

Case Report

BLOOD CYST OF THE PULMONARY VALVE: A CASE REPORT

Fulya Çakalağaoğlu* / K. Cantürk Çakalağaoğlu**
Cem Alhan** / Sevgi Küllü*

* M.D., Department of Pathology, School of Medicine, Marmara University, Istanbul, Turkey.

** M.D., Department of Cardiovascular Surgery, Siyami Ersek Thoracic and Cardiovascular Surgery Center, Istanbul, Turkey.

ABSTRACT

Blood-filled cysts of the heart valves are commonly reported at postmortem examination of infants but are rare in older children and adults. We present a case with blood cyst on the pulmonary valve that was surgically removed at the open heart operation. We also review the relevant literature.

Key Words: Blood cyst, Pulmonary valve, Factor VIII.

INTRODUCTION

Blood-filled cysts (BFC) of the heart valves are frequently seen as a postmortem finding on the atrioventricular valves of infants aged <6 months. They are usually small, rounded, pinpoint to pinhead nodules on the submarginal part of the atrial surface of the cups (1-3). Although these cysts seem to disappear after 6 months of age, rare cases have been described in children and adults (4-5). We describe a child who underwent successful surgical excision of a blood cyst from the pulmonary valve.

CASE REPORT

A 12 year old boy was admitted to the Siyami Ersek Thoracic and Cardiovascular Surgery center with exertional dyspnea and cyanosis. He looked healthy. Physical examination was normal except for the heart. There was a palpable systolic thrill, and palpable pulmonary valve closure in the second left intercostal space. An opening click of the pulmonary valve and a grade 2 to 3/6 systolic ejection murmur was heard at the same area. A pulmonic second sound was

accentuated and relatively fixed splitting was established.

His pulse was regular with a rate of 78 beats/minute. Blood chemistry and other laboratory tests were all within normal limits. His electrocardiogram showed features of right ventricular overload and cardiac catheterization disclosed a systolic pressure of 80-120mmHg in the right ventricle with a pressure gradient of 65 mmHg after advancing the catheter into the pulmonary trunk. The cardiac index and left ventricular ejection fraction were within normal limits. These findings are showed pulmonary stenosis.

On January 15, 1998, open heart surgery was performed. At operation, the pulmonary artery was seen to be greatly dilated and a systolic thrill was palpated over the pulmonary trunk. During surgery, a cystic mass was found near the free margin of the left posterior pulmonary valve cusp on the atrial surface. It was successfully resected. During the excision the cyst was opened and bloody fluid flowed out.

Histopathological Finding: The specimen received consisted of a round cyst-like structure, measuring 4 x 6 mm in diameter (Fig 1). It had a 2 mm thick wall of white fibrous tissue that was translucent enough to show dark blue content. The outer surfaces were smooth and glistening. On sectioning, it oozed old bloody fluid and left an empty space with some blood clot. The surface of the cystic space was also smooth and glistening. Microscopically, the cyst wall was composed of dense collagenous tissue which was consistent with cardiac valvular tissue. Both the luminal and outer surfaces were lined with a monolayer of flat cells (Fig 2). A blood clot and calcified bodies of small thrombi were found within the cyst.

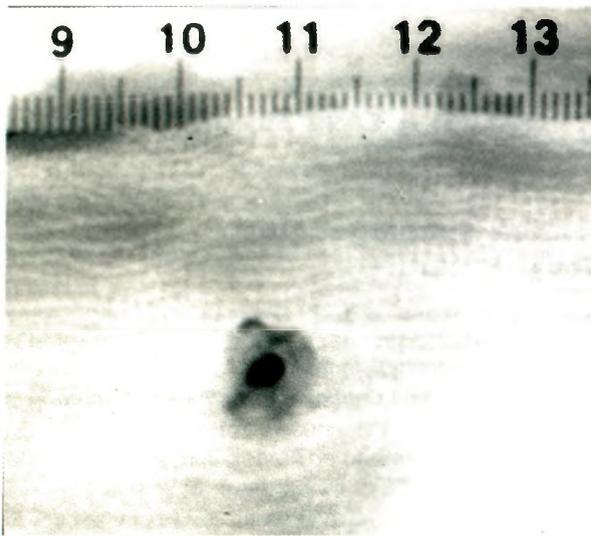


Fig.1: Gross photograph of the cyst.

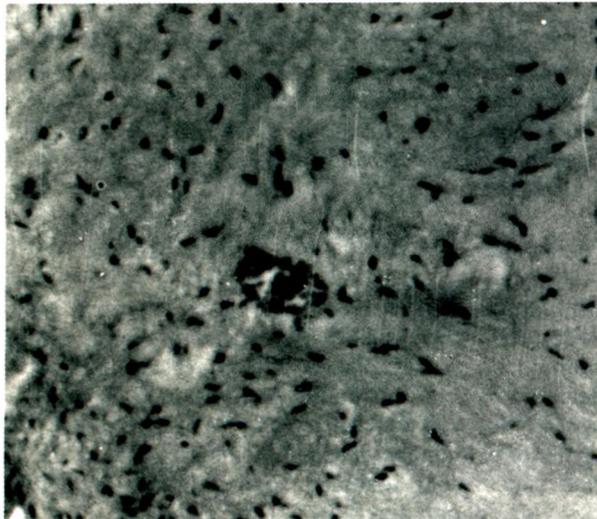


Fig.2: The cyst wall was lined by a monolayer of flat cells. (Hematoxylin-Eosin stain, 40X).

We performed immunostaining using standard avidin biotin peroxidase complex methods with antibody against factor VIII (Biogenex). Flat cells were stained with that antibody. The result was consistent with endothelial cells as was thought histologically.

After operation, no murmur was heard. The patient had an uneventful recovery.

DISCUSSION

Blood filled cyst of the heart valves was first reported in 1844 by Elsasser (1). BFC are frequently found on

the atrioventricular valves of necropsies newborn infants when their incidence ranges from 25% to almost 100% (1-5). They are usually seen as small, rounded, multiple nodules on the atrial surfaces of the atrioventricular valves, but are also seen less often on the ventricular surfaces of the semilunar valves (1-3).

The single valvular blood cysts occurring in older children and adults have seldom been reported. To date Liese et al, Cumming and Ferguson and Sakakibara et al, Paşaoğlu et al, Minato et al reported 12 of BFC of the pulmonary valve that were successfully treated by surgical removal.

Several explanations have been given regarding the origin of blood cysts. According to a well-known theory cysts are formed when blood enters into crevices on the surface of the valve cusp, with a subsequent sealing off of the portal entry (1-3). In our case, the cyst was filled with bloody fluid. This finding was sufficient to support Boyd's pathogenetic theory mentioned above. Another opinion is that in the case of sudden occlusion, such as that caused by inflammation, vagal stimulation, anoxia and hemorrhagic diathesis branches of the circulation behave as end arteries and their occlusion leads to hematoma of the subvalvular region (1-5).

The blood cysts of the pulmonary valve were usually on the arterial surface of the posterior cusp (1-3). The cysts projected above the surface of the valves near the free margin frequently in both infants or adults. In our case, it was on the arterial surface of the right cusp near the free margin.

Histologically, the cysts were usually lined with flat endothelium-like cells on both sides and contained a bloody fluid. In present case endothelial cells were immunopositive, confirming their endothelial nature (1).

Clinically, a blood cyst should be considered when echocardiograph shows a mass on a cardiac valve in patients with a systolic murmur and symptoms suggesting valvular stenosis (4-5).

Usually pedunculated lesions on the valves are readily excised, but simple incision of the cyst might be more effective than excision because the latter would result in the deformity of the valve (5-7). There have been no reports of its recurrence after surgery.

In summary, we reported a case of a blood cyst of the pulmonary valve, with review of the literature. Blood cysts of the cardiac valves usually have no clinical significance, but when they are on the pulmonary valve, they often contribute to stenosis of the valves.

REFERENCES

1. Minato H, Manabe T, Masaki H, Kawahara Y. Blood cyst of the pulmonary valve in an adult: Report of a case and review of the literature. *Hum Pathol* 1997;28:252-255.
2. Zwart HHJ, Jenson CB, Knight JA, et al. Blood-filled cyst on heart valves. *J Pediatr Surg* 1973;8:333-334.
3. Paşaoğlu I, Doğan R, Demircin M, et al. Blood cyst of the pulmonary valve causing pulmonic valve stenosis. *Am J Cardiol* 1993;72:493-494.
4. Ohmoto Y, Tsuchihashi K, Tanaka S, et al. Giant endocardial blood cyst in left ventricle resected by transaortic valve approach. *Chest* 1993;103:965-966.
5. Paşaoğlu I, Doğan R, Oram A, et al. Giant blood cyst of the left ventricle. *Japan Heart J* 1991;32:147-151.
6. Raskoff WJ, Wagner S, Hill JD. Pulmonary valve cyst in an adult. *J Thorac Cardiovasc Surg* 1976;72:450-453.
7. Sim EKW, Wong ML, Tan TK, Sim SK. Blood cyst of the tricuspid valve. *Ann Thorac Surg* 1996;61:1012-1013.