A newborn infant with intrapericardial rhabdomyosarcoma: a case report

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Cardiac tumors are uncommon in neonates and most of them are histologically benign. The most common cardiac tumor in neonates and infants is rhabdomyoma. Malignant cardiac tumors are considerably rarer, and rhabdomyosarcoma (RMS) is the leading malignancy.

To our knowledge, only one case of intrapericardial RMS was reported in the literature, in a seven-month-old baby. Here we present another newborn baby with intrapericardial RMS.

Key words: rhabdomyosarcoma, pericardium, newborn.

Primary cardiac tumors are uncommon in the fetus and neonate, but with improved imaging techniques such as two-dimensional echocardiography, ultrasonography and magnetic resonance imaging, increasing numbers have been reported in the last decades. Primary tumors are found considerably more frequently than metastatic ones, and most of them are benign. The most common type of heart tumors in children is rhabdomyoma followed by teratoma and fibroma[1,2]. Holley et al.[3] revealed an incidence of primary cardiac tumor as 0.14% in 14,000 fetal echocardiographies over an eight-year period. Of these tumors, 89% were rhabdomyoma.

Malignant cardiac tumors are considerably rarer in neonates and infants. Only a few cases of malignant tumors have been reported[4-6], with the predominant tumor being rhabdomyosarcoma (RMS). Here we present a newborn baby with intrapericardial RMS. To our knowledge, it is the second RMS case in childhood detected in the pericardium.

Case Report

A 21-day-old newborn infant was admitted to the hospital with respiratory distress and cyanosis. On admission, her weight was 3200 g (25-50 p), length 54 cm (75-90 p), and head circumference 36 cm (75-90 p). Vital signs were as follows: temperature: 37°C, heart rate: 170/min, respiratory rate: 68/min and blood pressure: 93/58 mmHg. She was tachypneic and tachycardic. Breath and heart sounds were diminished on the left side of the thorax. The remainder of the physical examination was unremarkable.

Laboratory investigation showed a normal blood count and biochemical values. Capillary blood gases were within the normal limits. C-reactive protein was 9.4 mg/L. A chest radiograph suggested pericardial effusion. Echocardiography showed thickening of the pericardium and profuse effusion without disturbing diastolic enlargement of the heart. Pericardiocentesis was carried out under ultrasound imaging and 120 ml serohemorrhagic effusion was drained. There were abundant erythrocytes in microscopic examination. There was no bacterium or leukocyte on smear, on Gram staining and with Ehrlich-Ziehl- Neelsen, and culture was negative. The fluid protein was: 3.94 g/dl, lactate dehydrogenase (LDH): 443 U/L, and glucose: 46 mg/dl.

On the ninth day of admission, peri-cardio-pulmonary fenestration and drainage had to be performed due to increased effusion. Ceftazidime

and vancomycin were prescribed prophylactically. She had anemia and was transfused with packed red cells. Pericardial biopsy showed fibrous pericarditis. After drainage, thorax tomography revealed no abnormality except for minimal pleural effusion. On the 20th day of admission, echocardiography showed a thickened, fibrotic pericardium and loculated pericardial effusion, and anterolateral partial pericardiectomy was performed. Pericardium was gray-yellow, thick and adhered tightly to the adjacent tissue. Pathologic examination demonstrated that the tumor was extended in all pericardial tissue, had a myxoid stroma, and included abundant racquet cells, and hypercellular and necrotic areas (Fig. 1). Defined cells were positive with periodic acid-Schiff (PAS), vimentin, keratin, S-100, and epithelial membrane antigen (EMA). Desmin was also positive (Fig. 2). Embryonal RMS was diagnosed. On the 23rd day of admission, echocardiography showed a hyperechoic mass in the pericardium surrounding the posterior wall of the left ventricle and apex, and a large mass in front of the pulmonary artery in modified aortic short axis (Fig. 3a, 3b). Thorax tomography revealed a mass surrounding the heart and mediastinal vascular process that could not be differentiated from the pericardium and other vascular processes, presenting small calcification areas (Fig. 4). Abdominal, pelvic and cranial tomographies were normal. She was treated with vincristine, actinomycin-D, and cyclophosphamide chemotherapy protocol. On the subsequent days she had neutropenia and respiratory insufficiency and she was supported on ventilator. In spite of intensive supportive therapy, she died on the 34th day of admission.

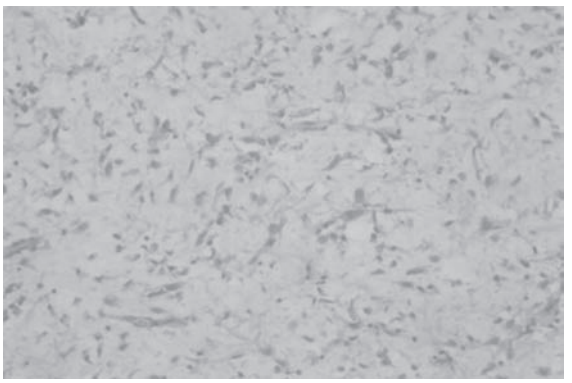

Fig. 1. Tumor with a myxoid stroma, including abundant racquet cells, and hypercellular and necrotic areas.

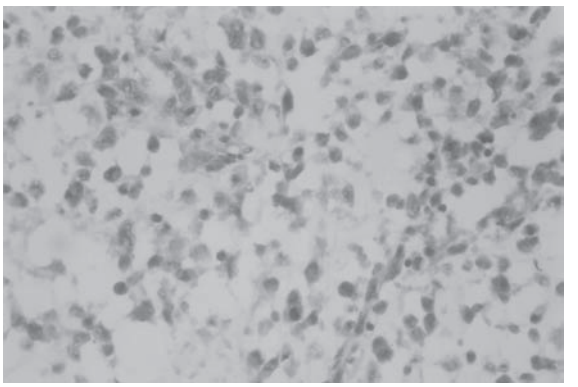

Fig. 2. Racquet cells positive with desmin.

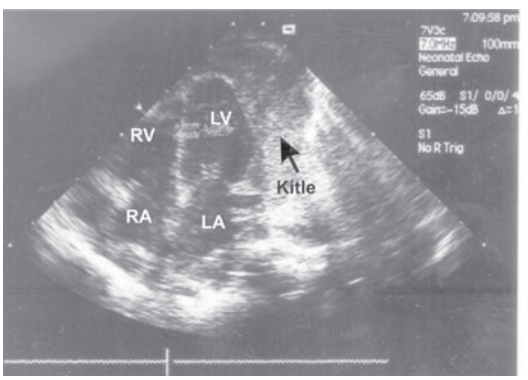

Fig. 3a. In four-chamber position, echocardiography shows hyperechoic mass in pericardium surrounding posterior wall of left ventricle and apex.
RV: Right ventricle. RA: Right atrium. LV: Left ventricle. LA: Left atrium. Arrow shows the mass.

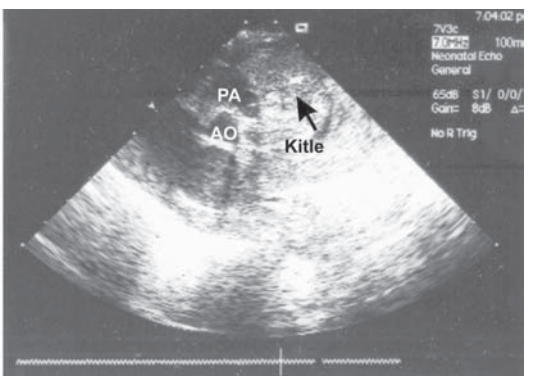

Fig. 3b. In modified aortic short axis, echocardiography shows a large mass in front of pulmonary artery.
PA: Pulmonary artery. AO: Aorta. Arrow shows the mass.

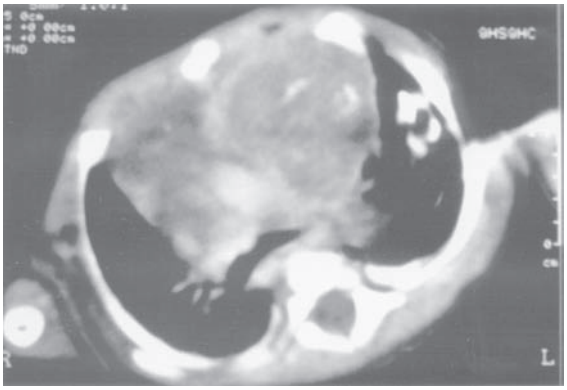

Fig. 4. Thorax tomography showing a mass surrounding the heart and mediastinal vascular process that could not be differentiated from pericardium and other vascular processes, presenting small calcification areas.

Discussion

Primary cardiac RMS is very rare in infants and children. It usually presents clinically with cyanosis, murmur, congestive heart failure or arrhythmia, or obstruction if it extends into the cardiac chamber[4]. The present patient was admitted to hospital with respiratory distress and cyanosis.

Traditionally, RMS was classified as embryonal, botryoid (a subtype of embryonal), alveolar, and pleomorphic types[7]. Embryonal type accounts for about 53% of all cases, occurring in infants, children, and young adults and generally having an intermediate prognosis[8]. Embryonal type is characterized histologically by small spindle- or tadpole (racquet)-shaped rhabdomyoblasts with many mitoses. Vimentin and muscle markers desmin and actin are immunoreactive. Other characteristic findings are myxoid areas and necrotic foci. In the present patient, the tumor had myxoid stroma including abundant racquet cells, and hypercellular and necrotic areas. Tumor cells were positive with PAS, vimentin and desmin.

Cardiac tumors are best diagnosed by two-dimensional echocardiography. In Beghetti et al.'s[1] article, 55 of 56 patients with primary cardiac tumors were reported to be diagnosed by echocardiography. Even meticulous prenatal ultrasonography can reveal cardiac tumors[3]. In Elderkin and Radford's[9] article evaluating primary cardiac tumors over a 20-year period, they found 12 patients, five of whom were diagnosed by antenatal ultrasonography.

Cardiac RMS is very rare in neonates, and to our knowledge, only one case with intrapericardiac RMS was reported in the literature, by Beitzke et al.[10], in a seven-month-old infant. That case was first reported as myxosarcoma histologically at three months of age and in spite of chemotherapy and radiation, the infant died due to multiple intracerebral metastases. It was later diagnosed as RMS in postmortem biopsy. Generally, neonates with primary cardiac malignant tumors have grave prognosis with essentially no survivors[4,11]. Our case died due to septic shock during chemotherapy regimen.

In conclusion, although cardiac tumors are rare in newborns, a tumor must be kept in mind in patients with pericardial thickening and pericardial effusion, and the patient should be followed up by serial echocardiographic studies.

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