A rare case of LVOT obstruction

Saji Subramanian¹, V.L Jayaprakash²

A full term female baby born to a primigravida mother was referred for evaluation of a murmur at 45 days of age. The baby was asymptomatic at presentation. Physical examination showed “ash-leaf macule” over the abdomen (Fig.1), normal pulse, JVP and no cardiomegaly. There was a grade 3/6 high pitched ejection systolic murmur in the left upper sternal border which was conducted widely.

Chest X-ray and ECG were normal. Echocardiography revealed multiple echogenic masses in the myocardium of the left ventricle (Fig.2). A mass protruded into the left ventricular out flow tract causing obstruction to blood flow with a doppler derived peak systolic gradient of 56 mm of Hg (Fig.3 & 4). A diagnosis of multiple neonatal cardiac rhabdomyomas was made.

An association with tuberous sclerosis was suspected. CT head revealed cortical tubers and ependymal nodules (Fig.5). Retinal examination showed retinal astrocytoma.

Cardiac MR was done which showed features of rhabdomyoma (Fig.6)

A diagnosis of tuberous sclerosis with Rhabdomyoma and LVOT obstruction was made.

Discussion

Neonatal cardiac tumours are rare entities. Autopsy series in children have reported an incidence of 0.027–0.08%. Rhabdomyomas are the most common primary cardiac neoplasm in children. It is estimated

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Neonatal cardiac tumours are rare entities. Autopsy series in children have reported an incidence of 0.027–0.08%. Rhabdomyomas are the most common primary cardiac neoplasm in children. It is estimated that 50–60% of tuberous sclerosis patients will develop cardiac rhabdomyomas. Conversely 51–86% of the rhabdomyomas are reported to be associated with tuberous sclerosis1.

Rhabdomyomas are generally multiple, well-circumscribed tumours that can occur anywhere in the heart, most commonly in the ventricles. They can be intramural or pedunculated and encroaching on the intracavitary space. They are hamartomas with no malignant potential.

The clinical presentation of cardiac rhabdomyomas depends on their number, size and position.

They may be detected prenatally on a routine foetal ultrasound scan or may present as hydrops foetalis. Postnatally they may be totally asymptomatic, may present with an asymptomatic cardiac murmur or may present variously as congestive cardiac failure, low cardiac output due to intracardiac flow obstruction, arrhythmias of various types or as sudden infant death.

Left ventricular out flow tract obstruction seems to be one of the rarer presentations of cardiac rhabdomyomas in infants2. But unlike our patient, most of these children are ill at diagnosis.

Manifestations of Cardiac tumours:

Benign tumours

Cardiac myxomas
- Obstructive cardiac symptoms: pulmonary oedema or progressive cardiac failure
- Embolic symptoms
- Constitutional symptoms

Papillary fibroelastoma
- Embolic symptoms of obstruction of coronary or cerebral circulation. Sudden death by prolapse into coronary ostia or occlusion of large coronary branch

Rhabdomyoma
- Dependent on size, might result in cardiomegaly, congestive heart failure, and cardiac arrhythmias.
- Sudden death or stillbirth

Fibroma
- A third are asymptomatic, the remainder usually present with heart failure, cyanosis, arrhythmias, syncope, chest pain, or sudden death

Atrioventricular nodal tumours
- Asymptomatic; sudden death

Cardiac lipomas
- Asymptomatic
- Rare extrinsic compression of heart dependent on size and location
Malignant tumours

Angiosarcoma
- Non-specific, possible chest pain, shortness of breath, malaise, and fever

Osteosarcoma
- Atrial presents with respiratory symptoms
- Ventricular presents with recurrent ventricular tachyarrhythmia

Leiomyosarcoma
- Pulmonary presents with dyspnoea, chest pain, and non-productive cough
- Cardiac might present with right heart failure, valve stenosis, rhythm alterations, conduction abnormalities, haemopericardium, and sudden death

Rhabdomyosarcoma
- Non-specific symptoms, sometimes pleuropericardial symptoms and distal embolisation
- Arrhythmias and obstructive symptoms

Cardiac lymphoma
- Cardiac tamponade, heart failure, exertional dyspnoea, atrial fibrillation, and features of right-sided heart obstruction.

The natural history of cardiac rhabdomyomas in infants and children has been well studied. All the studies have shown that these tumours have a propensity for spontaneous regression. Younger the age at diagnosis, higher the chance for spontaneous regression. Complete regression is more common in the first 4 years of life. This has important implications for therapy.

Surgical intervention is indicated only in the event of haemodynamic compromise or intractable arrhythmias. If complete surgical resection is not possible because of the location of the tumour, a partial resection can be done and the residual tumour usually regresses. The mortality from surgery for cardiac tumours in children is about 5%.

References:
2. Verhaaren H A, Vanakker O, DeWolf D, Suys B,