Pediatric Right Atrial Mass With Pulmonary Embolization

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INTRODUCTION

Pediatric cardiac masses involving the right atrium (RA) are extremely rare but have the potential for severe complications. The differential for a right atrial mass is broad, including benign tumors, metastatic disease, and thrombi, and they share similarities on imaging, making the diagnosis challenging.[1,2] The range of complications depends on the type, shape, size, and location of the mass. In cases of cardiac myxomas and thrombi, pulmonary embolization may occur.[3,4] The management of a right atrial mass needs to be individualized based on the pathology and clinical presentation; however, anticoagulation and surgical resection are important options.[4] We present an infant with a right atrial mass and pulmonary embolization requiring urgent surgical resection and tricuspid valve (TV) repair.

CASE PRESENTATION

A 22-month-old fully immunized girl with noncontributory past medical, surgical, and birth history initially presented with cough, intermittent congestion, a few episodes of nonbloody, nonbilious emesis, decreased oral intake, and periorbital edema. Their symptoms progressed to lethargy and generalized weakness, prompting presentation to a clinic where the patient was found to be tachypneic and pale with perioral cyanosis and signs of poor perfusion: cool extremities and delayed capillary refill. Physical examination was remarkable for pallor, tachycardia, bilateral lower and upper extremity edema, delayed capillary refill without murmurs or gallops, and absence of hepatosplenomegaly.

Laboratory studies revealed severe microcytic anemia with iron deficiency (hemoglobin, 1.7 g/dL; mean corpuscular volume 50.4 fL; Fe 17, μ/dL; ferritin, 11 ng/mL; transferrin saturation, 4.7%; red cell distribution width, 32.2%; 1.32%, reticulocytes). The peripheral blood smear showed hypochromic red blood cells without schistocytes, spherocytes, Howell-Jolly bodies, basophilic stippling, dysplastic white cell morphology, or blasts. Venous blood gas demonstrated a pH of 7.042 and bicarbonate 5.6 mmol/L with marked elevation in serum lactic acid to 14.6 mmol/L, suggesting significant metabolic acidosis. D-dimer and pro b-type natriuretic peptide were elevated at 1.44 μ/mL and 29,455 pg/mL, respectively. The labs also revealed a mild coagulopathy with an international normalized ratio of 1.4 and prothrombin time of 17.2 seconds. Infectious etiology workup was unremarkable with a negative blood culture, viral panel, and urinalysis. Initial chest x-ray demonstrated bilateral hazy opacities without cardiomegaly. A point-of-care ultrasound revealed a right atrial mass with pericardial effusion.

Further diagnostic imaging was pursued given the presentation of acute hypoxic respiratory failure and a low cardiac output state. A chest computed tomography (CT) angiogram (Figure 1) reveled a nonocclusive embolus within a subsegmental branch of the pulmonary artery supplying the left lower lobe and apparent filling defect within the RA and extending into the inferior vena cava (IVC) that was concerning for a pathologic mass.

A transthoracic echocardiogram demonstrated a large, pedunculated, nonobstructive mobile mass in the RA (measuring 44 mm $\times$ 13 mm; Figure 2, Videos 1 and 2) that appeared to be attached to the IVC with an in-and-out mobility into the right ventricle (RV) through the TV (Figures 3 and 4, Videos 3 and 4).

A moderate pericardial effusion was noted anterior to the RA (20 mm) without evidence of tamponade physiology. Mild mitral regurgitation and tricuspid regurgitation (TR) were seen. The TR jet velocity was approximately 2.6 m/sec, suggesting minimally elevated right heart pressure. There was no evidence of septal flattening. There was moderate biventricular dilatation with normal systolic function (left ventricular ejection fraction of 64%). Presence of a solitary kidney was incidentally discovered on CT.

The patient was admitted to the intensive care unit for management of severe microcytic anemia and a large right atrial mass with left subsegmental pulmonary embolism. The patient was initially supplemented with increased fraction of inspired oxygen of 100% to acutely augment tissue oxygen delivery in the setting of low hemoglobin. The diagnosis of severe nutritional iron deficiency anemia (IDA) was made based on laboratory findings of iron deficiency, hypochromic red blood cell morphology visualized on peripheral blood smear, and history of inadequate iron intake. Given the severity and suspected chronicity of the patient's IDA, gradual serial transfusions of 5 mL/kg packed red blood cells were pursued to maintain hemodynamic stability and prevent pulmonary edema, with appropriate rise in hemoglobin to 7.6 g/dL. Iron deficiency was managed with intravenous iron supplementation. Anticoagulation with a direct thrombin inhibitor was initiated with addition of aspirin for its antiplatelet function in the setting of reactive thrombocytosis to 1,231 10^3/uL. Given the possibility of an intracardiac thrombus, an evaluation for hypercoagulability was pursued; however, it was found to be inconclusive—homozygous pattern without prothrombin G20210A mutation with normal activated protein C resistance, indicating absence of factor V Leiden mutation. The family history was unremarkable for any hereditary hypercoagulability without reported history of thrombosis, coagulopathies, or anemia.

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VIDEO HIGHLIGHTS

Video 1: Two-dimensional transthoracic echocardiogram, subcostal sagittal view, demonstrates a large echobright mass attached to wall of the IVC and highly mobile within the RA.

Video 2: Two-dimensional transthoracic echocardiogram, apical 4-chamber view, demonstrates mild biventricular dilation, normal biventricular systolic function, and a large echobright mass in the RA crossing the plane of the TV.

Video 3: Two-dimensional transesophageal echocardiogram, midesophageal RV-focused 4-chamber (−4°) view, demonstrates normal RV systolic function and a large echobright mass in the RA crossing the plane of the TV.

Video 4: Two-dimensional transesophageal echocardiogram, midesophageal, basal short-axis RV outflow tract (50°) view, demonstrates a normal aortic valve and a large mobile echobright mass in the RA.

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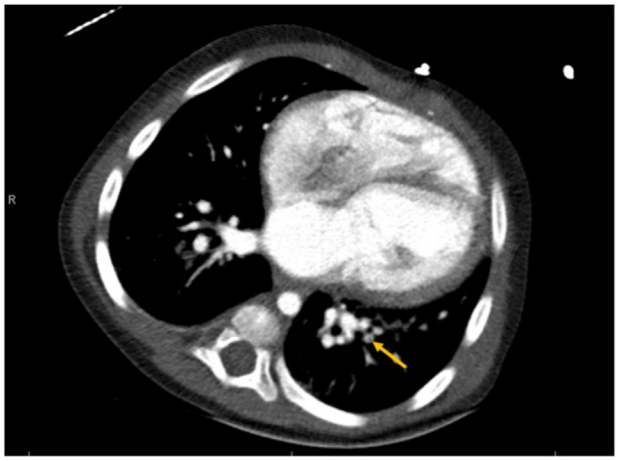

Figure 1 Contrast-enhanced CT of the chest, axial display, demonstrates biventricular dilation and a filling defect in the RA and in the subsegmental left lower lobe pulmonary artery branch (*arrow*).

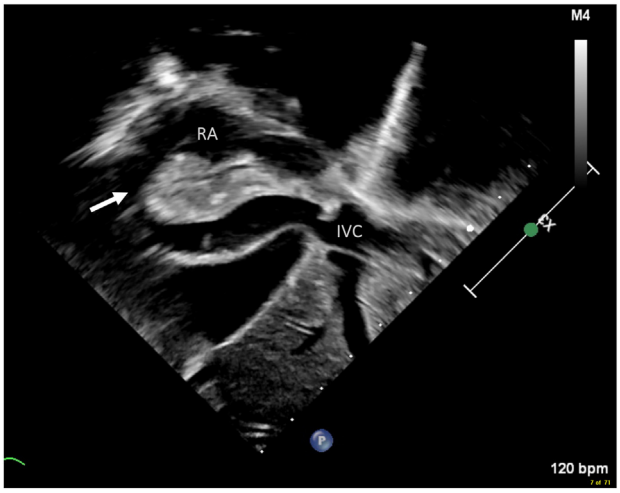

Figure 2 Two-dimensional transthoracic echocardiogram subcostal sagittal midsystolic view demonstrates a large echobright mass (*arrow*) attached to wall of the IVC and protruding into the RA.

Given the size of the right atrial mass and risk for continued embolization, emergent cardiac surgery for removal of the mass was pursued within 2 days of admission once the patient had favorable hemodynamics with improved acidosis. The patient's hemoglobin and pro b-type natriuretic peptide improved to 7.7 g/dL and 8,628 pg/mL, respectively, prior to surgery. Intraoperatively, a highly mobile oval mass was visualized within the RA and was in contact with the TV (Figure 5).

The dilated RV and enlarged TV annulus suggested the chronicity of the mass. The excised mass measured 45 mm in length × 15 mm in width and was attached to the inside of the IVC with a 1 mm stalk (Figure 6). There was evidence of damage to the TV, demonstrated by thickened edges of the TV leaflets, suggesting chronic mechanical stimulation from prolapse of the right atrial mass during the cardiac cycle. The mass was completely excised without residual findings on the postoperative echocardiogram. A TV repair with annuloplasty was completed to reduce the annulus size with no significant residual TR on postoperative echocardiogram. Pathology results of the excised intracardiac mass revealed a degenerated blood clot with calcifications. The patient made a satisfactory postoperative recovery and was eventually discharged on long-term anticoagulation with a competitive inhibitor of factor Xa. Following discharge, the patient was closely followed by hematology. They had complete resolution of IDA following iron supplementation and dietary changes. Anticoagulation was continued with plans for repeat imaging to reassess the pulmonary embolus after 6 months of therapy.

DISCUSSION

Initial considerations for any pediatric patient with an intracardiac mass should include benign or malignant primary cardiac tumors, metastases, or thrombi.[5] Despite being relatively uncommon in children, and comprising ~10% of benign cardiac masses,[2] cardiac myxoma was initially the suspected etiology in our case. The appearance of the mass on echocardiogram and the absence of significant risk factors for thrombus formation guided our reasoning. Typically, myxomas are friable, gelatinous masses arising in the left atrium (75%). In 15% of cases, however, they can arise in the RA.[6] Echocardiogram findings play an important role in the diagnosis of myxomas; they appear heterogenous with irregular calcifications.[1,7] Structurally, they contain a single large, highly mobile component that attaches to the atrial wall by a short, broad-based pedicle.[7] The pedunculated nature of myxomas allows for possible intracardiac obstruction or prolapse of the highly mobile component through the atrioventricular valve. Additionally, fragments of the tumor can detach, leading to pulmonary embolism.[3] Therefore, they are typically removed surgically upon diagnosis.

Intracardiac thrombi are rare in pediatric patients and are usually provoked by underlying conditions such as hypercoagulability, arrhythmias, or low cardiac output.[8] Additionally, studies have shown increased incidence in postoperative patients following Fontan or cardiac transplantation, presumedly due to altered physiology and

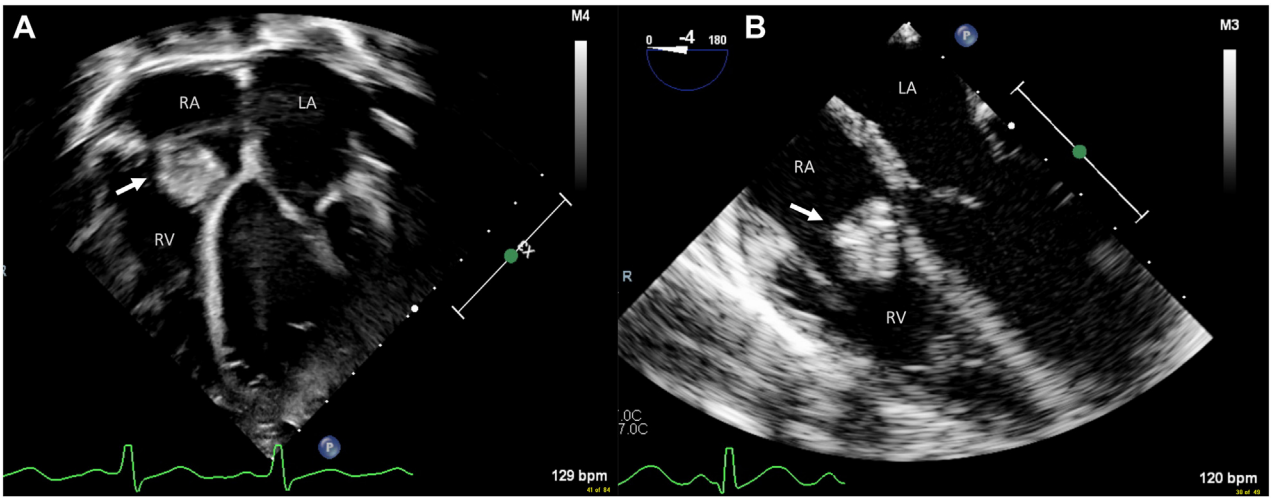

Figure 3 Two-dimensional transthoracic echocardiogram in the 4-chamber view **(A)** and two-dimensional transesophageal echocardiogram midesophageal, RV-focused 4-chamber view **(B)** in middiastole demonstrate mild biventricular dilation and a large echobright mass (*arrow*) in the RA crossing the plane of the TV. *LA*, Left atrium.

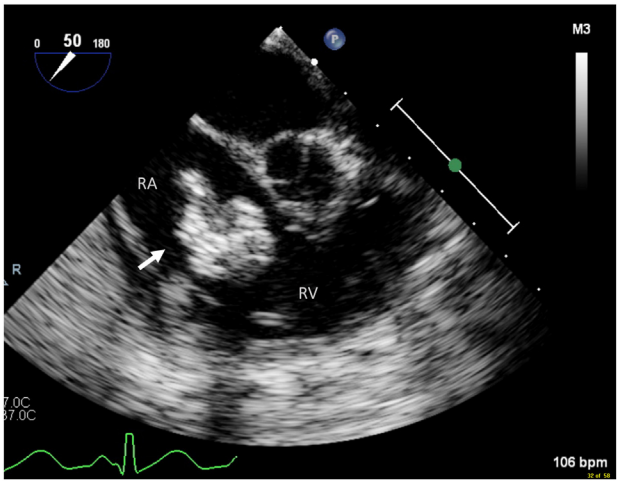

Figure 4 Two-dimensional transesophageal echocardiogram, midesophageal, basal short-axis RV outflow tract (50°) diastolic view, demonstrates a normal aortic valve and a large, mobile echobright mass (*arrow*) in the RA.

inflammation.[8] Intracardiac thrombi appear as homogenous masses on echocardiogram. They can develop within any of the cardiac chambers, forming pedunculated or sessile masses attached to the walls of vasculature or endocardium.[2] While other benign tumors such as rhabdomyoma and cardiac fibromas are more common in pediatric patients, they were unlikely in our patient based on clinical presentation and imaging.

On primary evaluation of our patient, the presence of a pedunculated, highly mobile mass with scattered calcifications was concerning for either a myxoma or intracardiac thrombus. It can be difficult to distinguish myxoma from intracardiac thrombus on imaging features. Both conditions have a wide variety of documented shapes, sizes, locations, and clinical presentations. Given the lack of a known hypercoagulable condition that may precipitate clotting, thrombus was considered less likely. The location of the mass and its attachment to the IVC, however, was unlikely for a myxoma.[1] Advanced imaging modalities such as cardiovascular magnetic resonance imaging and cardiac CT may aid in the detection and characterization of intracardiac masses; however, given the fragile hemodynamics of our patient, we proceeded to intervention. Administration of an ultrasound-enhancing agent can improve intracardiac thrombus detection by providing contrast opacification of the cardiac chambers and show an intracardiac thrombus as a filling defect. This is especially valuable in patients with poor echocardiographic windows and suspicion for a thrombus. In the current case, as there was clear visualization of the thrombus on multiple two-dimensional views, contrast echocardiography was not attempted.[9] Contrast echocardiography is also valuable in visualizing vascularity, aiding in the distinction between a tumor or thrombus, and should be considered in similar cases.[10]

The presence of a sizable intracardiac mass with evidence of pulmonary embolization, regardless of its histological origin, demanded prompt management. Pulmonary embolism in the present case is not unexpected as it is a well-described complication of right heart thrombi.[4,11] Previous studies have reported pulmonary embolism in nearly 71% of those with identified right heart thrombi.[4] In the case of atrial or ventricular shunts or structurally abnormal intracardiac connections, systemic embolization may occur.[2] In a previous study of intracardiac thrombi, the shape and mobility, rather than size, were significant predictors of risk of embolization.[12] Thrombocytosis, causing platelet aggregation, has previously been identified as an additional important risk factor for embolization among those with an intracardiac mass.[3] For this reason, previous studies have proposed the role of low-dose aspirin in these circumstances.[13] Iron deficiency anemia, in the absence of thrombocytosis, has also been previously associated with thrombosis.[13] This rate of thrombosis is nearly doubled in cases of both IDA and thrombocytosis.[13] The synergistic effect of these variables on thrombus formation was manifested in the present case.

The guidelines on the management of right atrial masses differ based on their pathology, and their optimal management is still unclear. In case of an intracardiac thrombus, management options include surgical thrombectomy, anticoagulation, or thrombolysis. A

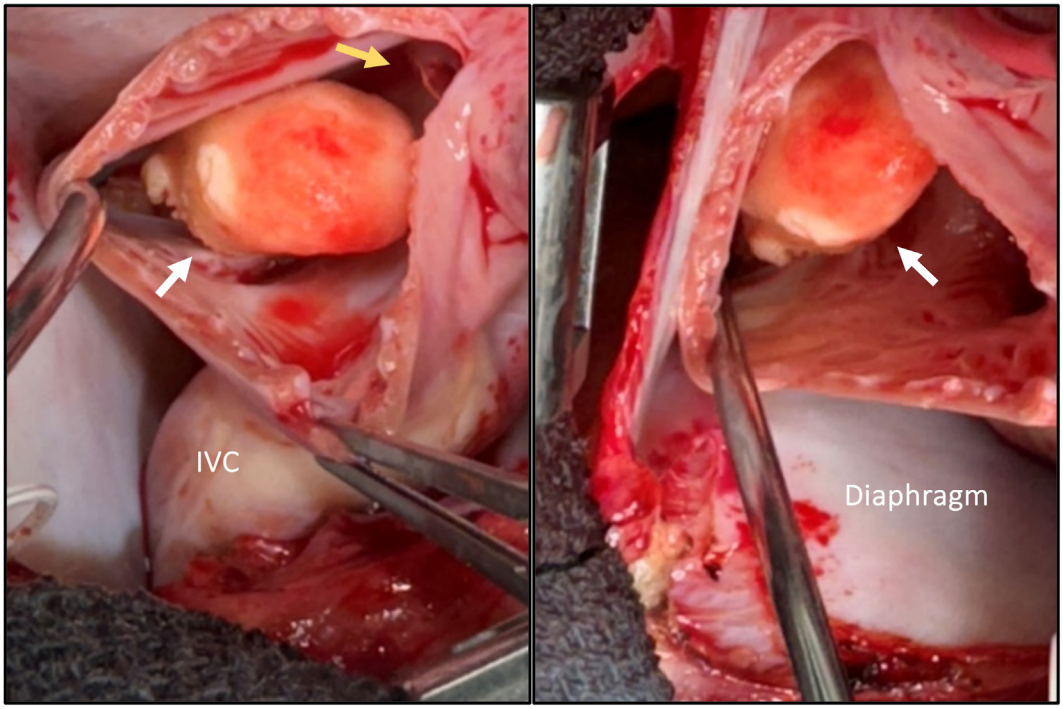

Figure 5 Intraoperative images from the surgeon's perspective of the mass (*white arrows*) within the RA rotated slightly (*left panel*) to demonstrate the proximity to the TV (*yellow arrow*) and the anatomically correct orientation (*right panel*) with the diaphragm positioned inferiorly.

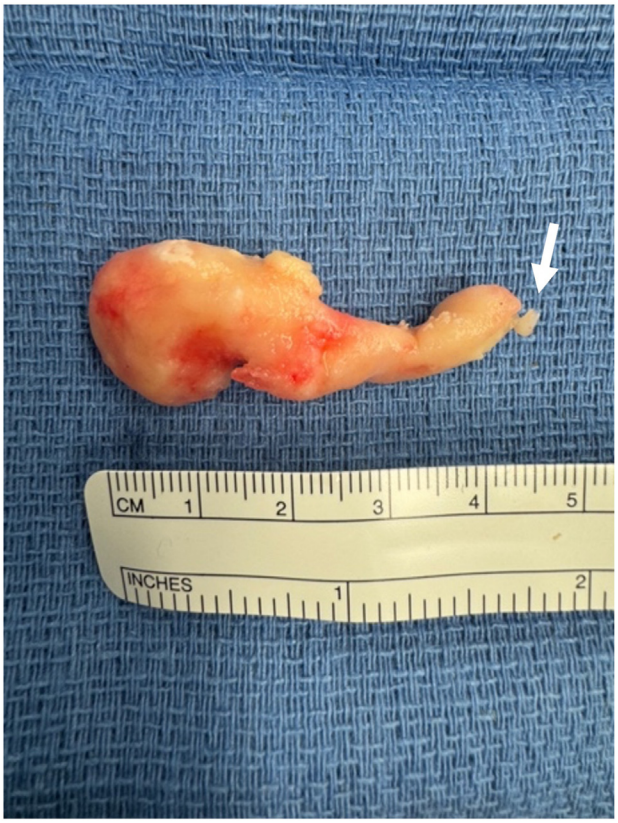

Figure 6 Gross specimen of the resected mass measuring 45 × 15 mm. The tip of the mass (*arrow*) was attached deep within the IVC near the level of the diaphragm.

previous study pursued by the European Working Group on Echocardiography aimed to identify management based on thrombus morphology and site of thrombus origination.[14] Thrombi characterized as type A were mobile and originated outside of the heart, while type B were largely immobile and arose within the heart.[14] Mortality was found to be higher in patients with type A features compared to those with type B characteristics regardless of management strategy. Additionally, the mortality for patients with type A thrombi was significantly lower in those receiving surgical intervention compared to anticoagulation alone.[14] In accordance with these guidelines, given the mobility of the mass, the evidence of pulmonary thromboembolism, and the potential for valvular damage observed, surgical excision was pursued.

Based on a through literature review of the prevalence and causes of right atrial thrombus formation in pediatric populations, a vast majority of right atrial thrombi in children are associated with central venous catheterization, premature birth, and underlying malignancy. There is a paucity of literature describing cases of unprovoked thrombus formation. Here we report a detailed account of our diagnostic approach, interventions, and long-term management, thereby offering insights that may enhance clinical practice.

CONCLUSION

The presence of a sizable, mobile right atrial thrombus and subsequent pulmonary artery embolus had significant hemodynamic consequences in this patient. Although the mechanism of thrombus formation is not entirely explained, we must consider the concurrent impact of IDA and thrombocytosis in the risk of embolization and role of anticoagulation. Echocardiography was an important tool in the

characterization of the mass and its effects on adjacent structures, guiding its resection.

ETHICS STATEMENT

The authors declare that the work described has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) for experiments involving humans.

CONSENT STATEMENT

The authors declare that since this was a non-interventional, retrospective, observational study utilizing de-identified data, informed consent was not required from the patient under an IRB exemption status.

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DISCLOSURE STATEMENT

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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SUPPLEMENTARY DATA

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REFERENCES

1. Scheffel H, Baumueller S, Stolzmann P, Leschka S, Plass A, Alkadhi H, et al. Atrial myxomas and thrombi: comparison of imaging features on CT. Am J Roentgenol 2009;192:639-45.
2. Poterucha TJ, Kochav J, O'Connor DS, Rosner GF. Cardiac tumors: clinical presentation, diagnosis, and management. Cur Treat Options Oncol 2019; 20:66.
3. He DK, Zhang YF, Liang Y, Ye SX, Wang C, Kang B, et al. Risk factors for embolism in cardiac myxoma: a retrospective analysis. Med Sci Monit 2015;21:1146-54.
4. Lahmidi I, Assoweh DC, Haddiya I, Bentata Y, El Ouafi N, Ismaili N. Clinicopathological features of adult right-sided cardiac masses: analysis of 19 cases. Ann Med Surg 2022;77:103613.
5. Ghadimi MM, Lu JC, Rigsby CK, Krishnamurthy R, Dorfman AL, Agarwal PP. MRI of pediatric cardiac masses. AJR Am J Roentgenol 2014;202:971-81.
6. Wu JC. Chapter: 37 - primary and secondary tumors. In: Solomon SD, Wu JC, Gillam LD, editors. Essential Echocardiography. Philadelphia, PA: Elsevier; 2019:385-391.e381.
7. Uzun O, Wilson DG, Vujanic GM, Parsons JM, De Giovanni JV. Cardiac tumours in children. Orphanet J Rare Dis 2007;2:11.
8. John JB, Cron SG, Kung GC, Mott AR. Intracardiac thrombi in pediatric patients: presentation profiles and clinical outcomes. Pediatr Cardiol 2007;28:213-20.
9. Cohen A, Donal E, Delgado V, Pepi M, Tsang T, Gerber B, et al. EACVI recommendations on cardiovascular imaging for the detection of embolic sources: endorsed by the Canadian Society of Echocardiography. Eur Heart J Cardiovasc Imaging 2021;22:e24-57.
10. Cotter B, Raisinghani A, DeMaria AN. Established and emerging roles for ultrasound enhancing agents (contrast echocardiography). Clin Cardiol 2022;45:1114-22.
11. Rose PS, Punjabi NM, Pearse DB. Treatment of right heart thromboemboli. Chest 2002;121:806-14.
12. Abe Y, Asakura T, Gotou J, Iwai M, Watanabe Y, Sando M, et al. Prediction of embolism in atrial fibrillation: classification of left atrial thrombi by transesophageal echocardiography. Jpn Circ J 2000;64:411-5.
13. Song AB, Kuter DJ, Al-Samkari H. Characterization of the rate, predictors, and thrombotic complications of thrombocytosis in iron deficiency anemia. Am J Hematol 2020;95:1180-6.
14. The European Cooperative Study on the clinical significance of right heart thrombi. European Working Group on Echocardiography. Eur Heart J 1989;10:1046-59.