

Mature Teratoma in the Right Ventricle



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We present an exceedingly rare case of right ventricular outflow tract obstructing mass in an adult patient who presented with dyspnea and dizziness. Transthoracic echocardiography and computed tomography angiogram of the chest showed a large mass in the right ventricle obstructing the outflow tract. A dual right ventriculotomy and right atriotomy surgical approach was taken to completely resect the mass from the interventricular septum, which subsequently confirmed histopathologically the mass as a mature cystic teratoma.

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Tumors of the heart can be classified as primary benign, primary malignant, and secondary metastatic tumors. Primary cardiac tumors are said to be 30 times less common than secondary tumors in an adult patients.¹ More than 58% of primary benign tumors of the heart are myxoma, and the fact that more than 50% of these adult tumors occur in the left atria makes ventricular tumors more rare. The next most common benign intracardiac tumor is fibroelastoma, affecting mainly the aortic valve.¹

Teratoma is a tumor of embryonic germ cells composed of various proportions of endodermal, mesodermal, and ectodermal origin cells. A mature teratoma is defined as when more than 50% of its tissue composition is well-differentiated tissue.² The most common site for teratoma is the gonads followed by mediastinum. Adult intracardiac teratoma is exceedingly uncommon. When it occurs most cardiac teratoma is intrapericardial tumor. Histologically mature teratoma is a benign tumor with a rare chance of malignant transformation. However intracardiac tumors are malignant clinically by virtue of their location, causing obstruction of blood flow, conduction block, or sudden cardiac death.²

There are a handful of case reports of pediatric patients younger than 8 years of age with primary benign teratoma growing inside the heart. However we found

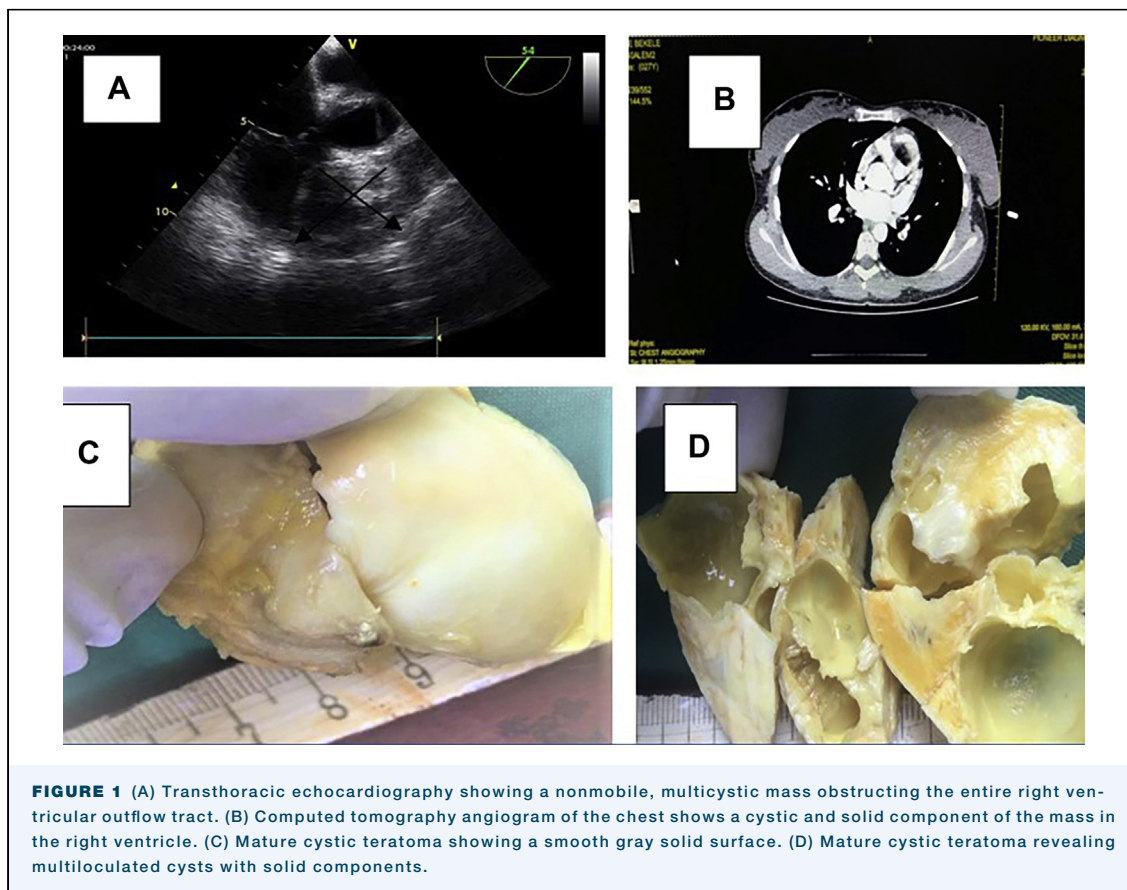
only 1 case report of intracardiac benign cystic teratoma in a 30-year-old African American man in the English literature, which was reported by Mori and colleagues in 2015.³ Here we report another case of benign mature cystic teratoma from the interventricular septum (IVS) of the right ventricle, which we handled with a different surgical technique from the first case report.

A 27-year-old woman presented with a complaint of dyspnea and dizziness for the last 4 years. The patient had an infant aged 1 year, 4 months, and the delivery at the time was uneventful. Dyspnea and discomfort increased after delivery. On physical examination there was a grade IV ejection systolic murmur best heard at the left lower parasternal area with a wide fixed splitting of the second heart sound. One year after the infant was born the patient underwent an echocardiography and was told to have a mass inside the right heart.

The patient had complete right bundle branch block on the electrocardiogram. Both transthoracic and transesophageal echocardiography showed a multicystic heterogeneous mass arising from the IVS measuring 40 mm × 33 mm. The right ventricular outflow tract (RVOT) gradient was 35 mm Hg. The computed tomography angiogram of the chest found a calcific enhancing mass measuring 5.8 × 4.2 cm arising from IVS of the right ventricle causing significant RVOT obstruction (Figure 1).

Surgery was decided, a midline sternotomy was done, and selective aortic and bicaval cannulation were established. The aorta was cross-clamped, and St Thomas cold blood cardioplegia was delivered through the aortic root. The heart was arrested, and the right atrium was opened parallel to the atrioventricular groove after snaring the superior and inferior vena cava. A vent was put in the left atrium through the right superior pulmonary vein. The exposure of the mass through the tricuspid valve was limited, and a vertical 4-cm ventriculotomy over the RVOT below the pulmonary valve into the right ventricle was performed with a safe margin to the right coronary artery branches. A wide-base, attached, smooth surface, well-circumscribed cystic and hard heterogeneous mass was embedded in the endocardium of the IVS, extending from the attachment of the chordae septal and anterior leaflets to most of the RVOT. Complete excision of the mass measuring 6 × 4.5 cm was achieved with cross-clamp time of 74 minutes and total bypass time of 133 minutes.

Using hard Teflon buttressing the margins of the ventriculotomy, the first interrupted 4:0 Prolene suture was followed by a continuous suture used to achieve hemostatic closure of the right ventricle. The heart had a normal sinus rhythm without conduction



block except for the preoperative right bundle branch block. Transesophageal echocardiography showed no ventricular septal defect, no tricuspid regurgitation, and no turbulence or regurgitation across the pulmonary valve. The patient's postoperative course was uneventful, and she discharged from the hospital after 5 days.

Pathologic evaluation of the gross mass revealed a well-circumscribed, yellowish-gray, smooth external surface, solid mass measuring $6 \times 4 \times 4$ cm. A cut section disclosed variably sized, multiloculated cysts containing mucus and a greasy material with a solid and bony hard area. Formalin-fixed, paraffin-embedded, hematoxylin and eosin-stained sections disclosed well-differentiated skin epithelium with adnexal structures, mucous gland, fat tissues, fibrous connective tissues, respiratory epithelium, and lamellar cortical bone. The overall microscopic appearances were in keeping with a mature cystic teratoma (Figure 2).

COMMENT

The only case report of an adult intracardiac benign teratoma was in a 30-year-old African American patient reported in 2015 by Mori and colleagues.³ In this case

report the tumor arose from the free wall of the right ventricle, and surgical excision was possible through the right atriotomy and tricuspid valve. In our case report however the mass was a wide-base attachment mass to the IVS, and mobilization of the mass was possible only through a right ventriculotomy. Of the 2 adult primary malignant intracardiac teratomas in the English literature, 1 was a case report of adult intraventricular malignant teratoma in 1973 by Cabanas and Moore,⁴ which was successfully addressed surgically, and the other was an adult malignant intracardiac teratoma reported in 1946 arising from the interatrial septum after postmortem examination.⁵

In general echocardiography has an excellent sensitivity to intracardiac masses arising from the endocardium because of the sharp echo contrast between the mass and the echo-lucent chambers.⁶ However computed tomography of the chest is superior to echocardiography in the sense that it can detect tissue calcification within the intracardiac mass and better shows involvement of myocardium or pericardium. The unique utility of magnetic resonance imaging is its precision during equivocal intracardiac mass diagnosis by echocardiography. In addition magnetic resonance imaging permits the accurate

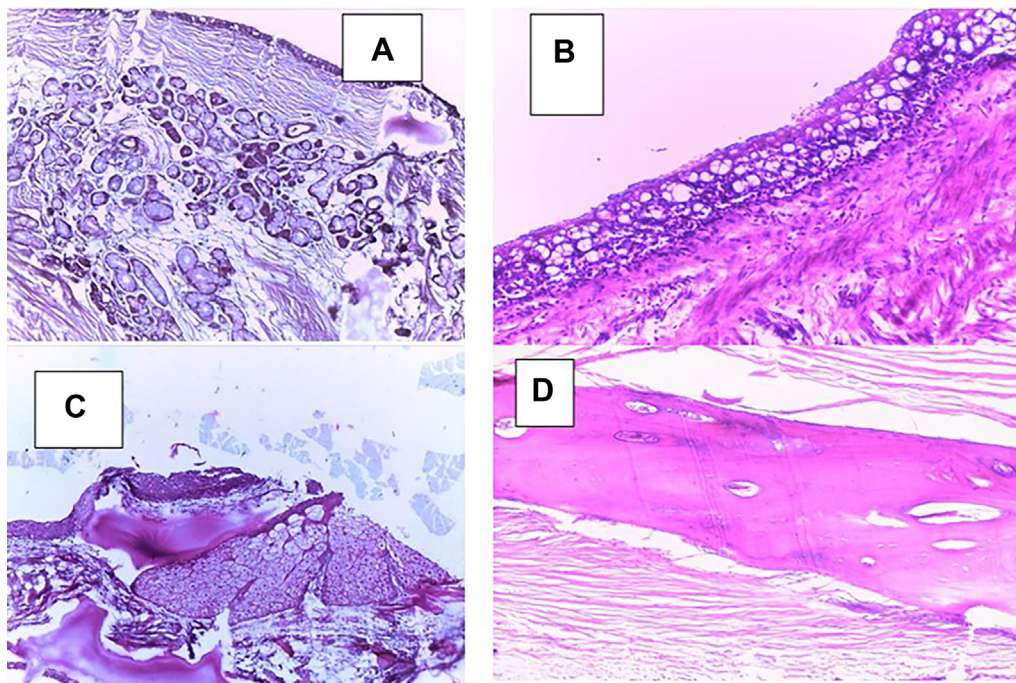


FIGURE 2 (A) Mucous glands within fibrous fatty stroma and respiratory epithelium. (B) Ciliated respiratory lung epithelium (hematoxylin and eosin, 200×). (C) Mature squamous epithelium with skin appendages. (D) Plate of lamellar cortical bone (hematoxylin and eosin, 200×).

diagnosis and description of primary and secondary intracardiac malignancies.⁶

Mature cystic teratoma is not sensitive to chemotherapy or radiation. Complete surgical excision is curative. When complete excision without affecting the vital structures is not possible, partial resection for unobstructed blood flow is an option.⁷ The right atriotomy approach through the tricuspid valve may provide only partial exposure to the

mass, and complete excision for a wide-based, attached mass on the IVS obstructing RVOT may be difficult.

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