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Case Reports

Unique Association of a Rapidly Growing Right Atrial Myxoma in a Child with Double-Outlet Right Ventricle

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Abstract. We describe a patient with double-outlet right ventricle in whom a large right atrial myxoma developed over approximately 6 months. This patient represents the first case described of a right atrial myxoma occurring in an unoperated patient with congenital heart disease other than an isolated atrial septal defect. Because the child was followed with serial echocardiograms, we can document the rapid growth of the tumor.

Key words: Atrial myxoma — Congenital heart disease — Double-outlet right ventricle

The occurrence of right atrial myxoma during childhood is uncommon. Atrial myxomas are considered to be slow-growing tumors, although tumor growth rates have not been established. Upon review of the literature, there have been no previous reports of myxomas in children with underlying congenital heart disease other than isolated atrial septal defects. The following case describes rapid growth of a large atrial myxoma in a patient with double-outlet right ventricle (DORV) and pulmonic stenosis.

Case Report

The patient was diagnosed with DORV, dextroposition, and a large omphalocele in utero by fetal ultrasonography. During the immediate neonatal period an echocardiogram further delineated her anatomy as DORV (S,D,D), atrial septal defect, large malalignment-type ventricular septal defect, and pulmonic stenosis in the setting of dextroposition. Shortly after birth, she underwent primary repair of the omphalocele.

During early infancy she developed mild congestive heart failure, which was adequately managed with digoxin, furosemide, and spironolactone. She was followed expectantly, with plans for surgical repair of her structural heart disease at a later date. Her follow-up cardiac evaluations included serial echocardiograms.

At diagnosis she presented with a several-week history of lethargy and daily fever spikes, with no obvious source of infection. Blood cultures were negative. Echocardiography revealed a large homogeneous mass in the right atrium that appeared to partially obstruct tricuspid inflow (Fig. 1A). The mass measured 4.7 × 2.3 cm and was attached to the inferior portion of the atrial septum. A review of an echocardiogram obtained approximately 6 months prior to this study revealed no cardiac masses (Fig. 1B).

She was brought to the operating room for excision of the right atrial mass and repair of the underlying congenital heart disease. The stalk of the large right atrial tumor was resected from the atrial septum. Grossly, it was described as a moderately firm, lobulated mass of tissue measuring 5 × 4 × 2 cm in its largest diameter (Fig. 2). The cut surface was smooth, light pink, and of myxomatoid appearance. Microscopic sections were consistent with a cardiac myxoma formed by spindle-shaped cells in bands and a whorl-like pattern arising from a thick endocardium with areas of inflammatory infiltrate (Fig. 3). The patient defervesced postoperatively and was discharged home after an uneventful recuperation. Cardiac follow-up has failed to reveal any recurrence of the intracardiac myxoma.

Discussion

Intraatrial masses including tumors, vegetations of endocarditis, and thrombi are rare in children [6, 11]. Myxomas are the most common type of primary cardiac tumor in the adult population, and rhabdomyomas are the most common cardiac tumor in children [7, 11]. The formation of a myxoma within the right atrium of a child with congenital heart disease has not been previously reported. All reported cases of atrial myxomas have been in patients with structurally normal hearts or with isolated atrial septal defects [3, 5, 16].

Myxomas are most frequently located in the left atrium arising from the fossa ovalis [7]. Approximately 25% are located in the right atrium [8]. Most cardiac myxomas have benign histology, although they may

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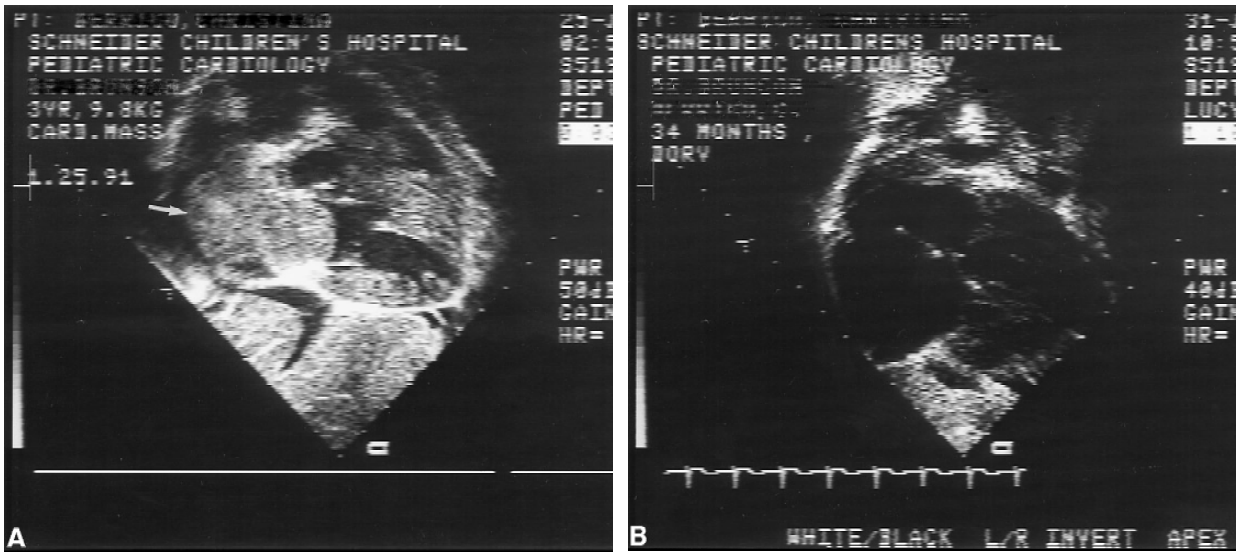

Fig. 1. (A) Echocardiogram showing a large right atrial mass arising from the atrial septum and obscuring the tricuspid valve inlet. **(B)** Echocardiogram in a similar projection performed 6 months earlier showing the area to be free of the mass.

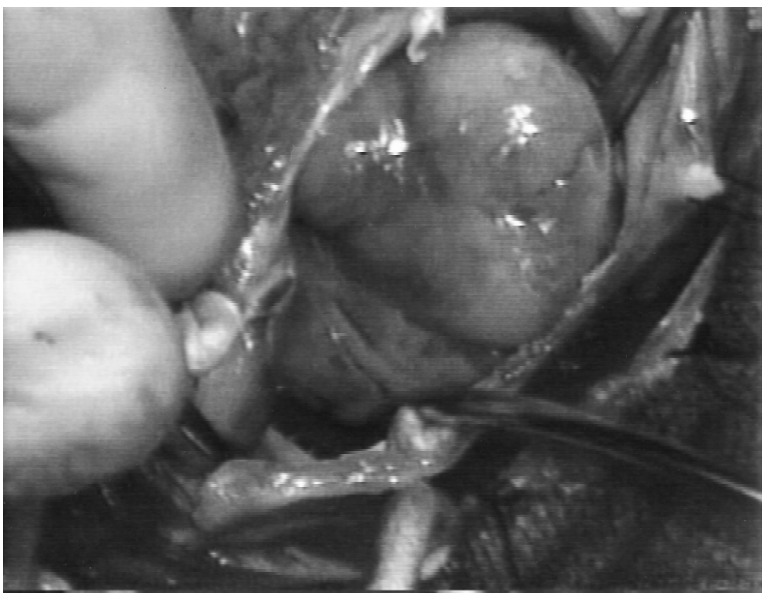

Fig. 2. Intraoperative photograph of the myxoma prior to excision.

cause life-threatening problems. The clinical signs and symptoms with which affected patients present are often nonspecific and may simulate other disease entities [2, 4, 12, 13, 17]. The patient described presented with a history of protracted fevers and lethargy. In a child with congenital heart disease and these symptoms, bacterial endocarditis would be considered the most likely diagnosis, but it must be remembered that fever of undetermined origin can be one of the presenting symptoms of cardiac myxoma as well [7]. Atrial myxomas may affect cardiac function by obstructing flow across the atrioventricular valves, with resultant symptoms of low cardiac output and congestive heart failure. Pulmonary and systemic embolization of tumor fragments have been reported [2, 8]. Right atrial myxomas in the setting of an atrial septal communication may result in paradoxical emboli to the systemic circulation. Rarely, a large tumor obstructs tricuspid valve flow and results in clinical cyanosis due to right-to-left shunting across the atrial septal defect [4]. The latter was obscured in this case by coexisting cyanotic congenital heart disease.

Intracardiac masses can be diagnosed accurately by two-dimensional echocardiography, obviating the need for cardiac catheterization. Although histologic tissue characterization is not possible from echocardiography, certain echocardiographic features (e.g., echo-density,

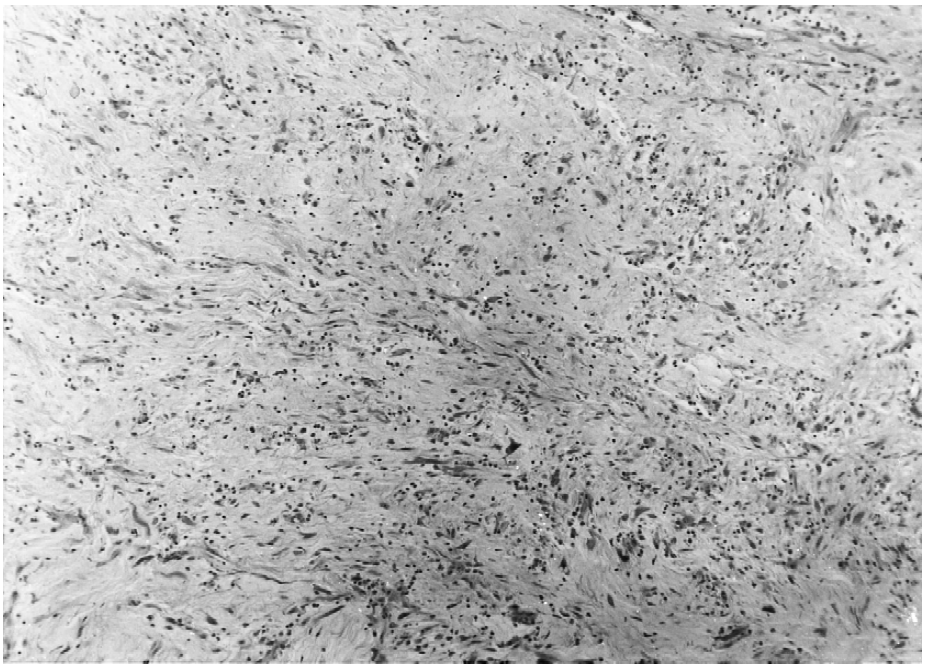

Fig. 3. Cardiac myxoma showing spindle-shaped cells and hyperchromatic nuclei segregated by abundant myxomatoid stroma (H & E, original magnification ×120).

size, shape, and location of the mass) in conjunction with clinical data may prove helpful for categorizing the intracardiac mass. As a result of the unique occurrence of a right atrial myxoma in a child with congenital heart disease, we had the opportunity to view serial echocardiograms performed on our patient as part of her routine cardiology follow-up. An echocardiogram performed approximately 6 months prior to the diagnosis of the right atrial myxoma and all previous echocardiograms showed the right atrium to be free of tumor, suggesting a relatively rapid growth rate for this cardiac myxoma. This finding has been described in reports involving primary and recurrent left atrial myxomas [9, 10, 14, 15]. Aggressive surgical resection is the therapy of choice for these lesions [1]. After the patient was diagnosed as having a right atrial mass consistent with a tumor, she underwent surgical resection of the tumor and repair of her underlying congenital heart disease. Her outcome has been excellent.

In summary, we have described the first case of a cardiac myxoma in a child with congenital heart disease, other than an isolated atrial septal defect. It is unlikely that there is a relation between these two lesions because DORV is not an infrequent congenital heart lesion and no previous association with myxomas has been reported. Additionally, because we had performed a premorbid echocardiogram 6 months prior to diagnosis of the atrial myxoma, we were able to document the rapid growth rate of this tumor.

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