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Case Report

Right coronary artery fistula with giant pseudoaneurysm presenting as intrapericardial mass

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ABSTRACT

Coronary artery fistulas are rare congenital anomalies usually discovered incidentally on imaging studies. Coronary artery pseudoaneurysms are unusual complications of coronary artery fistulas, which can be due to atherosclerosis, inflammatory, traumatic or iatrogenic causes. We present a case of a 55 year old female with no known atherosclerotic risk factors, history of trauma or connective tissue disease referred because of recurrent palpitations. Work ups revealed a cardiac mass with an initial assessment of pericardial cyst. A multi modality approach of two dimensional echocardiography with Doppler studies, multidetector computed tomography and coronary angiogram revealed coronary artery fistula draining into a pericardial mass. The patient underwent surgical excision of the mass and ligation of the feeding vessel. Histopathology revealed features suggestive of a pseudoaneurysm. Postoperative course was uneventful and she was discharged stable and improved. Coronary artery fistula complicated by pseudoaneurysm is a rare clinical entity especially in patients without history of trauma or other risk factors. It can have an unusual presentation which can confound the diagnosis. Multimodality imaging is essential and adjunctive in order to determine a conclusive assessment.

< **Learning objective:** Coronary artery pseudoaneurysm secondary to a congenital coronary artery fistula is an unusual cardiovascular pathology and can present as a rare diagnostic challenge for the clinician. This case emphasizes the importance of meticulous integration of both clinical assessment and complementary multimodality imaging approaches to better define the best therapeutic plan and facilitation of definitive surgical management.>

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Introduction

A coronary artery fistula is a rare defect that involves a fistulous communication with either a cardiac chamber or another vascular segment of the pulmonary or systemic circulation. They can either be congenital or acquired and can be asymptomatic or present with varying symptomatology. Coronary artery aneurysms are rare complications associated with a fistula can result from either atherosclerotic, inflammatory or traumatic causes and are rarer than coronary artery fistulas [1]. Pseudoaneurysms are characterized by a contained rupture of the myocardium or a blood vessel with blood

flow into a cavity contained by pericardium, thrombus or adhesions. Most reported cases of coronary artery pseudoaneurysms are due to traumatic or post procedural complications [2]. Due to its rarity, the incidence of non traumatic coronary artery pseudoaneurysm secondary to a coronary artery fistula is unknown.

Case report

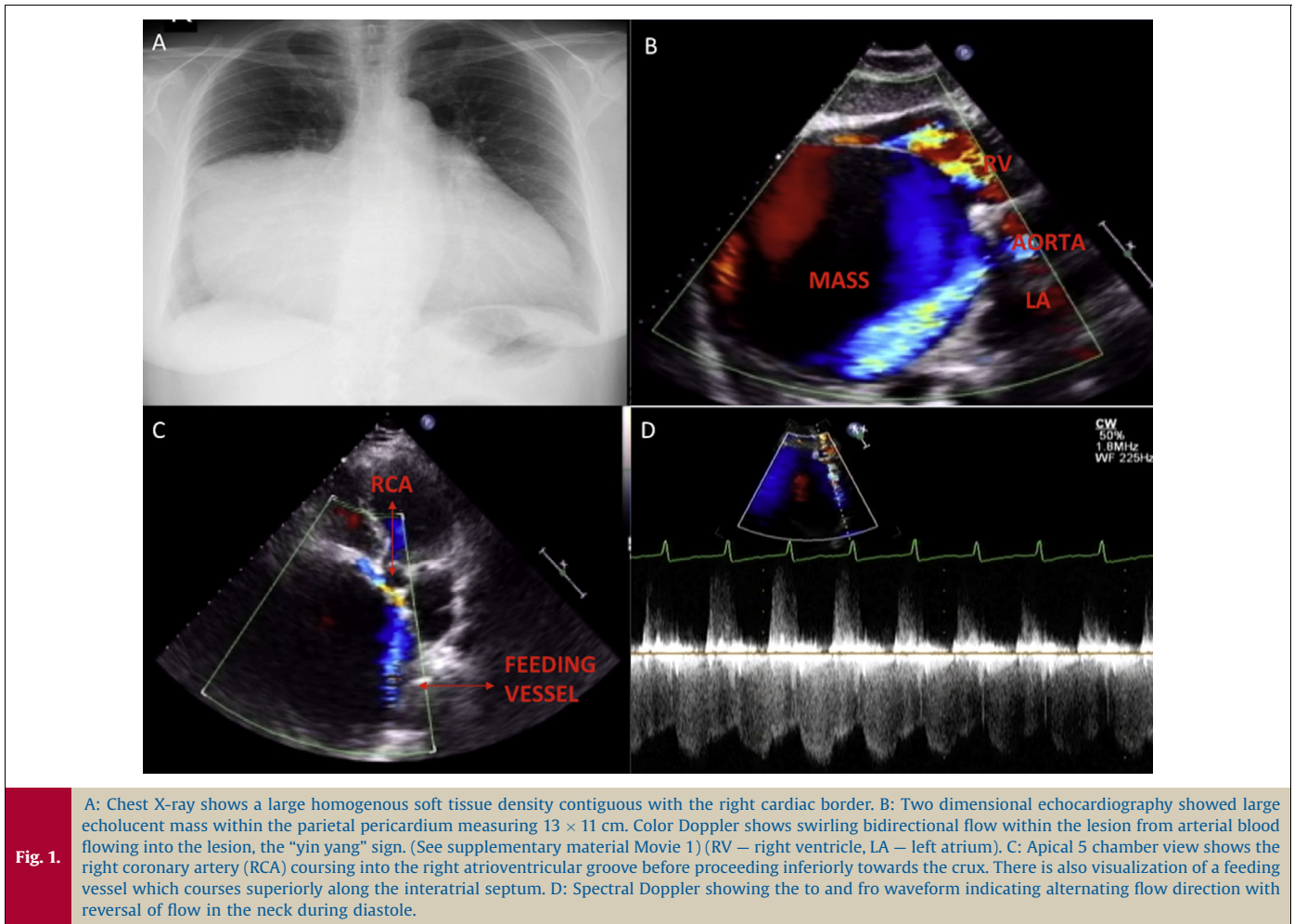
A 55 year old female with no cardiac risk factors has been having recurrent palpitations that would resolve spontaneously for the past 3 years. On initial check up it was noted that she had atrial flutter that would spontaneously convert to sinus rhythm. She was referred to a cardiologist where work ups revealed the presence of a cardiac mass. The initial consideration was that of a pericardial cyst. She was then referred to our institution for further evaluation. Her past history and physical examination was unremarkable.

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Chest radiograph showed a large rounded homogenous right sided soft tissue density with a partially calcified wall (Fig. 1A). Two dimensional transthoracic echocardiogram showed normal ejection fraction with no regional wall motion abnormalities. There was an echolucent mass anterior to the right atrium and intrapericardially located. Blood flow within the mass reveals a swirling “yin yang” blood flow pattern (Fig. 1B, supplementary material Movie 1). Further interrogation shows a dilated right coronary artery with a feeding vessel coursing into the mass (Fig. 1C). Doppler analysis reveals a “to and fro” spectral waveform pattern (Fig. 1D).

Coronary CT angiogram shows a large intrapericardial mass which is probably vascular in origin while compressing surrounding structures and measures 12 × 12 × 13 cm (Fig. 2A). The branch of the dilated right coronary artery directly appears to be draining into the mass (Fig. 2B and C). The patient was then submitted for coronary angiography which showed a normal left coronary system. The right coronary system divides into 2 branches. The first branch traverses the usual course of the right coronary artery while the other branch appears to be an anomalous dilated vessel draining into a huge sized chamber (Fig. 2D).

Patient underwent sternotomy excision of the mass with ligation of the feeding vessel under cardiopulmonary bypass. Intraoperative findings showed a 12 cm blood filled intrapericardial mass located lateral to the right ventricle and insinuates between the superior vena cava and the pulmonary veins. There was note of a vessel arising from the proximal portion of the ascending aorta which fills the mass and a draining sinus into the

inferior vena cava (Fig. 3A). The mass was opened and blood was drained via cardiotomy suction (Fig. 3B). Staining was positive for elastic fibers with replacement by fibromyxoid areas (Fig. 3C). Additional stains highlighted the vasa vasora. With clinico pathologic correlation, the pathology report was signed out as fibrocollagenous tissues suggestive of elastic artery with extensive fibromyxoid change and calcification probably pseudoaneurysm with no evidence of malignancy or infection.

Her postoperative course was unremarkable. She had stable vital signs and was in sinus rhythm with no recurrence of arrhythmia. Repeat transthoracic echo showed normal wall motion and contractility with absence of the previously noted intrapericardial mass. She was eventually discharged and improved on follow up.

Discussion

Coronary artery fistulas are rare congenital anomalies present in 0.002% of the general population and are discovered incidentally in 0.25% of patients undergoing catheterization procedures. The right coronary artery or its branches is the most common site of origin in 55% while the left coronary system is involved in 35% of cases [3]. With regards to the site of drainage over 90% of fistulas drain into venous structures including right-sided chambers, pulmonary artery, coronary sinus, and superior vena cava but drainage into the left-sided chambers is less frequent. Fistulous drainage occurs into the right ventricle in 40%, right atrium in 26%, pulmonary artery in 17%, left ventricle in 3%, coronary sinus in 7%,

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