

Continuous Heart Murmur-Two Cases of Rare Causes and Role of Imaging in Diagnosis

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Abstract

A continuous murmur is defined as a murmur that begins in systole, continues through the second heart sound and into part or all of diastole. Among a number of differential diagnoses, anomalous systemic arterial supply to the normal basal segments and Rupture of Sinus of Valsalva (RSOV) are two rare causes of continuous heart. These two entities, which are often overlooked clinically, may have potentially fatal complications. We herein present two cases of uncommon causes of continuous heart murmur; demonstrate the radiological features and corroborate the role of Electric Cardiogram-Gated Computed Tomography (ECG-gated CT) in diagnosis.

Introduction

Continuous heart murmurs are murmurs those involve the whole cardiac cycle, begin in systole, continue through the second heart sound and into part or all of diastole. There are a number of differential diagnoses for continuous heart murmur: Patent Ductus Arteriosus (PDA), cervical venous hum, hyperdynamic circulation of pregnancy, Arterio-Venous (AV) fistulas of coronary arteries, great vessel or haemodialysis etc. Nowadays, ECG-gated CT provides excellent anatomic depiction and also simultaneous evaluation of coronary arteries, great vessels and lungs. In this article, we would like to present two cases of uncommon causes of continuous heart murmur; demonstrate the radiological features and corroborate the role of ECG-gated CT in diagnosis.

Case 1

A 30-year-old male non-smoker with good past health was noted to have a Grade 3 continuous heart murmur at the precordium during his pre-employment health check-up in the private sector in May 2011. An approximately 2.0 cm x 4.0 cm shadow was also spotted at left retrocardiac region, with dilated tubular structures, in his pre-employment chest radiograph (CXR) (Figure 1a). Respiratory physician suspected that it was a pulmonary Arterio-Venous Malformation (AVM); and subjected the patient to Magnetic Resonance (MR) study to measure the shunt ratio, as the consideration for embolization

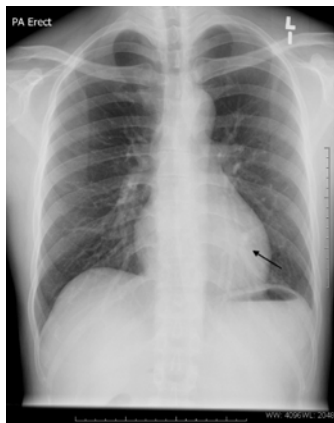


Figure 1a: Chest Radiography (CXR) Posterior-anterior (PA) projection demonstrates an approximately 2.0 cm x 4.0 cm shadow at left retrocardiac region (arrow), with dilated tubular structures, suspicious of feeding arteries and draining veins, adjacent to it.

depended on the degree of Arterio-Venous (AV) shunting. However, MR images were not typical of pulmonary AVM. Magnetic Resonance Angiography (MRA) demonstrated serpiginous vascular tangle at left lower lobe, with systemic arterial blood supply and pulmonary venous drainage (Figure 1b). Cine phase contrast for flow volume showed Qp:Qs=1:1.1, indicative of no significant AV shunting. Magnetic Resonance (MR) pulmonary arteriogram showed attenuation of segmental branches of basal segments of Left Lower Lobe (LLL) (Figure 1c). Bronchopulmonary sequestration was suspected at that moment.

A Computed Tomographic (CT) scan was subsequently performed and demonstrated a ~1.0 cm in caliber anomalous systemic artery from the descending aorta at T8 level to the basal segment of left lower lobe (Figure 1d-1g). Dilated arteries were paralleling the normal bronchi.

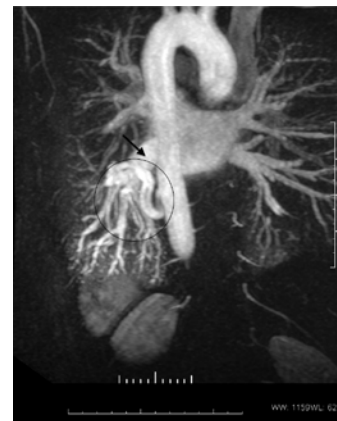


Figure 1b: Magnetic resonance angiography (MRA) demonstrates serpiginous vascular tangle at left lower lobe, with systemic arterial blood supply (circle) and pulmonary venous drainage (arrow).

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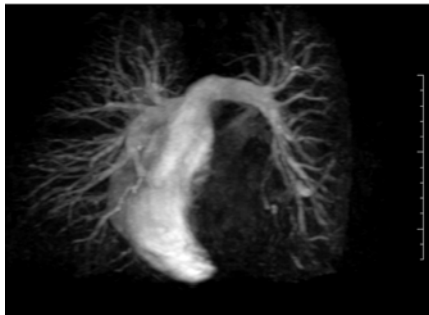


Figure 1c: Magnetic resonance (MR) pulmonary arteriogram shows attenuation of segmental branches of basal segments of left lower lobe (LLL).

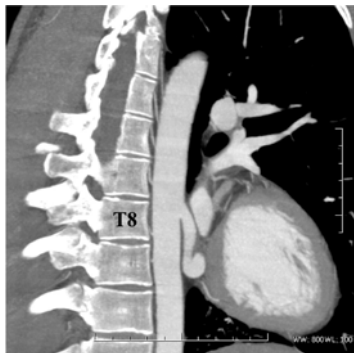


Figure 1d: Electric cardiogram (ECG)-gated computed tomography (CT) (1d,1e) demonstrates an anomalous systemic artery from the descending aorta at T8 level to the basal segment of left lower lobe. Dilated arteries are paralleling the normal bronchi. No direction communication with the pulmonary veins is evident. (1f) Tributaries of the left inferior posterior vein (LIPV) in left lower lobe are hypertrophies, with variceal formation. Branches of left pulmonary artery (LPA) supplying the basal segments, distal to origin of the apical segment branch, are however attenuated. (1g) Underlying lung parenchyma is normal, which makes bronchopulmonary sequestration less likely. Overall findings are in favour of anomalous systemic arterial supply to normal lung parenchyma in the basal segments of left lower lobe.

No direction communication with the pulmonary veins was evident. Tributaries of the Left Inferior Posterior Vein (LIPV) in left lower lobe were hypertrophies with variceal formation. Branches of Left Pulmonary Artery (LPA) supplying the basal segments, distal to origin of the apical segment branch, were however attenuated. Underlying lung parenchyma was normal, which made bronchopulmonary sequestration less likely. Overall findings were in favour of anomalous systemic arterial supply to normal lung parenchyma in the basal segments of left lower lobe, with associated Pulmonary Vein (PV) dilatation and attenuated Pulmonary Artery (PA) branches. Since our patient was asymptomatic, with no significant shunting, he was managed conservatively.

Case 2

A 25-year-old young man, who was a chronic smoker and recreational drug abuser, was incidentally found to have a rough systolic murmur, loudest over the precordium, during a consultation for flu-like symptoms by a general practitioner in September 2011. He was referred to the cardiac clinic for further assessment and investigation. On further enquiry by the cardiologist, he volunteered a 2-month history of exertional dyspnea. Physical examination revealed a mildly engorged jugular vein and mild leg edema. Cardiac auscultation revealed a Grade 3/4 continuous murmur along the right sternal



Figure 1e: Electric cardiogram (ECG)-gated computed tomography (CT) (1d,1e) demonstrates an anomalous systemic artery from the descending aorta at T8 level to the basal segment of left lower lobe. Dilated arteries are paralleling the normal bronchi. No direction communication with the pulmonary veins is evident. (1f) Tributaries of the left inferior posterior vein (LIPV) in left lower lobe are hypertrophies, with variceal formation. Branches of left pulmonary artery (LPA) supplying the basal segments, distal to origin of the apical segment branch, are however attenuated. (1g) Underlying lung parenchyma is normal, which makes bronchopulmonary sequestration less likely. Overall findings are in favour of anomalous systemic arterial supply to normal lung parenchyma in the basal segments of left lower lobe.



Figure 1f: Electric cardiogram (ECG)-gated computed tomography (CT) (1d,1e) demonstrates an anomalous systemic artery from the descending aorta at T8 level to the basal segment of left lower lobe. Dilated arteries are paralleling the normal bronchi. No direction communication with the pulmonary veins is evident. (1f) Tributaries of the left inferior posterior vein (LIPV) in left lower lobe are hypertrophies, with variceal formation. Branches of left pulmonary artery (LPA) supplying the basal segments, distal to origin of the apical segment branch, are however attenuated. (1g) Underlying lung parenchyma is normal, which makes bronchopulmonary sequestration less likely. Overall findings are in favour of anomalous systemic arterial supply to normal lung parenchyma in the basal segments of left lower lobe.

border. Transthoracic Echocardiography (TTE) revealed alternating jet-flows into the Right Atrium (RA) during different phases of the cardiac cycle; and structurally normal valves, with trivial Tricuspid Regurgitation (TR).

Chest Radiographs (CXRs) revealed mild pulmonary congestion with cardiomegaly. (Figure 2a) The patient refused to undergo Transesophageal Echocardiography (TEE) even after a comprehensive description of the procedure was provided. Electric Cardiogram (ECG)-gated Computed Tomography (CT) was performed, and it revealed a tubular jet of contrast from the anterior base of the right sinus of Valsalva, into Right Atrium (RA), with a maximum diameter of ~5.8 mm (Figure 2b-2d). The hole of the right cusp was surgically

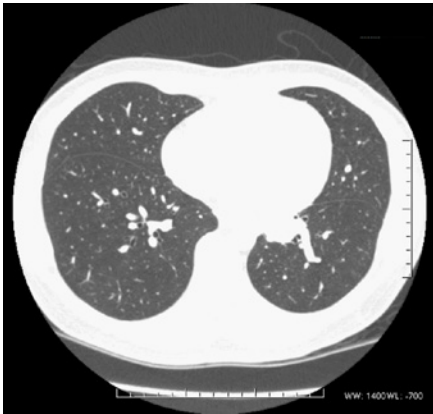


Figure 1g: Electric cardiogram (ECG)-gated computed tomography (CT) (1d,1e) demonstrates an anomalous systemic artery from the descending aorta at T8 level to the basal segment of left lower lobe. Dilated arteries are paralleling the normal bronchi. No direction communication with the pulmonary veins is evident. (1f) Tributaries of the left inferior posterior vein (LIPV) in left lower lobe are hypertrophies, with variceal formation. Branches of left pulmonary artery (LPA) supplying the basal segments, distal to origin of the apical segment branch, are however attenuated. (1g) Underlying lung parenchyma is normal, which makes bronchopulmonary sequestration less likely. Overall findings are in favour of anomalous systemic arterial supply to normal lung parenchyma in the basal segments of left lower lobe.

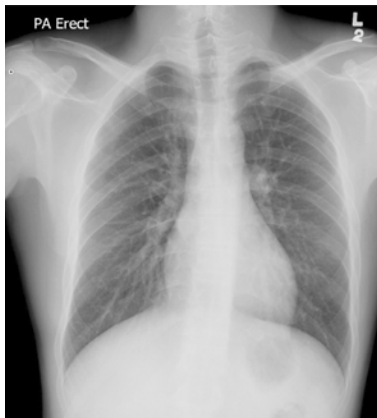


Figure 2a: Chest Radiography (CXR) Posterior-anterior (PA) projection reveals mild cardiomegaly and pulmonary congestion.

repaired. The patient recovered uneventfully, with no long-term cardiac abnormality.

Discussion

Continuous heart murmur is a common clinical finding in asymptomatic patients and patients with various presentations, such as chest discomfort and shortness of breath. It is defined as a murmur that begins in systole, continues through the second heart sound and into part or all of diastole. There are a number of differential diagnoses for continuous heart murmur: Patent Ductus Arteriosus (PDA); cervical venous hum; hyperdynamic circulation of pregnancy; Arterio-Venous (AV) fistulas of coronary arteries; great vessel or haemodialysis; coarctation collaterals, truncus arteriosus; pseudo-truncus; total anomalous pulmonary venous connection; Cruveilhier-Baumgarten Syndrome; and post-operative status such as Blalock-Taussig shunt; Potts operation; and Waterston operation. Anomalous systemic arterial supply to the normal basal segments and rupture of sinus of Valsalva

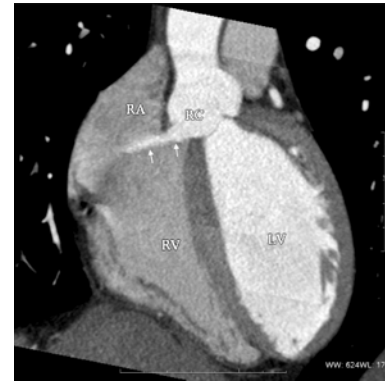


Figure 2b: Electric cardiogram (ECG)-gated computed tomography (CT) was performed, with retrospective reconstruction and reformat of images in oblique coronal views (2a and 2b) and 4-chamber view (2c). A tubular jet of contrast (white arrows) is revealed from the anterior base of the right sinus of Valsalva, into right atrium (RA), with a maximum diameter of ~5.8 mm (2b). (RA=right atrium; RC=right cusp; RV=right ventricle; LA=left atrium; LV=left ventricle).

(RSOV) are two rare and vital causes of continuous heart murmur, as they are potentially fatal.

Radiological features of anomalous systemic arterial supply to the normal basal segments are often confused with pulmonary AVM or bronchopulmonary sequestration. In plain radiographs, the anomalous systemic artery can appear as dilated tubular structures or ill-defined nodular area of increased opacity in retrocardiac region, as in our cases. The anomalous systemic artery can silhouette out the outline of the lower thoracic descending aorta as focal obscuration. Anomalous systemic arteries are also well demonstrated on CT scans as vascular structures, with the same attenuation as the thoracic aorta in the left lower lobe. Moreover, CT can also provide information about the morphology of the bronchial tree and the pulmonary parenchyma, which are both normal in this condition. Another CT finding is the absence or attenuation of the interlobar artery distal to the origin of the superior segmental artery [1,2]. Bronchial tree and the pulmonary parenchyma are normal in this condition. Although MRI can demonstrate an anomalous systemic artery, yet it would not be easy to determine confidently whether the lung tissue supplied by a systemic artery is normal. Therefore, CT is a useful technique for the diagnosis of anomalous systemic arterial supply to the normal basal segments [3]. Patients having anomalous systemic arterial supply to the normal basal segments of left lower lobe are often asymptomatic, while occasional cases of hemoptysis are recognized. Without the presence of clinical symptoms such as hemoptysis and congestive heart failure, treatment is conservative; despite surgery is always imperative for the correction of left-to-right shunt. An anomalous systemic artery can be ligated if there is pulmonary supply to the involved segments of the lung. If, on the other hand, the aberrant artery represents the sole source of blood flow, lobectomy is required. In either case, contrast-enhanced CT is indispensable for correct diagnosis and proper treatment.

In patients suffering from RSOV, CXR may reveal cardiomegaly, right heart enlargement and pulmonary congestion. ECG-gated CT is superb in identifying the location of the rupture hole before surgical evaluation; providing early diagnosis of RSOV; delineating an aorto-cardiac shunt; and evaluating tricuspid valve involvement [4,5]. The high contrast resolution of CT makes it possible to delineate an aorto-cardiac shunt, if present, and to identify a ruptured aneurysm by depicting a jet of contrast material extending from the aneurysm into the adjacent cardiac chamber. The high contrast resolution of CT is also



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invaluable for evaluating Tricuspid Valve (TV) involvement. Tricuspid regurgitation is a serious consideration if there is TV involvement, and may increase the risk of endocarditis and the difficulty of surgical repair. If TV involvement is suspected, images with better quality focus around the TV can be obtained. Although RSOV may have potentially fatal complications, the prognosis is excellent after treatment. Thus, prompt and accurate diagnosis is crucial.

There are many different causes for continuous murmur and some of them may have potentially fatal complications. Anomalous systemic arterial supply to the normal basal segments and Rupture of Sinus of Valsalva (RSOV) are two rare causes of continuous heart murmur. These two entities are important, as they may have potentially fatal complications. Early diagnosis and prompt management are crucial. Nowadays, ECG-gated CT provides excellent anatomic depiction, and simultaneous evaluation of coronary arteries, great vessels and lungs. A combined approach to diagnoses contributed by both cardiologists

and radiologists often reveals the nature of pathology better and aids successful management.

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