cyanosed with digital clubbing. The jugular venous pressure was noted to be elevated. On cardiac auscultation, the second heart sound at the pulmonary area was found to be accentuated. No cardiac murmur was heard. Examination of the lungs revealed no abnormalities. The patient's chest X-ray and 12-lead electrocardiogram are shown in Figs 1 and 2 respectively. What is the diagnosis?

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DIAGNOSIS

Atrial septal defect (ASD) with Eisenmenger syndrome

DISCUSSION

Eisenmenger syndrome is a term used to describe congenital intra-cardiac systemic pulmonary communication with a reversal of the left-to-right shunt resulting from severe pulmonary hypertension. The term was first coined by Victor Eisenmenger in 1897, who described a 32-year-old man with a large ventricular septal defect (VSD)[1].

The clinical presentation of patients with Eisenmenger syndrome varies according to the level and the severity of the shunt. Patients born with a large VSD or aorticopulmonary window may present with failure to thrive or severe heart failure from the first few months to 2 years of life. However, patients with ASD tend to be asymptomatic in their early life even though the shunts are large. In such patients, progressive pulmonary hypertension will ensue and Eisenmenger syndrome will usually develop in late adulthood with consequent shortening of lifespan. Atrial arrhythmias, syncope, haemoptysis, angina-like chest pain and right-sided heart failure are late manifestations of this condition.

Common findings on physical examination of patients with Eisenmenger syndrome from ASD include: (1) central cyanosis with clubbing in severe cases; (2) elevated jugular venous pressure with a prominent 'a' wave; (3) left parasternal heave due to right ventricular hypertrophy, and (4) a loud pulmonary component of the second heart sound at the pulmonary area. An early diastolic murmur of pulmonary regurgitation, indicative of pulmonary hypertension, may also be present at the left sternal edge.

The main electrocardiographic features (Fig 1) of this patient are: (1) right axis deviation, and (2) right ventricular hypertrophy, as indicated by R/S ratio in V1 greater than one, R wave amplitude in lead V1 exceeding 7 mm and R in V1 plus S in V5 or V6 exceeding 11 mm (Sokolow's criteria)[2]. Left ventricular hypertrophy in ASD with Eisenmenger syndrome is uncommon except in those patients with underlying single ventricle or co-existing patent ductus arteriosus (PDA).

The most striking feature in the chest X-ray (Fig 2) is the presence of: (1) a giant main pulmonary artery; (2) grossly dilated right pulmonary artery, and (3) narrowing of the distal portion of the segmental pulmonary arteries resulting in the classical "pruning" radiologic pattern which is seen in long-standing pulmonary hypertension. However, the radiological distinction between decreased pulmonary vascularity and normal lung fields may be difficult at times. No pulmonary arterial calcification was seen in the chest X-ray. Marked cardiomegaly due mainly to dilatation of the right ventricle was present. The left atrium and the left ventricle are usually normal in size in Eisenmenger syndrome from ASD. In Eisenmenger syndromes arising from shunting at ventricular and great artery levels, the main and lobar pulmonary arteries are usually not so markedly dilated. Cardiomegaly is frequently seen but is usually moderate. The presence of a prominent aortic knob may suggest that the underlying shunt is at the great artery level such as in a PDA rather than an intra-cardiac shunt.

An important differential diagnosis that needs to be considered in this patient is primary pulmonary hypertension. In this condition, the electrocardiogram[3] and chest X-ray may exhibit similar features as in Eisenmenger syndrome. However in the chest X-ray, the main and lobar arteries are usually not as dilated as in Eisenmenger syndrome and the cardiomegaly is less severe. Primary pulmonary hypertension is a diagnosis of exclusion, which should be made only after other secondary causes of pulmonary hypertension such as Eisenmenger syndrome has been ruled out.

In our patient, two-dimensional echocardiography and Doppler colour flow mapping confirmed the diagnosis of a large secundum ASD with severe pulmonary arterial hypertension (calculated pulmonary artery systolic pressure of 85 mmHg) and bi-directional shunting through the ASD. The patient is being managed medically as surgical closure of the ASD is contraindicated at this late stage.

REFERENCES

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3. Kanemoto N. Electrocardiographic and hemodynamic correlations in primary pulmonary hypertension. Angiology 1988; 39:781.