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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.

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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).

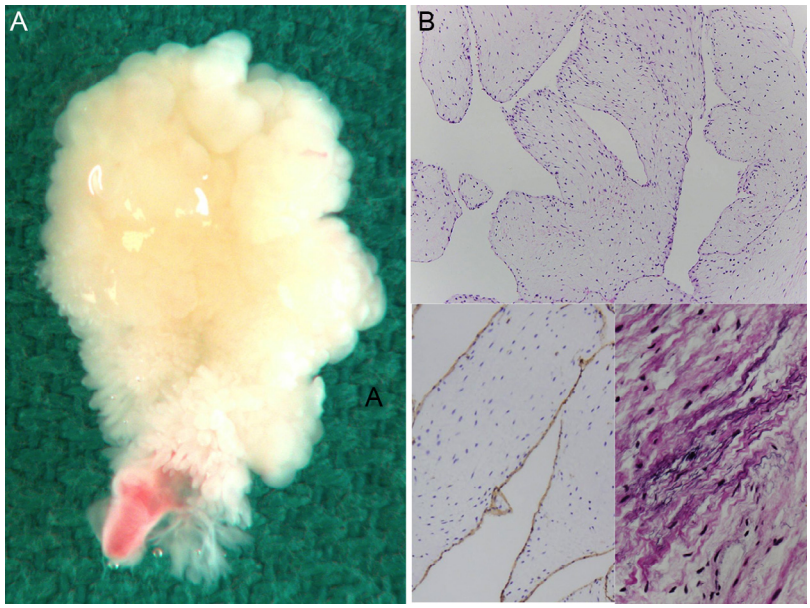

Fig. 1 – (A) Gross examination of the tissue revealed a myxoid appearance with papillary fronds, measuring 3.0 cm in the largest dimension. (B) Microscopic examination demonstrated paucicellular papillary fronds. The fronds were surfaced by CD31-positive endothelium, but no internal vessels were seen.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

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