Primary vascular tumors of the heart in infancy. Report of a case with successful surgical management.

M Ilbawi, S DeLeon, T Riggs and R Hernandez

Chest 1982;81;511-512
DOI 10.1378/chest.81.4.511

Primary Vascular Tumors of the Heart in Infancy*

Report of a Case with Successful Surgical Management

Michel Ilbawi, M.D., F.C.C.P.;
Serafin DeLeon, M.D., F.C.C.P.;
Thomas Riggs, M.D.; and Ramiro Hernandez, M.D.

A six-week-old infant with a large vascular tumor of the right atrium was diagnosed preoperatively and treated successfully. This case illustrates the feasibility of radical excision of extensive atrial tumors and restoration of the atrial cavity using autologous pericardium.

Primary vascular tumors of the heart are exceedingly rare, especially in infancy, as is evident by the paucity of such reports in the literature. Diagnosis may be difficult because the symptoms may be similar to those produced by a host of other congenital and acquired cardiac lesions.[1] Surgical experience with these tumors is limited and is complicated by the need for extensive resection of the involved cardiac structures along with the tumor. Satisfactory results are achieved only in patients with solitary well circumscribed lesions.[2,3]

We have successfully resected a hemangioendothelioma of the right atrium in a six-week-old infant. This case is being reported because of its unique nature.

Case Report

The patient is a six-week-old full-term girl who presented with progressive tachypnea, slow feeding, and poor weight gain. Physical examination revealed mild nasal flaring, distant heart sounds, and hepatomegaly.

There was massive cardiomegaly on the chest roentgenogram. Two-dimensional sector echocardiography indicated the presence of moderate pericardial effusion. A cystic mass was seen within the right atrium (Fig 1). Pericardiocentesis yielded straw-colored fluid with no malignant cells. Computerized tomography (CAT scan) following intrapericardial injection of carbon dioxide and contrast medium (Renografin) failed to reveal extracardiac extension of the tumor. Cardiac catheterization and angiography confirmed the presence of a vascular tumor in the right atrium and a small atrial septal defect.

At surgery, external inspection revealed a tumor mass involving most of the atrial free wall without extension into the pericardium (Fig 2). A diagnosis of "benign vascular tumor" was made from a frozen section examination of a biopsy taken from the involved atrial wall.

Using cardiopulmonary bypass and moderate hypothermia, the right atrium was opened. The tumor was vascular, sessile, and attached to the endocardial surface of the free atrial wall. It extended to the junction with the superior vena cava and involved the atrioventricular groove anteriorly and part of the interatrial septum superiorly. The sulcus terminalis and the entrance of the inferior vena cava were free of tumor. The mass was excised with a rim of normal atrial wall; however, a narrow strip of involved tissue near the atrioventricular groove could not be removed due to its proximity to the right coronary artery. The interatrial septal defect was closed with direct sutures, and the atrial wall was reconstructed with pericardium.

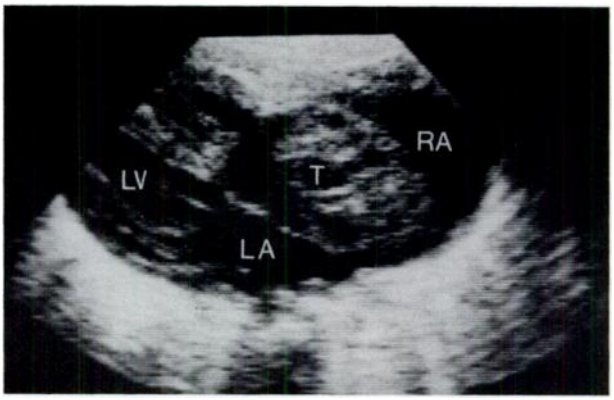

FIGURE 1. A two-dimensional sector echocardiogram showing a cystic tumor (T) in the right atrial cavity (RA), partially attached to the superior aspect of the interatrial septum. LA—left atrium, LV—left ventricle.

Microscopic examination of the tumor revealed numerous small vascular channels lined by flattened endothelial cells and surrounded by spindle-shaped cells. No atypia or mitosis were noted. Reticulin stain confirmed the vascular origin of the tumor and established the diagnosis of benign hemangioendothelioma (Fig 3).

The postoperative course was uneventful. Follow-up sector echocardiogram ten months later demonstrated no residual tumor. Electrocardiogram showed ectopic atrial focus with recurrent runs of atrial arrhythmias.

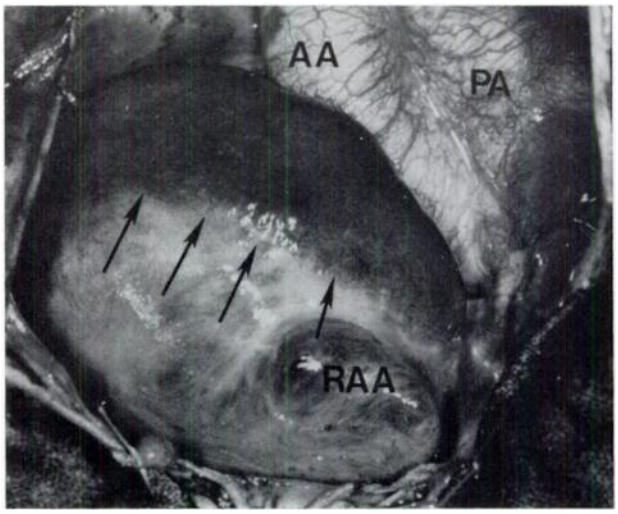

FIGURE 2. An intraoperative view of the tumor. Note the proximity of the tumor to the aortic root. The arrows delineate the portion of the atrial wall grossly involved with tumor. RAA—right atrial appendage; AA—ascending aorta; PA—pulmonary artery.

*From the Divisions of Cardiovascular-Thoracic Surgery and Cardiology (Willis J. Potts Children's Heart Center), and Department of Radiology, The Children's Memorial Hospital, and the Departments of Surgery and Pediatrics, Northwestern University Medical School, Chicago.
Funded by the A. C. Buehler Foundation.
Reprint requests: Dr. Ilbawi, 2300 Children's Plaza, Chicago 60614

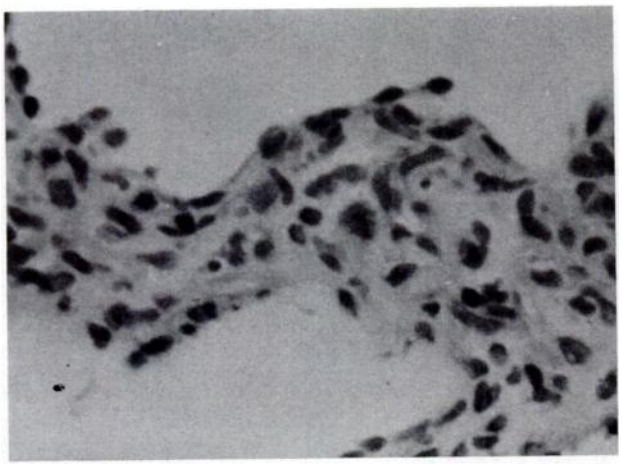

FIGURE 3. The microscopic appearance of the tumor. The vascular spaces are lined with benign endothelial cells.

DISCUSSION

There are few reports in the literature describing the various types of primary cardiac vascular tumors.[3-5] Most of these were in adult patients, and only five cases have been described in infants at postmortem examination (Table 1). To our knowledge, our patient may be the only infant who has had successful radical excision of a vascular tumor of the heart.

The pathogenesis of these vascular tumors is uncertain and it is not easy to determine whether they represent angiomas, hamartomas, telangiectases, varices, blood filled cysts, or even granulation tissue.[6] The majority, except for blood cysts and varicosities, have been subendocardial in origin and without infiltrative features.[4] The right atrium is most commonly involved.[5] They are usually localized and sessile, but could be associated with multiple angiomas of the skin and visceral organs.[7]

Early and accurate preoperative diagnosis was almost impossible in the past because of the wide range of atypical symptoms generated by these tumors.[8] Recently, with the advent of echocardiography, radioactive nucleotide scanning, and computerized tomography, the diagnosis can be made early and with relative ease.

Complete excision of cardiac vascular tumors may not always be possible in spite of early detection because of the extent of the lesion. In our patient, a major portion of the right atrial wall, part of the interatrial septum, and the proximal portion of the superior vena cava were removed and replaced with pericardium. This extensive reconstruction with an akinetic wall does not seem to seriously effect the hemodynamic function of the atrium, and has been used in the surgical treatment of extensive tumors of the atria in older patients.[9,10] However, atrial arrhythmias may result, especially if the sinoatrial node is involved.

Although no longterm survival has been reported in adults with hemangioendothelioma of the heart,[27] the early radical treatment, the benign microscopic appearance of the tumor, and the young age of this patient may improve the prognosis.

Our experience with this case emphasizes the importance of early diagnosis of cardiac vascular tumors in infants, and illustrates the feasibility of successful radical excision of extensive atrial tumors and the use of autologous pericardium for restoration of the atrial cavity in this age group.

Table 1—*Reported Cases of Cardiac Vascular Tumors in Infancy*

Author	Age	Pathology	Diagnosed at
Lymburner[11]	10 mos	Lymphangioma	Autopsy
Nichols[12]	18 days	Endothelium-lined vascular spaces (Lambl's excrescences)	Autopsy
Boyd[13]	1 hour	Endothelium-lined Vascular spaces (Lambl's excrescences)	Autopsy
Schuster[14]	Newborn	Cavernous hemangioma	Autopsy
Gasul[15]	–	Hemangioma	Autopsy
Present case	6 weeks	Hemangioendothelioma	Surgery

REFERENCES

1 Van der Hauwaet LG. Cardiac tumours in infancy and childhood. Br Heart J 1971; 33:125-32
2 Gerbode F, Kerth WJ, Hill JD. Surgical management of tumors of the heart. Surgery 1967; 61:94-101
3 Warembourg H, Niquet G, Ribet M, et al. Hemangiome du coeur. Presse Med 1971; 79:2074-76
4 Sulayman R, Cassels DE. Myocardial coronary hemangiomatous tumors in children. Chest 1975; 68:113-15
5 Tabry IF, Nassar VH, Rizk G, et al. Cavernous hemangioma of the heart; case report and review of the literature. J Thorac Cardiovasc Surg 1975; 69:415-20
6 Bigelow NH, Klinger S, Wright AW. Primary tumors of the heart in infancy and early childhood. Cancer 1954; 7:549-63
7 Berlov GA, Pigarev YUG. Unusual systemic tumor-like angiomatosis (malignant hemangioendothelioma of the pericardium, angiomatous polyp of the endocardium and progressive hemangiomatosis of the liver). Arkh Pat 1964; 26:74-78
8 Longino LA, Meeker IA. Primary cardiac tumors in infancy. J Ped 1953; 43:724-31
9 Cuilliford AT, Isom OW, Trehan NK, et al. Benign tumors of right atrium necessitating extensive resection and reconstruction. J Thorac Cardiovasc Surg 1978; 76: 178-82
10 Kilman JW, Craenan J, Hosier DM. Replacement of entire right atrial wall in an infant with a cardiac rhabdomyoma. J Ped Surg 1973; 8:317-24
11 Lymburner RM. Tumors of the heart: histological and clinical study. Canad MAJ 1934; 30:368-73
12 Nichols JL. Angiomata in valves of heart of a newly born child. J Exper Med 1908; 10:368-70
13 Boyd TAB. Blood cysts on the heart valves of infants. Am J Path 1949; 25:757-59
14 Schuster H. Haemangioma Cavernosum im Herzen cines Neugeborenen. Virchow's Arch f Path Anat 1914; 215: 335-39
15 Gasul BM, Arcilla RA, Lev M. Heart disease in children; diagnosis and treatment. Philadelphia: J. B. Lippincott, 1967; 1086