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Disseminated Tuberculosis and Cardiac Rhabdomyomata

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Primary cardiac tumors are rare in children. Rhabdomyomas constitute more than 60% of cardiac tumors and generally present within the first year of life(1,2). We report a case of multiple rhabdomyoma of the heart in a 10-month-old patient with disseminated tuberculosis.

Case Report

The patient was a full term male boy, born to a 22-year-old primigravida. The mother was an open case of pulmonary tuberculosis receiving anti-tubercular treatment. The child had completed his primary immunization including BCG which was given in the neonatal period. The child achieved normal milestones but had frequent upper respiratory tract infections, gastroenteritis and failure to thrive. At 3 months of age he was admitted to the hospital with 4 days history of high fever, breathlessness and productive cough. On examination, he was severely malnourished (marasmus) and moderately dehydrated. The abdomen was distended with free fluid. There was an ejection systolic murmur grade 2/6 in the mitral and aortic areas. The roentgenogram of the chest showed miliary mottling. A 2-dimensional echocardiogram showed congenital bicuspid aortic valve with stenosis. The child died 3 days after admission.

At autopsy, the pleural and pericardial surfaces and the lung parenchyma were studded with miliary tubercles. The hilar lymph nodes were enlarged and caseous. The liver, spleen, kidneys and adrenals also showed multiple tubercles. The loops of the intestines were matted with numerous enlarged caseous lymph nodes in the mesentry.

The heart was enlarged and weighed 90 g. There were grey-white nodular masses on the epicardial surface. The right atrium, right ventricle and the pulmonary valves had pin head sized greyish nodules on the endocardium. The left ventricular cavity was distorted by large nodular masses protruding into the outflow and sinus portions. The interventricular septum was thickened and bulging onto the right and left side (*Fig. 1.*).

Multiple sections taken from various organs were processed and stained with hematoxylin and eosin. Sections from the

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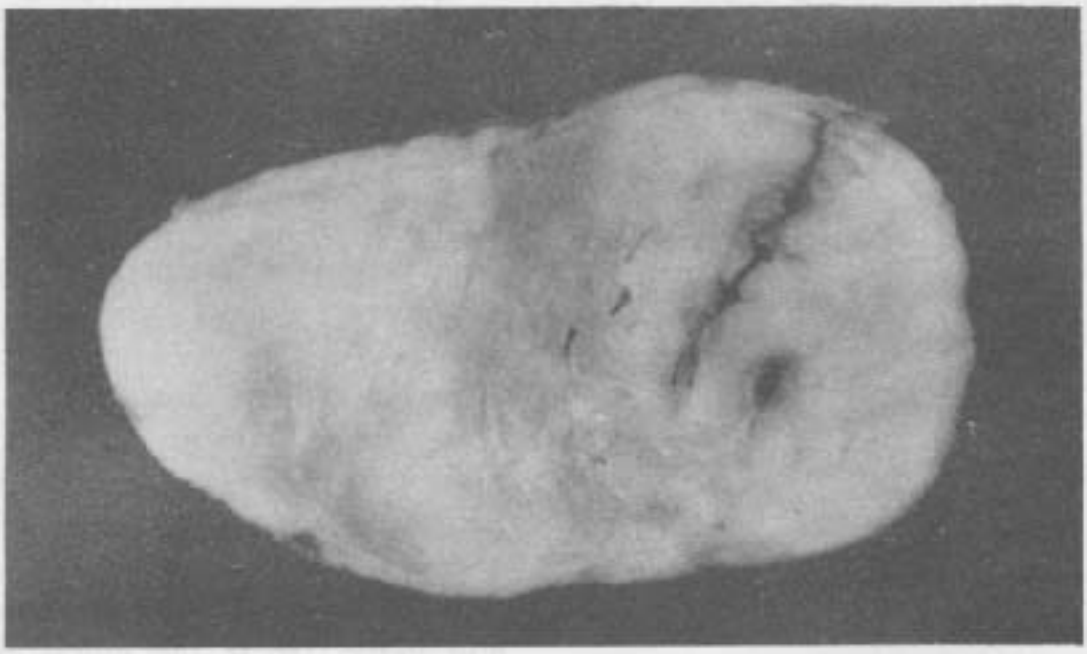

Fig. 1. Tranverse section of the heart showing slit like left ventricular cavity; right and left ventricle and inter-ventricular septum show grey-white tumor masses producing marked thickening of the wall.

lungs, pleura, liver, spleen, lymph nodes, intestines, kidneys and adrenals showed foci of caseous necrosis and granulomatous inflammatory reaction. Zeihl-Neelsen staining showed the presence of acid fast bacilli.

The nodular masses in the heart were composed of large cells with clear cytoplasm and spider-like cytoplasmic processes. These nodules were well demarcated from the surrounding normal myocardium. Such nodules were also seen abutting on the A-V node but were not seen in the S-A node. The histological appearance of the cardiac nodules was consistent with a rhabdomyoma (*Fig. 2*). There were no tubercular granulomas in the heart. A diagnosis of disseminated tuberculosis and cardiac rhabdomyomata was made.

Discussion

Cardiac tumors are rare in children but rhabdomyomas constitute the largest category(2). Most cases present below one year of age with valvular or outflow obstruction, congestive cardiac failure and arrhythmias(1,2). The condition may occur in association with tuberous sclerosis, kidney tumors and adenoma sebaceum of the skin(2). Fenoglio and his associates in an analysis of 36 cases of rhabdomyomas found left ventricular involvement and multiple lesions in over 90% cases(3). In a clinico-pathological study of 16 cases of cardiac rhabdomyomas by Burke and Virmani, ranging from birth to 9 years, an association with tuberous sclerosis was described in 10 patients and with congenital heart disease in 4(4). Surgical therapy for intramural nodular tumors is usually ineffective(2). However, there are occasional reports of successful excision of the tumor(1,4,5). Diffuse rhabdomyomatosis has been reported but most of these patients are still born, though rarely survival upto 21 years has been observed(6,7).

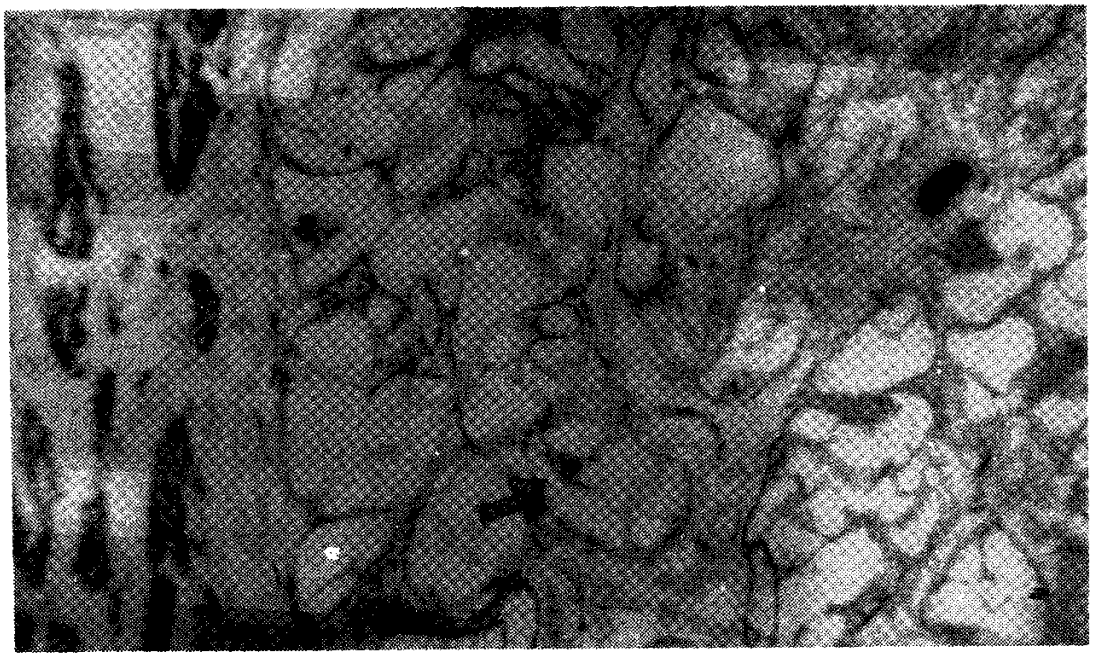

Fig. 2. Photomicrograph showing normal myocardium and well circumscribed tumor composed of large round to oval vacuolated cells with spider like cytoplasmic processes (arrow showing spider cell) (hematoxylin and eosin × 160).

The present case had clinical evidence of outflow obstruction which has been described in 50% cases of rhabdomyoma(1). There were multiple nodules in the chambers of the heart including the left ventricle. There was no evidence of tuberous sclerosis, kidney tumor or adenoma sebaceum. There was no associated congenital heart disease.

Congenital tuberculosis is rare even in developing countries(8,9). Less than 300 cases of congenital tuberculosis have been reported in medical literature(9) and only about 3 cases from India. Criteria for diagnosing congenital tuberculosis have been established by Biezk in 1935(10). The patients present in early neonatal period, either with obstructive jaundice due to the enlarged hepatic lymphnodes or a respiratory disease simulating bacterial pneumonia.

In the present case, the clinical history and postmortem findings support the diagnosis of congenital tuberculosis. However, due to the late clinical presentation of the child and the non availability of the placenta for examination, the intrauterine nature of tuberculosis cannot be confirmed. Therefore, here two possibilities have to be considered—congenital tuberculosis with an hepatic primary complex with widespread dissemination or a postnatally acquired disseminated tuberculosis.

In the present case, two unusual conditions—congenital disseminated tuberculosis and multiple cardiac rhabdomyomata were present. The association of these two relatively rare conditions in the patient is incidental but nevertheless uncommon.

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Fatal Rabies Encephalomyelitis Despite Chick Embryo Vaccine Prophylaxis

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Rabies encephalitis was one of the earliest known diseases, and because it is uniformly fatal, prevention of virus invasion is of great importance. With the introduction of the first rabies vaccine in 1885, post exposure prophylaxis assumed a new dimension. The initial vaccines were all purified neural vaccines, but a high rate of adverse reactions led to the introduction of the duck and chick embryo vaccines and finally the human diploid cell vaccine in 1974. Concomitant administration of rabies immune globulin is necessary for complete protection(1,2), and upto 1985, no case of fatal rabies encephalitis following this combination, prophylaxis has been reported.

Case Report

A 12-year-old boy was brought with complaints of high grade fever with bodyache for 8 days. There was progressive difficulty in walking, and episodes of diplopia and delirium. He had been bitten on the right forearm, and scratched on the right buttock and left leg by a dog 25 days ago. The same dog had bitten several others but its fate was unknown. The wound had been cleaned with hydrogen peroxide and povidone iodine, and treated with silver sulfadiazine cream. He had also received chick embryo rabies vaccine on days 0, 3, 7, and 14. There was no history of seizures, focal deficits, aerophobia or hydrophobia.

Examination revealed stable vital parameters. All the wounds had healed with

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