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Cardiac

# Radiopathologic correlation of a tricuspid valve papillary fibroelastoma detected in an infant

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ABSTRACT

Papillary fibroelastomas are benign primary cardiac tumors that usually arise from the valve apparatus and are rare in the pediatric population. Involvement of the tricuspid valve is even less common with only a few cases reported in the literature. Cardiac magnetic resonance imaging is a valuable examination that aids in differentiating a tumor from a thrombus. We present the case of an 11-month-old girl referred by her pediatrician to investigate a murmur noted since birth. To our knowledge, this is the first report of a pathologically proven papillary fibroelastoma arising from the tricuspid valve characterized by magnetic resonance imaging in an infant.



## Introduction

Papillary fibroelastomas are benign primary cardiac tumors that are rare in children and characteristically arise from the valvular apparatus. Few cases of fibroelastomas arising from the tricuspid valve have been reported in the literature. Patients with papillary fibroelastomas are typically asymptomatic but can present with serious complications such as pulmonary or systemic embolization, depending on their location. These tumors are usually detected by 2D (transthoracic) echocardiography. When the diagnosis is in question, cardiac magnetic resonance imaging (MRI) is obtained to help discern between a tumor and a thrombus.

## Case report

An 11-month-old girl with a murmur noted since birth was referred to cardiology by her pediatrician. The patient was born from an in-vitro fertilization with a donor egg via C-section at 37 weeks due to placental abnormalities. The patient had been asymptomatic and achieving appropriate

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developmental milestones. Family history was negative for congenital heart disease, seizures, or genetic disorders. The patient's physical examination revealed a grade III-VI unusually low-pitched continuous murmur heard best along the left lower sternal border but was otherwise normal. The initial workup included an electrocardiogram that showed normal sinus rhythm with a nonspecific intraventricular conduction delay and a transthoracic echocardiogram (TTE) that demonstrated a large mass within the right ventricle, in the vicinity of the basal interventricular septum. Given that the TTE had not been able to demonstrate other mass characteristics, a cardiac MRI was ordered for further investigation, which showed a mobile 15-mm oval mass within the right ventricle, in close proximity to the septal leaflet of the tricuspid valve. No valve inflow or right ventricular outflow tract obstruction was appreciated. The mass was faintly visualized on the cine gradient echo sequences (balanced turbo field echo or steady-state free precession [SSFP]) and showed isointense signal on T1-weighted sequence. The lesion had a targetoid appearance on T2-weighted sequences, with central hyperintense and peripheral hypointense signal, and also demonstrated peripheral enhancement on postcontrast images. Based on the MRI features of the mass, a papillary fibroelastoma arising from the tricuspid valve was suggested. After a multidisciplinary discussion, the cardiothoracic surgeon decided to proceed with resection. A preoperative Holter monitor did not show an arrhythmia and a contrast-enhanced computed tomography of the chest did not reveal pulmonary embolism. During surgery, the mass was attached to the septal leaflet and the chordae of the tricuspid valve, and was visually compatible with a fibroelastoma. After the mass was resected, an off-bypass intraoperative transesophageal echocardiogram revealed moderate tricuspid regurgitation that needed on-bypass annuloplasty. A subsequent repeat off-bypass intraoperative transesophageal echocardiogram showed only residual upper mild regurgitation. A postoperative TTE showed mild-to-moderate tricuspid regurgitation from poor coaptation of the leaflets and normal right ventricular function. There were no surgical complications. The patient was discharged on postoperative day 6 and has been doing well after surgery. The pathologic diagnosis was consistent with a papillary fibroelastoma.

## Discussion

The incidence of primary cardiac tumors in childhood is 0.3%, and among these, papillary fibroelastoma is the least frequently encountered, with only sporadic cases reported [1,2]. Although the pathogenesis of these masses has not been completely understood, some have proposed mechanical endothelial damage, atypical endocardial response to infection or trauma, organized embolization, and hamartomatous-congenital origin as plausible causes [2–4].

Most papillary fibroelastomas present as a solitary mass located along the surface of the valvar leaflets with half, a third, one-sixth, and one-tenth of the cases occurring in the aortic, mitral, tricuspid, and pulmonary valves, respectively [1–4]. Some cases of papillary fibroelastoma arising from nonvalvar endocardial surfaces, predominately in the left ventricle, have also been reported [3,4].

Grossly, papillary fibroelastomas resemble a sea anemone with numerous large papillary fronds emanating from a stalk [2,4]. The majority measure 0.1-3.0 cm but can measure up to 5 cm [4]. Microscopically, its papillary fronds consist of avascular collagenous connective tissue with myxoid change lined by endothelium [1,2] (Fig. 1A and B).

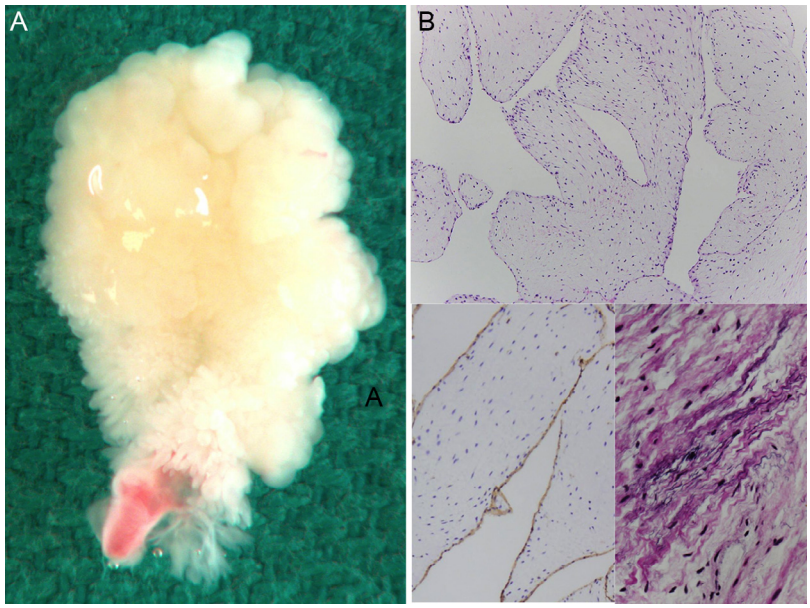


**Fig. 1 – (A) Gross examination of the tissue revealed a myxoid appearance with papillary fronds, measuring 3.0 cm in the largest dimension. (B) Microscopic examination demonstrated paucicellular papillary fronds. The fronds were surfaced by CD31-positive endothelium, but no internal vessels were seen. Focal elastic fibers were noted within the fronds by Verhoeff-van Gieson staining, consistent with a papillary fibroelastoma.**

Generally, papillary fibroelastomas are asymptomatic and found incidentally during evaluations for other cardiac symptomatology or at necropsy [1,3,4]. When papillary fibroelastomas are symptomatic, they classically present with embolism, either systemic—mostly cerebral or coronary—or pulmonary, due to the embolization of its fragile papillary fronds or the aggregation of platelets and fibrin [1,4]. Other reported symptoms include valvar dysfunction, cardiac failure, myocardial infarction, thrombocytopenia, pulmonary hemorrhage, and sudden death [3,4]. Additional diagnostic possibilities that should be considered in these cases are myxoma, lipoma, Lambl excrescences, thrombus, and vegetations, among others [1,2,4]. Although rhabdomyomas and fibromas are the first and second most common neoplasms in children, respectively, these classically arise within the myocardium and not from the valve apparatus [5].

Although 2D echocardiography has been widely used as the traditional modality in the assessment of patients with cardiac masses, cardiac MRI has proven to better characterize and differentiate between a wide range of tumors and thrombi [2,5]. Excluding the possibility of thrombus not only narrows the differential to intracardiac masses but also directs the following steps for management [2,5]. Moreover, a comprehensive cardiac MRI assessment with gradient echo (SSFP), T1-weighted with and without fat saturation, T2-weighted, first-pass perfusion, and delayed myocardial post–contrast enhanced sequences, may help predict the tissue diagnosis in pediatric patients presenting for the evaluation of an intracardiac mass [5]. In our case, TTE failed to demonstrate the close relationship of the mass to the septal leaflet of the tricuspid valve, a characteristic that was critical in our final assessment to favor papillary fibroelastoma over other masses.

The typical MRI features of papillary fibroelastoma are homogeneous signal on all sequences with hypo- to isointense signal on T1, as well as hyperintense and less commonly hypointense signal on T2, relative to the myocardium [5–8]. On traditional cine SSFP (turbo field echo [TFE]) sequences, papillary fibroelastoma is usually isointense relative to the myocardium and presents as a mobile and pedunculated mass arising from a valve leaflet or the endocardial surface [5,8]. Although homogeneous delayed enhancement has been broadly described in the literature, slow and progressive centripetal filling on dynamic postcontrast images has also been demonstrated [6–8]. Notably, papillary fibroelastoma can be difficult to appreciate on certain sequences such as T1 black-blood and cine gradient echo images [6]. Furthermore, first-pass perfusion and fat saturation do not reveal any signal abnormalities [7]. Our case demonstrated a mobile mass within the right ventricle attached to the septal leaflet of the tricuspid valve, which was faintly visualized on the gradient echo sequences. The mass exhibited isointense T1 signal, central hyperintense and peripheral hypointense signal on T2-weighted sequences, and peripheral enhancement on postcontrast images. As previously reported, these imaging characteristics correlate with the pathologic microstructure of the lesion with high-T2 signal representing myxoid elements and gradual centripetal enhancement reflecting the avascular fibroelastic components (Fig. 2A-E).

Other entities such as thrombus, myxoma, and valvular vegetations have different MRI features. For instance, thrombus shows low signal on both T1 and T2, whereas myxomas demonstrate hypo- to isointense signal on T1 and high signal on T2 [8]. Moreover, myxomas characteristically exhibit heterogeneous signal on T1, T2, and dynamic postcontrast images due

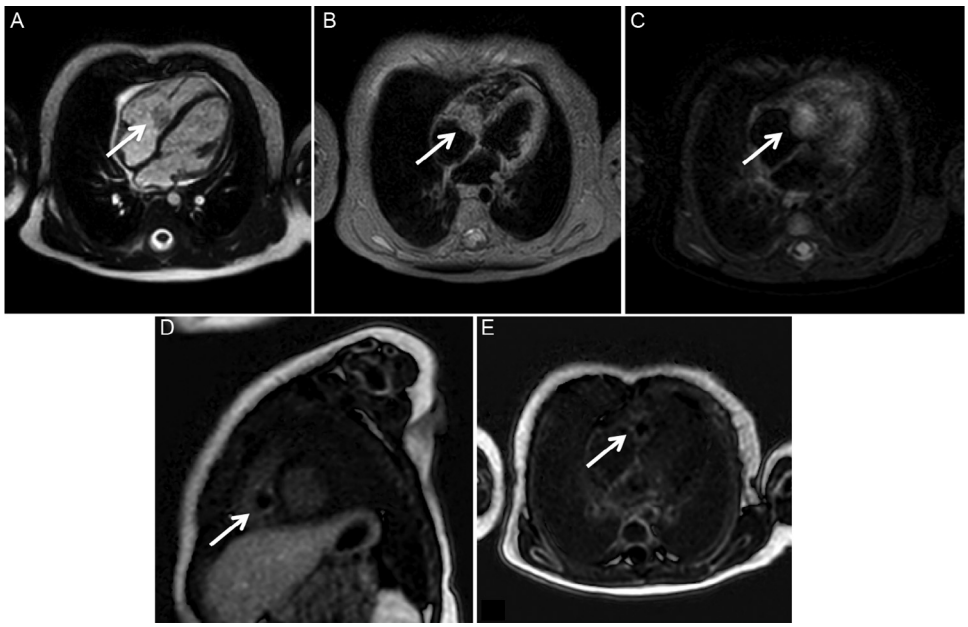


**Fig. 2 – (A) Four–chamber view cine gradient echo sequence shows a subtle, slightly hypointense mobile mass (arrow) within the right ventricle, attached to the septal leaflet of the tricuspid valve. (B) T1-weighted sequence with fat saturation in the 4-chamber geometry demonstrates the tumor is isointense to the myocardium. (C) On short TI inversion recovery sequence, also in the 4-chamber geometry, the mass exhibits a central high signal and a peripheral low signal. (D and E) Postcontrast sequences in the short-axis (D) and 4-chamber (E) views reveal a peripheral enhancement of the tumor.**

to the mixture of its high water content and fibrous components [8]. Although our lesion showed somewhat heterogeneous signal on T2-weighted sequences, the degree of heterogeneity was not diffuse, as it has been typically described in cases of myxomas [8]. Finally, valvular vegetations can be difficult to identify on T1 and T2 images, but can be apparent with low signal on TFE-SSFP images [8]. Overall, the location and the signal features of our patient's lesion revealed by cardiac magnetic resonance, in conjunction with her age, allowed us to suggest the diagnosis of papillary fibroelastoma over other possibilities such as myxoma.

The recommended treatment for papillary fibroelastomas is surgical resection, particularly if the mass is located along the mitral valve apparatus due to a potential risk of cardiac and neurologic complications [4]. Although surgery has been reported to be simple and safe overall, the treatment for asymptomatic papillary fibroelastomas in the right cardiac chambers remains a topic of controversy [4]. Despite the lack of published data documenting the recurrence of these masses, a close follow-up after resection is also recommended [4].

Although papillary fibroelastoma of the tricuspid valve is a rare entity in children, it can pose a diagnostic dilemma on 2D echocardiography. Our case illustrates that cardiac MRI with postcontrast sequences can serve as a complimentary tool to TTE when faced with the challenge of differentiating among a variety of masses and thrombus. Tissue characterization provided by cardiac MRI is important because it can help guide the management algorithm to surgical resection, especially in cases of papillary fibroelastomas, as they can potentially obstruct the atrioventricular valve inflow and occasionally embolize.

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