SUBMITTED 10 JAN 23
REVISION REQ. 23 MAR 23; REVISION RECD. 30 MAR 23
ACCEPTED 3 MAY 23
ONLINE-FIRST: MAY 2023
DOI: https://doi.org/10.18295/squmj.5.2023.037

Congenital Blood Cyst of a Child

A Case Report and Review of Literature

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Abstract

Blood-filled cysts of the heart valves are frequently reported at postpartum autopsies of infants. They are seen as round nodules mostly in the paediatric age group in infants less than two months of age and disappear spontaneously within six months of life. We present a unique case of an 11-month-old girl with a blood-filled cyst on the posterior leaflet of the pulmonary valve that was successfully treated. This case report highlights the characteristics and course of a paediatric patient with blood-filled cysts. Further studies are yet needed to better understand the diagnostic approaches to blood-filled cysts as well as treatment modalities to fill the gap in clinical settings.

Keywords: Blood filled cysts; Pulmonary valve; Pulmonary artery; Paediatrics; Cardiac tumor; Cardiology.

Introduction

Blood-filled cysts (BFCs) are rare benign tumors mainly reported as cardiac tumors.[1] Since primary cardiac valve tumors are very uncommon, autopsy studies have provided much of the current detailed information available in the literature.[2, 3] Numerous parameters, such as tumor classification, location size, growth rate, and susceptibility to embolize, affect the overall clinical presentation. Both myxomas and papillary fibroelastomas are the pathological conditions most likely to be related to embolism, with the latter making up most primary valve tumor types.[3, 4] Moreover, lipomas, myxomas, and rhabdomyomas are all reported as primary cardiac tumors as well.

BFCs of the heart valves were first reported in 1844 by Elsasser.[5] BFCs are often found on the atrioventricular valves of newborn infants' necropsies. 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] In this report we present a rare case of a paediatric patient with a BFC on the pulmonary valve leaflet.

Case Report

An 11-month-old girl presented to her general paediatrician for a non-cardiac cause at the beginning of the year 2022. She was noted to have a loud systolic heart murmur in the pulmonary area. The child was otherwise well, had no previous history of infections, and has normal clinical examination findings otherwise.

The echocardiogram demonstrated severe right ventricular outflow obstruction due to a possible cyst on the posterior leaflet of the pulmonary valve. The valve itself looked normal, and there was post-stenotic dilatation of the main pulmonary artery. The gradient across the pulmonary valve was 65 mmHg peak.

The patient underwent a right heart catheterization. The angiogram showed a cyst fixed on the surface of one of the pulmonary valve leaflets (Figure 1). The cyst was mobile with the leaflet, but not causing any regurgitation. No ballooning was done, and the

patient had a chest CT scan which showed a filling defect in the main pulmonary artery. The CT scan showed normal distal pulmonary arteries and branches, as well as normal lung parenchyma, mediastinum, and no lymphadenopathy. A comprehensive infectious and immunological assessment showed no underlying disease.

Following the discussion with our multidisciplinary team (MDT), we decided to surgically remove the cyst (Figure 2). The surgical procedure was uneventful. Resection of the whole cyst that was attached to the posterior leaflet of the pulmonary valve was performed, and the gradient decreased to less than 15 mmHg, with moderate pulmonary valve regurgitation.

The histopathology of the cyst (Figure 3) showed a 1*0.5*0.3cm multiloculated cyst, filled with blood, and composed of a thin fibrous wall with focal myxoid changes. The child made full recovery. At age of three, only a 12-mmHg peak gradient at the pulmonary valve was observed, with moderate regurgitation, and no new cysts were noted on any of the heart valves.

Informed and written consent for the patient's procedure and publication purposes for this case report was obtained from the parents.

Discussion

This report presents a unique case of a paediatric patient with a BFC on the posterior leaflet of the pulmonary valve that was successfully managed and treated. Unlike the paediatric age group, singular valvular BFCs are rarely reported in older children and adults, this is attributed to the fact that the cysts spontaneously regress in most patients as they age.[6] Liese et al., Cumming and Ferguson and Sakakibara et al., Pa§aoglu et al., and Minato et al. reported 12 BFCs of the pulmonary valve that were treated successfully by surgical resection.[5-12]

BFCs presents with symptoms of severe valvular stenosis due to the outflow obstruction, as well as regurgitation presenting with signs of cyanosis, although they have mostly

been reported to be asymptomatic and only discovered incidentally usually on CT scans done for non-cardiac reasons, especially in the infant age group.[6,12] The patient presented in this case report, initially went to the general paediatrician (GP) with no cardiac symptoms, and the leading cause of the BFCs discovery was due to a loud systolic murmur detected by the GP. Following that, she was referred to the cardiology department and an echocardiogram was done which identified the BFC. Echocardiogram is considered to be the gold standard for the diagnosis of BFCs, and in rare cases where a thrombus or bacterial vegetations are suspected, contrast echocardiogram might help differentiate it from cardiac cysts.[13]

To date, it is still unknown where the source of BFCs in cardiac valves arise from. Several animal studies revealed that 20% of all animal hearts contained BFCs, highlighting its high prevalence.[14] Regarding the development of BFCs, adult cases have been attributed to blunt trauma to the chest and complications during valvular surgeries, however, their cause in the paediatric age group is still unknown.[13] Tsutsui et al. suggested that BFCs may originate during the development of valves in early embryogenesis or during early period of life from blood entrapped in valvular crevices or tiny invaginations during development, therefore, a neonate with normal echocardiogram findings can still develop BFCs in early infancy or childhood, while this process is very unlikely to occur at later stages during adulthood.[15] Another hypothesis suggests that BFCs are primarily due to hematoma formation as a result of blocked small vessels.[12] Furthermore, based on the findings of the histological and ultrastructural analysis, BFCs could be due to the expansion of thin-walled valvular arteries in response to mechanical stress caused by the pressure gradient when atrioventricular valves are closed, developing a cyst.[14] However, the presence of BFCs in low-pressure structures like the pulmonary valve cannot be explained by the mentioned theory. Therefore, the mentioned hypotheses are quite hard to confirm and the definite formation of a BFC is not yet well established in the current literature.

In terms of management, Paşaoğlu et al. suggested surgically removing the BFCs in the heart at the time of diagnosis independent of the patient's symptoms.[10] This was the

course of action for the patient presented in this case report. On the other hand, Dencker et al. encouraged a more conservative approach in asymptomatic patients and stated that surgical approaches should be kept for symptomatic patients or if the cyst leads to cardiac dysfunction.[13] Surgical interventions are usually done in order to rule out malignancy and risk of strokes. Pharmacological therapies including anticoagulants and beta-blocker use are still controversial with very little evidence available in the current literature.[13] This emphasizes the need for more research exploring the outcomes of different management approaches.

Conclusion

In summary, we have reported a rare case of BFC above the pulmonary valve in addition to a literature review. Although several theories have been postulated regarding the origin of BFCs, it remains unknown. BFCs are rare and are seen as small round nodules on imaging modalities. However, a better understanding of the diagnostic approaches to BFCs as well as treatment modalities is required to ensure an overall better prognosis in both the adult and paediatric age groups.

Conflicts of Interest

The authors declare no conflict of interests.

Author Contributions

RK, IER and MK conceptualized and designed the work. RK, IER and MK collected analysed and interpreted the data. RK and MK drafted the manuscript. ZAA and MK revised the manuscript. All authors approved the final version of the manuscript. Rachid Kaddoura: drafting the manuscript.

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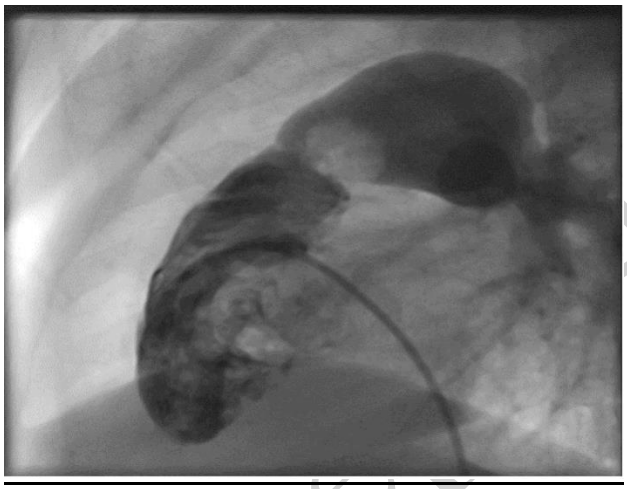

Figure 1: Right heart catheter showing circumscribed rounded cysts

Figure 2: Surgical removal of the cyst

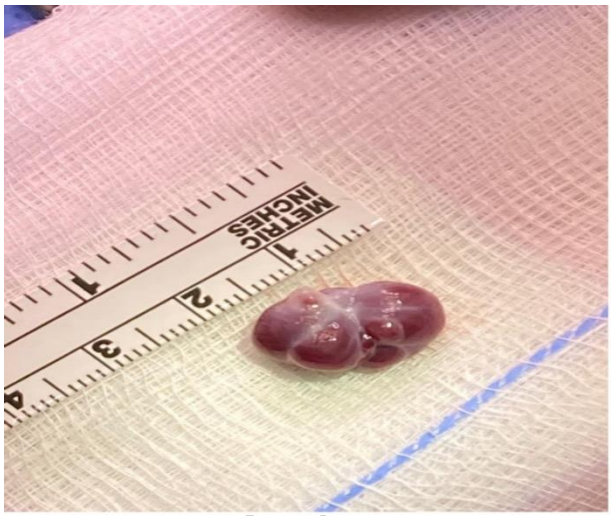

Figure 3: Gross appearance of the cysts