

A Mediastinal Mass due to Coronary to Pulmonary Artery Fistula with a Giant Aneurysm Formation

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ABSTRACT. A 64-year-old male with a congenital coronary artery to pulmonary artery fistula was admitted for evaluation of a mediastinal mass. A faint, high-pitched pansystolic murmur best heard in the second left intercostal space was changed by methoxamine into a continuous murmur. The phonocardiogram, echocardiogram and contrast-enhanced CT scan suggested a coronary artery fistula. It was confirmed by a selective coronary angiogram which revealed a giant oval aneurysm, corresponding to the localized bulging of the left cardiac border on X-ray. Surgical correction was successful. A fistulous aneurysm of a coronary artery should be considered in the differential diagnosis of a large mediastinal mass especially in the presence of a regurgitant heart murmur.

Key words: Mediastinal mass — Coronary artery fistula —
Coronary aneurysm — Continuous murmur

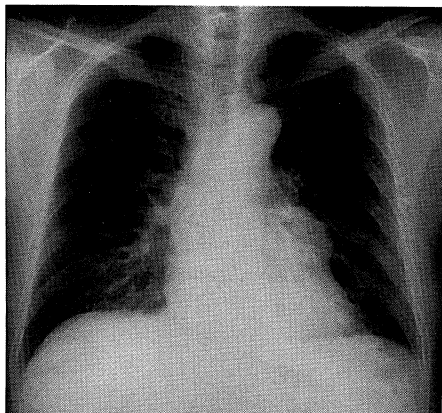
Congenital coronary artery fistulas are the most common type of coronary artery malformations. Although once considered rare, increasing numbers of surgically corrected cases are being reported, most of which were suspected because of a continuous murmur detected at routine examination. A coronary artery aneurysm and fistula on occasion may cause an abnormal cardiac silhouette as demonstrated by our patient with a coronary artery to pulmonary artery fistula associated with a giant aneurysm formation simulating a mediastinal mass.^{1,2)}

We report a case of coronary artery fistula whom phonocardiographic evaluation of the systolic murmur is useful in the differential diagnosis.

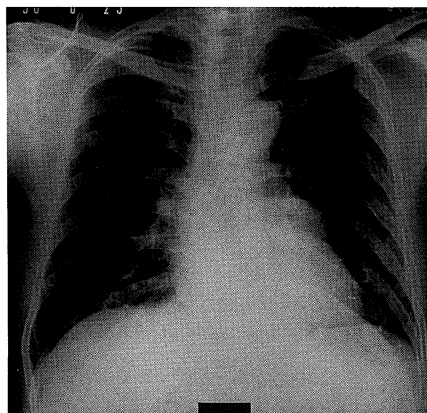
CASE REPORT

A 64-year-old male was admitted for evaluation of a mediastinal mass on chest X-ray which was first noticed 3 months prior to admission at a routine health examination. The patient had been asymptomatic with no previous cardiac history, and the family history was not contributory. For several years he had climbed trees in order to cut off large branches which he held under his

A



B



C

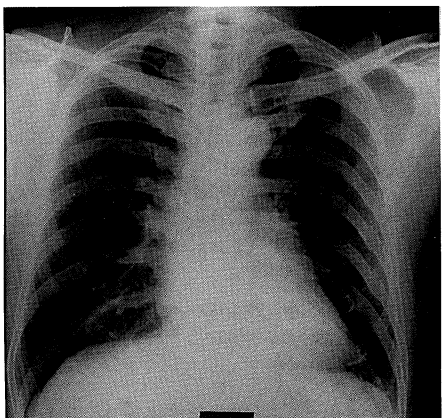


Fig 1. A serial chest X-ray film, showing a localized round bulge on the left cardiac border, suggested a mediastinal mass initially. The abnormal shadow has been gradually enlarging. (A). Chest X-ray film taken on admission. (B). Chest X-ray film taken 3 months before admission. (C). Chest X-ray film taken 6 months before admission.

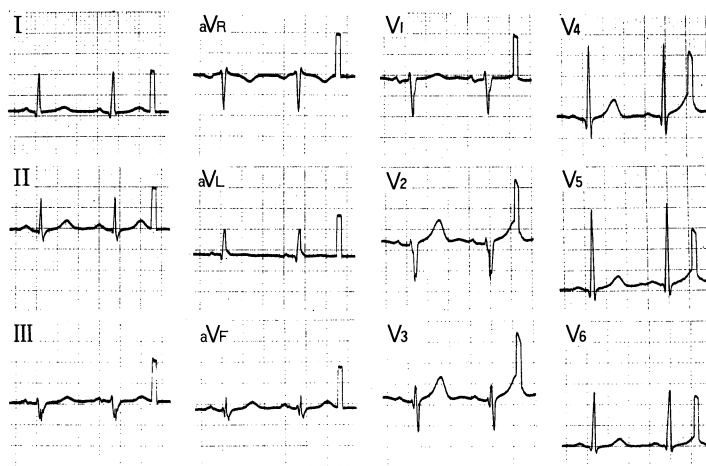
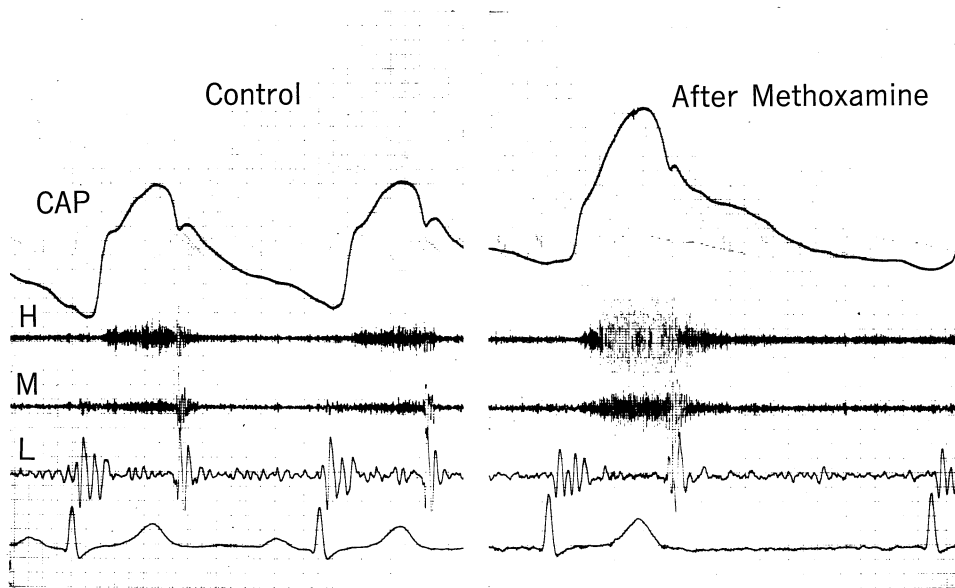


Fig 2. The electrocardiogram, recorded on admission.

A



B

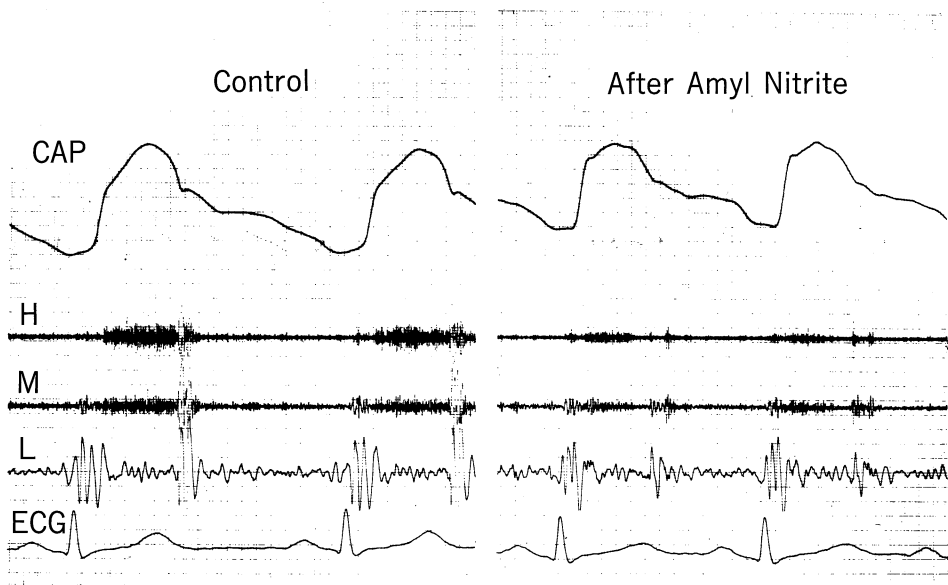


Fig 3. Control phonocardiogram, recorded at the left second intercostal space with a paper speed of 100 mm/sec, demonstrates a high-pitched pansystolic murmur. Intravenous methoxamine accentuated the murmur and made it continuous (A), while amyl nitrite inhalation decreased the loudness of the murmur (B). From top to bottom: CAP = Carotid artery pulse. PCG = phonocardiogram (H: High-frequency, M: Medium-frequency, L: Low-frequency). ECG = Electrocardiogram.

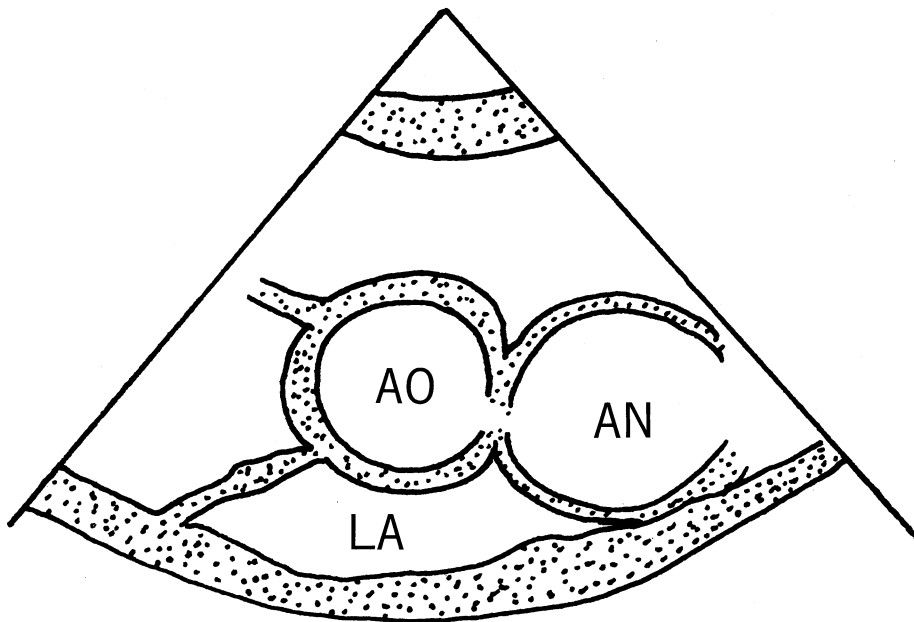
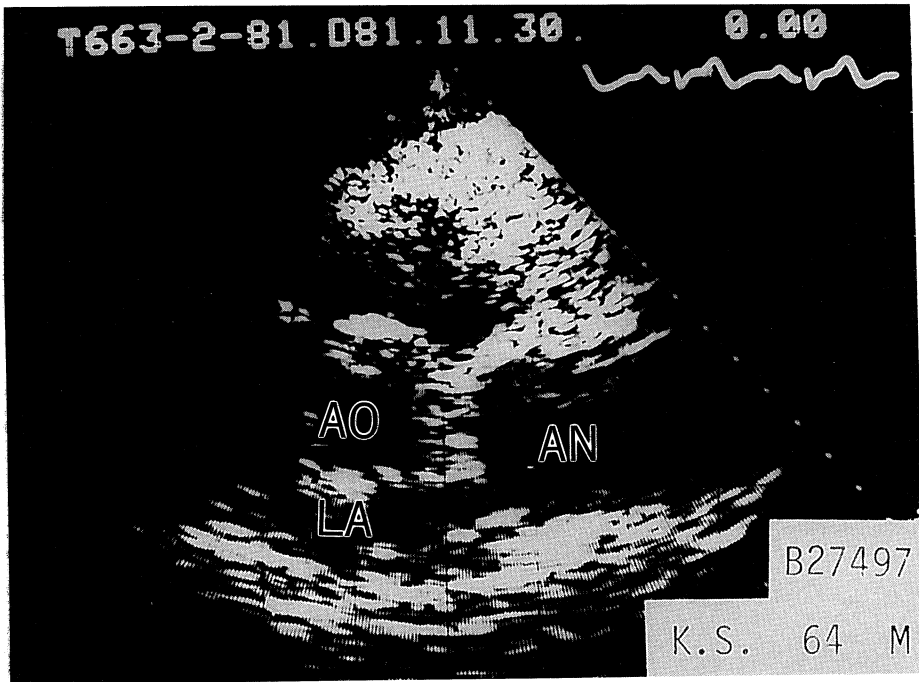


Fig 4. A short axis two-dimensional echocardiogram and corresponding schematic diagram, showing an aneurysmal dilatation at the origin of the left coronary artery.
AO = Aorta LA = Left atrium AN = Aneurysmal dilatation

left arm. The branches would usually strike his left chest forcefully when they were cut off.

On physical examination he appeared to be well-nourished, without cyanosis or clubbings. The peripheral pulses were all palpable. The blood pressure was 126/80 and his heart rate was 60. On auscultation his heart sounds were normal but a faint high-pitched pansystolic murmur was best heard in the second left intercostal space. The remainder of the physical examination was unremarkable.

A chest X-ray on admission showed a localized round bulge on the left cardiac border suggesting a mediastinal mass (Fig 1A). Pulmonary vascular markings were within normal limits. When comparing this film with the ones taken 3 months and 6 months before admission, the abnormal shadow had been gradually enlarging (Fig 1B and 1C). The electrocardiogram showed a poor R wave progression between V1 and V2 (Fig 2). A double Master two-step exercise test was negative. A phonocardiogram confirmed the auscultatory findings (Fig 3). Intravenous methoxamine accentuated the murmur and made it continuous (Fig 3A), while amyl nitrite inhalation decreased the loudness of the murmur (Fig 3B). These findings suggested the presence of a left-to-right shunt such as an arterio-venous communication, either in the heart or great vessels.

A short axis two-dimensional echocardiogram disclosed an aneurysmal dilatation at the origin of the left coronary artery (Fig 4). A CT scan revealed a 4×5 cm oval mass on the left posterior aspect of the pulmonary trunk. This lesion was enhanced by an intravenous injection of contrast medium and the

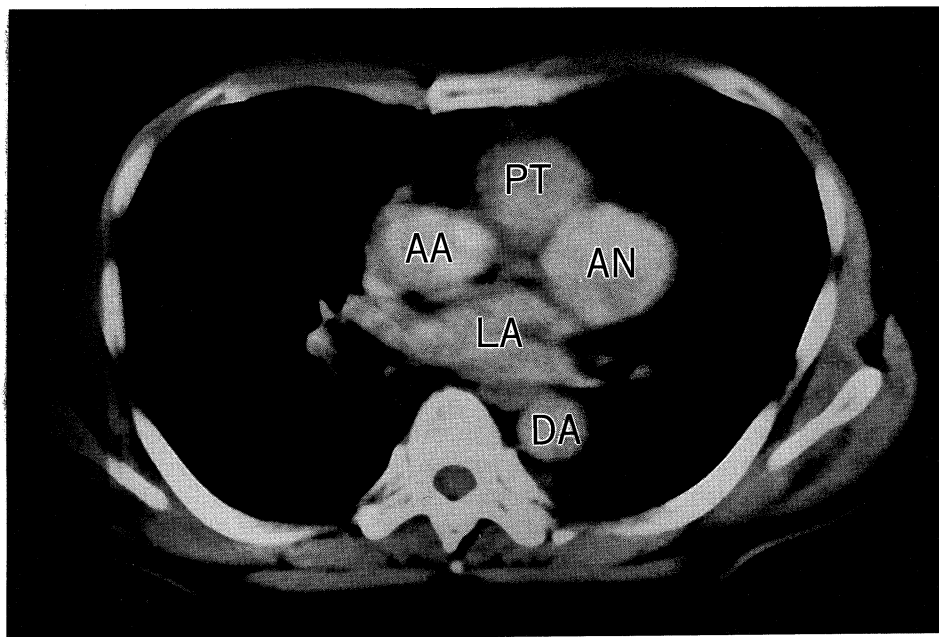


Fig 5. Contrast-enhanced CT scanning, demonstrating a 4×5 cm aneurysm (arrow) on the left posterior aspect of the pulmonary trunk.

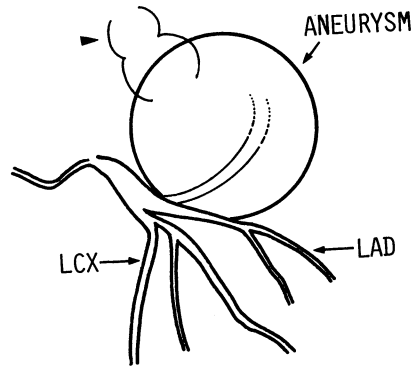
AA = Ascending aorta

PT = Pulmonary trunk

LA = Left atrium

DA = Descending aorta

A



B

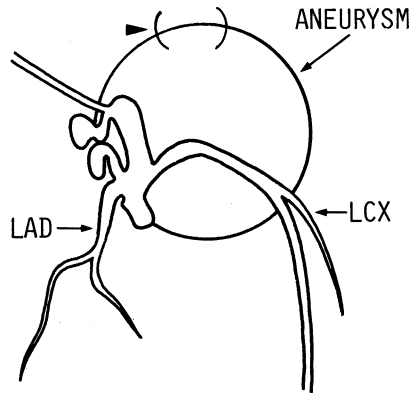
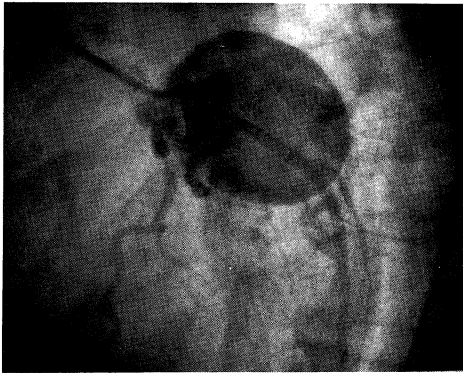


Fig 6. Selective coronary angiogram, showing the left anterior descending artery-to-pulmonary artery fistula with a giant oval aneurysm near its origin. Also note the left anterior descending artery with poor distal filling.

(A) Right anterior oblique projection
 (B) Left anterior oblique projection
 LAD = Left anterior descending artery
 LCX = Left circumflex branch
 Arrow head = fistulous vessel

findings confirmed the abnormal X-ray shadow as cardiovascular in origin (Fig 5). Cardiac catheterization data were essentially normal (Tab 1), but a selective coronary angiogram confirmed the left anterior descending artery-to-pulmonary artery fistula with a giant oval aneurysm near its origin (Fig 6). There was a poor filling of the left anterior descending artery distal to the fistula suggesting a coronary steal phenomenon and a lack of myocardial perfusion possibly accounting for the abnormal electrocardiographic precordial lead pattern and also for hypokinesis of the anterior wall on the left ventriculogram. The large aneurysm of the coronary artery fistula corresponded to the bulging of the left cardiac border seen on the chest X-ray (Fig 1). An urgent surgery was performed, since the aneurysm continued to enlarge and there was a risk of rupture. At operation a tortuous vessel arose from a branch of the left anterior descending artery which communicated with the pulmonary trunk. Proximally,

TABLE 1. Cardiac Catheterization

Position	Pressure (mean) mmHg	O ₂ Saturation (%)
PA wedge		92
right PA	20/10	70
left PA	24/12	68
main PA	20/ 8	71
RV outflow	24/ 5	68
RV inflow	24/ 4	68
RA high		8
RA middle		10
RA low		7
SVC		8
IVC		8
Femoral artery	140/80	90
Cardiac index	3.4L/min/m ²	

PA=pulmonary artery RV=right ventricle RA=right atrium
SVC=superior vena cava IVC=inferior vena cava

there was a grossly dilated aneurysm (4×5 cm) located over the anterior portion of the left atrial appendage, and densely adherent to the surrounding tissue. The aneurysm was resected under cardio-pulmonary bypass and it was found to contain an old thrombus. The terminal end of the fistulous vessel was doubly ligated and resected. The post-operative course was uneventful with the chest X-ray returning to normal and the murmur disappearing.

DISCUSSION

Congenital coronary artery fistulas are the commonest of the coronary abnormalities, and with the advance of selective coronary angiographic techniques it has been recognized with increasing frequency since first described in 1865.³⁻⁵⁾ About half of the patients are asymptomatic and most of them demonstrate a continuous murmur. A localized abnormal shadow on chest X-ray is rarely noted even in patients with a large tortuous coronary artery fistula, although in this patient the abnormal shadow found on the routine chest film was the only diagnostic clue to a giant coronary artery aneurysm. The chest film of a coronary artery fistula sometimes shows the characteristics of a large left-to-right shunt, but there are no reports of this condition presenting simply as a bulging of the left cardiac border on plain chest X-ray films. Dilatation of the circumflex branch of the left coronary artery may, however, produce a convexity easily mistaken for a left atrial appendage.¹⁾ When an unusual cardiac silhouette is seen, a markedly dilated and tortuous coronary artery should be suspected.^{2,6)} Usually the bulge is slight, but the aneurysm can be large enough to cause bizarre cardiac enlargement.⁷⁾ Aneurysms of the coronary arteries with or without fistulous communications may not only cause localized chest films convexities but may also show calcifications.^{8,9)}

The differential diagnosis includes an enlarged left atrial appendage, absence of pericardium over the left atrial appendage, and a mediastinal mass. Although over half of the patients with a coronary artery fistula have a

continuous murmur with the diastolic component often louder than the systolic, our patient had only a pansystolic murmur which was made continuous with methoxamine. Therefore, phonocardiographic evaluation with a pharmacological test is important in finding the cause of the murmur.¹¹⁾

Two-dimensional echocardiography may also be used to evaluate the proximal left coronary artery, especially dilated one such as with a coronary artery aneurysm.^{12,13)} Contrast-enhanced CT scanning is of value in differentiating whether a mediastinal mass is cardiovascular in origin.¹⁴⁾

Surgical intervention of a coronary artery fistula remains controversial, especially in asymptomatic patients.¹⁵⁻¹⁸⁾ In patients developing cardiac failure, myocardial ischemia, infective endocarditis, pulmonary hypertension, or considerable left-to-right shunting, surgical correction should obviously be performed. On the other hand, in patients with small shunts, minimal cardiac enlargement and few clinical symptoms, surgical management is questionable because very little is known about the long-term natural history of coronary artery fistulae. In the few patients that have been followed for prolonged periods, little anatomical or functional changes occurred with small to moderate shunts, although some tended to increase over the years.¹⁹⁾ However, one of the potential complications of a coronary artery fistula is the development of progressive coronary atherosclerosis. There is one report of rupture of a coronary artery fistula with cardiac tamponade.²⁰⁾

Previous chest films in our patient showed that the aneurysm had been rapidly growing and the cineangiogram revealed a possible coronary steal phenomenon and myocardial ischemia.²¹⁻²³⁾ It is possible that the recurrent trauma from the heavy branches striking his left chest wall might have caused the rapid growth of his fistulous aneurysm. It is our opinion that once an aneurysm is present, early surgical correction should probably be carried out in order to avoid rupture. Especially, a surgical intervention is recommended to the case, which the fistulous aneurysm is more than 3 cm in size or has been rapidly growing.²⁴⁾

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