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Multiple Rhabdomyoma of the Heart Presenting with a Congenital Supraventricular Tachycardia

— Report of Case with Ultrastructural Study —

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A case of congenital rhabdomyoma of the heart in a 5-month-old Korean infant is described. The patient presented with a congenital supraventricular tachyarrhymia that was detected in utero by fetal sonography. The tumor was multiple, but no obvious association with tuberous sclerosis complex was demonstrated. Microscopic examination revealed classic "spider cells" with rich glycogen content. Ultrastructurally, the cells contained numerous leptofibrils, clumped Z band material, and desmosome-like cell junctions. The case is a second documented case of cardiac rhabdomyoma in this country, and its presentation as an etiological factor of supraventricular tachycardia is a very unusual manifestation.

Key Words: *Heart, Rhabdomyoma, Supraventricular tachycardia*

INTRODUCTION

Among primary cardiac tumors, rhabdomyoma affects mainly infants and children, and a fairly large proportion of cases are congenital (Bigelow et al., 1954). The ventricles are the main sites usually affected with the atrial septum affected less frequently (Heath, 1968).

The associated symptoms with cardiac rhabdomyomas are chiefly related to obstruction of intracardiac blood flow; arrhythmias are only rarely associated (Fenoglio et al., 1976). A significant proportion of infants with cardiac rhabdomyomas are stillborn; these tumors usually occur in association with tuberous sclerosis. Only sporadic cases have been reported in literature and there is yet no documented case of cardiac rhabdomyoma with congenital arrhythmia reported in this country. The authors described a case of cardiac rhabdomyoma in a 5-month-old Korean girl. The lesion was multiple and one was closely located near the sinuatrial node causing supraventricular tachycardia.

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CASE REPORT

A 5-month-old Korean female was brought to the Pediatric Department of Seoul National University Children's Hospital (SNUCH) due to congenital tachyarrhythmia. During the in utero period, rapid fetal heart rate was detected by fetal sonography. The infant was delivered as normal full-term by Caesarean section at a general hospital and the birth weight was 3.25Kg (50-75 percentile). She was discharged despite persistent tachyarrhythmic attacks. Since then, there have been frequent bouts of tachyarrhythmia. She also showed poor appetite and weight gain. She was brought to another general hospital where echocardiography showed a left ventricular mass. She was managed with digoxin and diuretics, but the symptoms persisted, and she was transferred to SNUCH for further treatment. Upon admission, she was observed to be undernourished and slightly pale. Her heart rate was 230/min and body weight and height

were 5.2kg (3rd percentile) and 60cm (3rd percentile), respectively. Her breathing sounds were clear and the heartbeat was regular without murmur. The abdomen was slightly distended, and the liver was palpable by 2 fingerwidths below right costal margin. A whitish, slightly depigmented patch was noted on her back, but neurological abnormality was not present. Laboratory findings were unremarkable. A chest X-ray revealed mild cardiomegaly with a bulging of the left cardiac border. Electrocardiogram and Holter monitoring demonstrated ectopic, automatic supraventricular tachycardia, non-conducted premature atrial contractions with intermittent atrial bigeminy, and atrial escape beat. Echocardiography revealed increased echogenicity in both ventricles, left atrium, and sinuatrial node. MRI scan revealed intracardiac tumors involving atrial septum, interventricular septum, and posterior wall of the left ventricle. Brain CT was unremarkable. A thoracotomy was conducted, and near-total removal of left atrioventricular (2.5×2.5 cm), right ventricular (1×1 cm), and right atrium-superior vena caval junction (sinuatrial node region, 0.3×0.3cm) tumors was done. Postoperatively, the heart rate was normalized and her hospital course was uneventful. Three months following the operation, left ventricular aneurysm developed at the previous tumor site and pseudoaneurysmectomy was done.

PATHOLOGIC FINDINGS

Gross Findings:

The received specimen consisted of a small lump of solid soft tissue and fragments of dusky pinkish soft tissue. The main piece measured 1.7×1.6×0.5cm and weighed 0.8gm. The cut surface was homogeneously solid and pinkish red (Fig. 1). No secondary change such as necrosis or hemorrhage was observed.

Light Microscoic Findings:

The individual tumors were rather well circumscribed from surrounding myocardial fibers, and normal myofibers were occasionally observed in the periphery of the tumors (Fig. 2). The tumor cells had mainly vacuolated clear or weakly eosinophilic granular cytoplasm. The clear cytoplasm was traversed by radiating, thin cytoplasmic processes, comprising typical features of so called "spider cells" (Fig. 3). The glycogen content in clear cytoplasm was demonstrated on periodic acid-Schiff and diastase treated periodic acid-Schiff stainings on alcohol-fixed tissue. The cytoplasmic cross striations were easily identified on both hematoxylin-eosin and phosphotungstic acid-

Fig. 1. The rhabdomyomas have homogeneously solid, pinkish red cut surface. More yellowish uninvolved myocardium partially encircles individual tumors.

Fig. 2. The tumor cells undergo smooth transition with normal myocardium. The characteristic clear cytoplasm of rhabdomyoma cells are easily distinguished from surrounding myocardium.

hematoxylin staining. The nuclei were mainly oval and were mainly located in the center. Supporting stroma was scanty.

Ultrastructural Findings:

Most of the tumor cells had scattered glycogen particles in their clear cytoplasmic space upon light microscopic examination. Individual cells contained numerous, haphazard leptofibrils, clumped Z-band material, scattered small mitochondria, and endoplasmic reticulum (Fig. 4). The condensed cytoplasm was mainly composed of leptofibril complexes, and my-

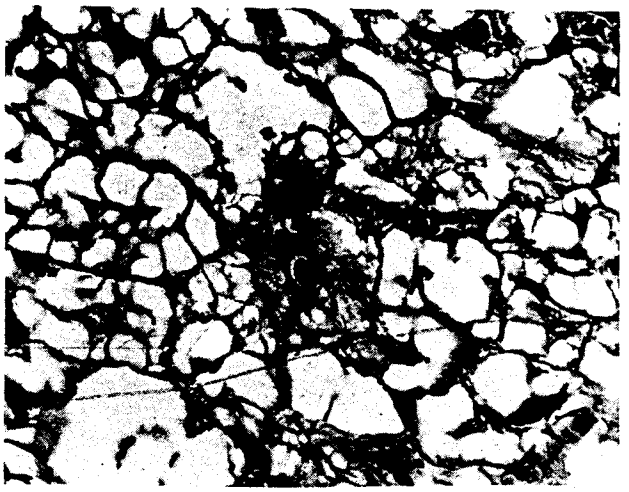

Fig. 3. The classic 'Spider cells' have clear cytoplasm divided by cytoplasmic processes. The cytoplasmic cross striations are frequently observed.

ofilaments were not infrequently observed. Membrane-bound vacuolar structures were not observed. The nuclei had a peripherally condensed chromatin pattern with relatively prominent nucleoli. Individual cells were in close contact and were connected by desmosome-like cell junctions which were composed of dense plaques (Fig. 5).

DISCUSSION

Cardiac rhabdomyoma is a rare occurrence with only sporadic cases reported in world literature (Fenoglio et al., 1976). In this country, only a case of cardiac rhabdomyoma in a tuberous sclerosis patient is present as yet (Yoon et al., 1989), and in the present study, the authors described a case of cardiac rhabdomyoma presented with congenital tachyarrhythmia of which classic morphologic features were verified.

Congenital rhabdomyoma is known to be associat-

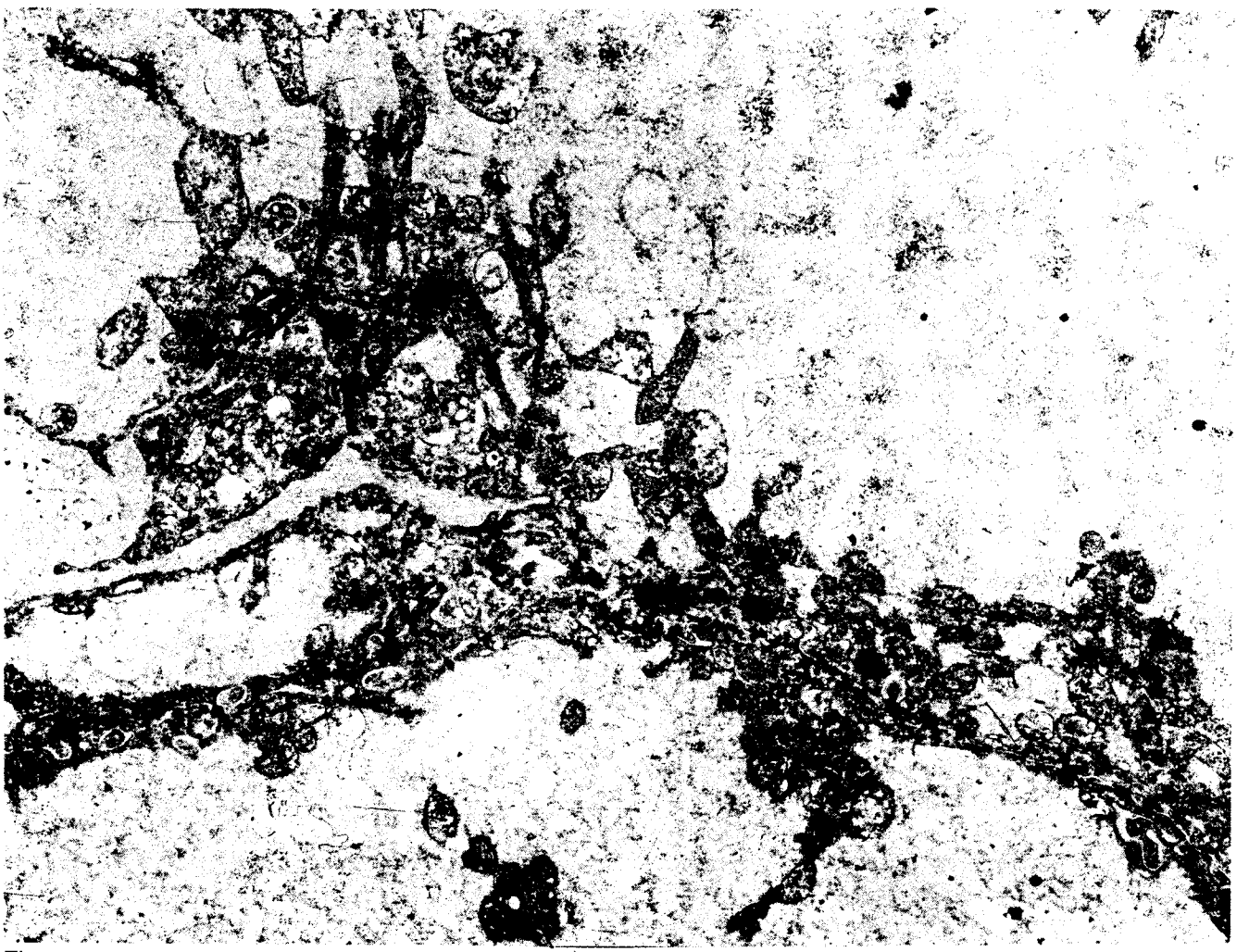

Fig. 4. Multiple intracytoplasmic leptofibrils, Z band-like materials, and a few dilated mitochondria are present in the cytoplasm (TEM, ×12,500).

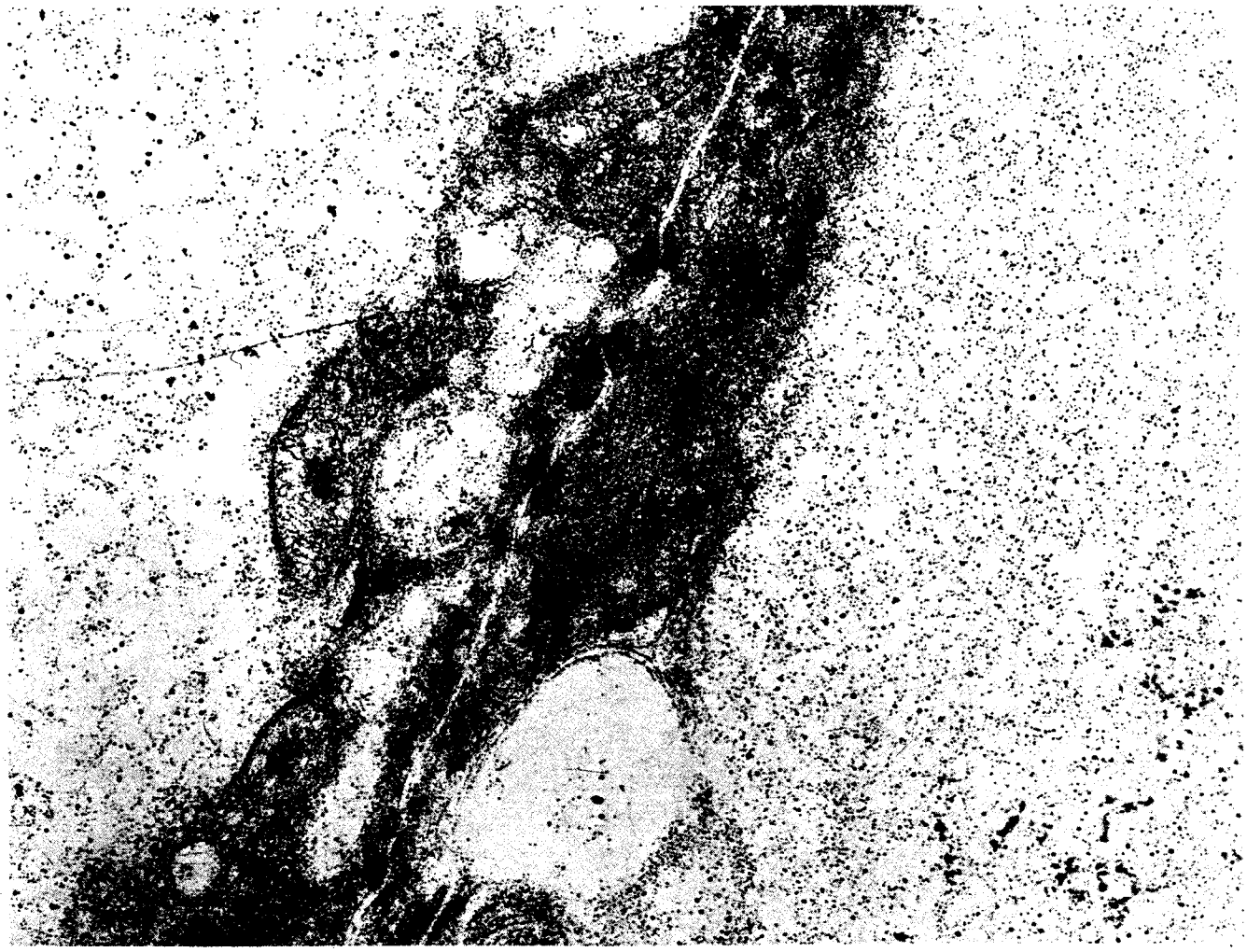

Fig. 5. The cells are connected by desmosome-like intercellular junctions composed of dense plaques (TEM, ×50,000).

ed with other congenital anomalies, and there is a high correlation with tuberous sclerosis (Bigelow et al., 1954). Though a depigmented lesion was found in the present case, the lesion differed from the typical salmon's patch of tuberous sclerosis and normal brain CT also ruled out any connection to tuberous sclerosis in the present case. Other evidences of organ involvement of tuberous sclerosis were not found during the whole course of hospital work up. The clinical features of cardiac rhabdomyoma are usually related to obstruction of intracardiac blood flow and arrhythmia may develop in a small proportion of cases (Fenoglio et al., 1976). Both atrial and ventricular involvement and the occurrence of paroxysmal supraventricular tachycardia due to sinuatrial nodal involvement, which was dramatically corrected after surgical removal, are regarded as unusual features of the present case.

The congenital and multiple nature of this neoplasm was regarded as a strong evidence for being a malformative process rather than a true neoplasm and Fenoglio et al thought that the rhabdomyoma is a kind of 'tumor-like malformation (fetal hamartoma)' on the basis of their observation that rhabdomyoma cells closely resemble embryonic cardiac muscle cells (Fenoglio et al., 1977). The ultrastructural findings of the present case coincided well with previous descriptions (Fenoglio et al., 1976, Fenoglio et al., 1977, Bruni et al., 1980), but type A and B cells of Bruni et al, were not definable.

Despite well known pathologic features, the reason for multiple involvement and site predilection has been unsolved in this peculiar entity.

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