A Case of Rhabdomyoma in Infancy

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SUMMARY

A case of cardiac rhabdomyoma was presented. She was diagnosed as intraatrial tumor by cardiac catheterization and angiocardiography at 7 months of age.

Two months later, she was undergone open heart surgery, which revealed large rhabdomyoma in the left atrium obstructing the mitral valve.

Additional Indexing Words:

Cardiac tumor	Congestive heart failure	Mitral stenosis
Pre-exciting syndrome	Surgical treatment	

RHABDOMYOMA is the most common primary cardiac tumor in infancy,¹⁾ but the number of cases diagnosed alive was very limited. Recently, the diagnosis can be confirmed by cardiac catheterization and angiography at an early stage. Some papers have reviewed the clinical presentations of this tumors in infancy.^{2),3)} In these papers, rhabdomyomas may cause obstruction of the aortic^{2),4)} or pulmonic valve,³⁾ but obstruction of the mitral orifice by a rhabdomyoma is very rare.

The purpose of this report is to describe the clinical data, including that of cardiac catheterization and angiography, of an infant with congestive heart failure due to mitral stenosis with insufficiency caused by a rhabdomyoma of the left atrium.

CASE REPORT

A 6-month-old girl was referred to our hospital for the further investigation of heart disease. At birth, she weighed 2.87 Kg and appeared normal. She had normally developed and had been active in her new-born period.

At 1 month of age, she was noticed to have failure to thrive and weak to cry.

At 3 months, she consulted a physician and was treated with digitalis after

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a clinical diagnosis of congenital heart disease with congestive heart failure had been made. But congestive heart failure persisted and upper respiratory infections frequently developed. Then she was transferred to our hospital. There was nothing in the family history to suggest neurological or cardiovascular disease.

On admission:

Physical examination revealed mild cyanosis on lips and nail-beds. Her weight was 4.89 Kg. The heart rate was 90 per minute and respiratory rate 60 per minute. Peripheral pulses were present and equal. The liver was palpated 3 cm below the right costal margin. On auscultation of the heart, a loud grade 4 regurgitant systolic murmur and grade 2 diastolic murmur were heard at the fourth intercostal space along the left sternal border which radiated to the apex and the second sound was single. The intensity of the murmur did not change on different occasions and positions of auscultation. There was no focal neurological signs and the skin was normal.

The chest X-ray film showed the evidence of cardiac enlargement with a cardio-thoracic ratio of 65% and there was a distinct protrusion on the left cardiac border (Fig. 1). Pulmonary vascularity was increased. The electrocardiogram showed right axis deviation ($+55^\circ$) in the frontal plane and right ventricular hypertrophy (Fig. 2). A short P-R interval with delta wave suggested pre-exciting syndrome. Ultrasoundcardiographic examination presented the evidence of left atrial tumor, there being multiple echoes during diastole behind the anterior mitral leaflet (Fig. 3).

Cardiac catheterization was performed (Table I). No shunt was detected, but the right ventricular and pulmonary artery pressures were elevated. Pulmonary wedge pressure curve was not so well traced, but c and v waves were high. No

Fig. 1. Chest X-ray film shows cardiomegaly with a hump at the left cardiac border.

Fig. 2. Electrocardiogram shows short P-R interval. The change of S-T segment suggested digitalis effect.

Fig. 3. Echocardiogram. Behind the anterior mitral leaflet, the tumor produces multiple band-like echoes during diastole.

obstructions of the left and right ventricular outflow tract were detected. Pulmonary artery injection of contrast medium with follow-through to the left atrium was performed and left atrial filling defect was noted. Left ventricular angiogram demonstrated moderate mitral regurgitation and again left atrial filling defect (Fig. 4). So left atrial tumor was believed to be present.

At 9-month of age, cardiac surgery was performed because of severe congestive heart failure. A muscle-like tumor was present in the atrial septum. It was protruding into the left atrium and had given rise to mitral stenosis and insufficiency. Total removal of the tumor seemed to be difficult, so it was dissected into

Table I. Cardiac Catheterization Report

Locations	Pressure (Mean) mmHg	O ₂ Saturation (%)
V. C.		66.9
R. A.	7/-4 (3)	64.4
R. V.	76/10 (30)	65.5
P. A.	80/28 (48)	68.7
P.A.W.	24/13 23/15 (17)	
F. A.	105/50 (68)	93.5
L. V.	118/15 (58)	
Ao.	118/55 (78)	

Cardiac Output 4.4 L/min/M²
P.A.R. 23.48 mmHg/L/min
T.P.R. 36.36 mmHg/L/min
T.S.R. 49.24 mmHg/L/min

Fig. 4. Selective left ventricular angiogram shows regurgitation of mitral valve and filling defect in the left atrium.

Fig. 5. Microscopic section of the tumor shows rhabdomyoma cells, spider cells and cross striations.

several pieces. Postoperatively, cardiac beats were weak and small. Recovery of beats was unsuccessful and the patient passed away.

Microscopically, the cardiac tumor showed large clear rhabdomyoma cells (Fig. 5).

As the postmortem examination could not be carried out, anomalies of the brain or kidneys were not detected.

DISCUSSION

Though in many reports were discussed the cardiac rhabdomyoma,²⁾⁻⁹⁾ the true nature remains obscure, while it is usually included in the tumors of the heart. It may form a single mass or multiple nodules and occur mainly in the walls of the ventricles or the ventricular septum. When large, it may project into the cavity of the ventricle. Histological sections present a spongy vacuolated appearance when the tissue has been fixed in aqueous formalin. However, if the tissue is fixed in alcohol, these vacuoles are found to contain a mucopolysaccharide that stains positively by the periodic acid-Schiff reaction, so it is probably glycogen.⁵⁾

According to Kidder,⁹⁾ the youngest subject reported on was a 6-month old fetus and the oldest patient was 45 years old. Forty % of the patients were dead until 6 months of age, 60% by the end of 1 year, and 80% by age of 5 years.

Information on the clinical presentation of cardiac rhabdomyomas in infants are scanty.¹⁰⁾ None the less, these reported cases with sufficient clinical data suggest that a rhabdomyoma of the heart can present itself in various way: obstruction of blood flow, myocardial involvement, and disturbance

of the cardiac rhythm. Obstruction of the left ventricular outflow tract by a rhabdomyoma may simulate subaortic stenosis^{2),4),10),11)} and that of the right ventricle resembles pulmonary stenosis.^{3),10),13)} Among reported cases, the patient of Golding and Reed¹²⁾ was diagnosed as tetralogy of Fallot by the cardiac catheterization data and cine-angiographic findings. He underwent successful open heart surgery and microscopic examination of muscle removed from the infundibulum demonstrated rhabdomyoma. But, to our knowledge, mitral valve obstruction produced by a rhabdomyoma has been reported only once (Harich⁴⁾). In that case, a huge tumor projected both into the left atrium and ventricle from the atrial and ventricular septum.

Congestive heart failure due to myocardial involvement is a common complication of cardiac rhabdomyoma and the symptom usually develops in very early stage.^{4),11),14)} In our case, however, the symptom of congestive heart failure has developed relatively late, so we supposed that the failure was not only due to myocardial involvement of rhabdomyoma but also due to mitral failure.

Arrhythmias are commonly observed in intramural tumors. Atrial arrhythmia such as atrial premature contraction, tachycardia, flutter or fibrillation,²⁾ and ventricular tachycardia¹⁰⁾ have been reported. A short P-R interval with bouts of reciprocal tachycardia was noted in the report of Van der Hauwaert¹⁵⁾ and he suggested that the mechanism of it was probably the impulse re-entry by way of accessory atrioventricular pathways in the tumor.

As an autopsy was not permitted, other anomalies could not be detected in our case. According to Shafer,²⁾ if the brain is examined at autopsy, tuberous sclerosis will probably be found in almost all cases.

No surgical treatment has been available till quite recently, but nowadays it is generally said that surgical removal of the tumor should be considered if it is producing significant obstruction of the blood flow.²⁾

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