Rare disease

Myocardial infarction due to coronary steal caused by a congenital unilateral absence of the right pulmonary artery: a rare case

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Summary

A 27-year-old male patient presented with symptoms that mimic an acute coronary syndrome and associated with significant myocardial perfusion ischaemic defects. The latter was detected by adenosine stress Tc-99m estamibi single photon emission CT. However, subsequently the CT angiogram revealed a normal coronary arteries outline and calibre with prominent coronary artery collaterals filling the distal branches of the absent right pulmonary artery.

BACKGROUND

In the literature, to our knowledge, there have only been five reported cases of unilateral absence of pulmonary artery (UAPA) with the affected lung supplied by the coronary artery collaterals. In two of those cases it was reported that the coronary collaterals supplying the affected lung produced negative coronary perfusion defects.

Our patient is a young man who presented with compressing retrosternal chest pain, exertional dyspnoea, new inverted T wave in the inferolateral leads in the ECG, minor troponin leak and myocardial perfusion defect by the myocardial perfusion isotope scan. This resembled the acute coronary syndrome.

CASE PRESENTATION

A 27-year-old gentleman presented to emergency room with a retrosternal chest pain, squeezing in character, and radiating to his left shoulder. Pain was continuous for 30 min and relieved by rest. He had several similar episodes during the week before this presentation. During the past few years he had experienced numerous attacks of right-sided chest pain and shortness of breath on exertion.

There was no recent history of fever or flu like symptoms. He had smoked 10 cigarettes a day for the last 10 years. He has no family history of ischaemic heart disease. He is known to have right unilateral emphysematous lung disease diagnosed by high resolution CT.

Clinical examination showed a tall, thin and underweight young gentleman. Chest examination revealed signs of right side emphysema. Cardiovascular and other systemic examination were unremarkable.

INVESTIGATIONS

- 1) ECG tracing showed inverted T wave in inferolateral leads (lead II, III, aVF, V5 and V6).
- 2) Troponin I was 0.2 (normal: less than 0.01, suspicious: up to 0.33 and 0.34 cut off for acute myocardial infarction).

- 3) Adenosine stress Tc-99m estamibi single photon emission CT (SPECT) results were suggestive of medium size inducible ischaemia of mild to moderate severity over mid- and basal inferior wall, mid- and basal inferolateral wall and mid-anteroseptal wall. Test also showed fixed mild ischaemia (during rest and stress) in basal inferior wall (figure 1).

- 4) CT coronary angiogram showed well-outlined coronary vessels with normal outline and calibre. The main extra cardiac abnormality was agenesis of the right pulmonary artery. The distal right pulmonary artery branches were formed from numerous collaterals from diaphragmatic arteries, intercostals arteries, internal mammary arteries, possibly from bronchial arteries, and from the coronary arteries.

Coronary arteries collaterals were arising from the main right and left coronary arteries, they were relatively small in size.

The prominent collaterals were arising from the mid part of the left circumflex artery and conus branch of right coronary artery supplying the right pulmonary artery branches (figures 2 and 3).

TREATMENT

We noticed that in this patient the use of B blockers improved his cardiac symptoms.

DISCUSSION

UAPA is a rare congenital anomaly,¹ it is either right or left and; it was first described by Fraentzel.² It is a developmental anomaly due to failure in the connection of the sixth aortic arch with the pulmonary trunk.³

Bouros *et al* reported that the prevalence of isolated UAPA is 1:200 000 and it occurs equally in male and female patients.⁴

Harkal *et al* reported that between 1978 and 2000, only 108 cases were published, only 14 of them were

Figure 1 Adenosine stress Tc-99m sestamibi single photon emission CT showing inducible ischaemia over mid and basal inferior wall, mid and basal infrolateral wall and mid anteroseptal wall with a fixed mild basal inferior wall perfusion defect.

asymptomatic. The median age of UAPA patients was 14 years (0.1–58 years).¹

Occasionally, an isolated unilateral absence of the pulmonary artery occurs without cardiovascular anomalies, but more frequently it is associated with cardiovascular abnormalities like falot's tetralogy, septal defects, right aortic arch and patent ductus arteriosus.^{5 6}

Unilateral pulmonary emphysema or unilateral hyperlucent lung was first described by Swyer and James.⁷ In 1954, MacLeod⁸ reviewed 10 cases of one lung transradiancy and analysed clinical, radiological and bronchographic features of the disease in detail. The main radiological feature of the disease is hypertransradiancy of the lung and diminishing of its size. Diagnosis is based on obliteration of the small bronchioles, hypoplasia of the pulmonary artery and absence of the peripheral vascular bed and emphysema.⁹ Ipsilateral emphysema has been reported with isolated unilateral pulmonary artery agenesis. Although the underlying pathogenesis is not fully understood. Other potential explanatory hypotheses for bronchial complications, including the effects of protease-antiprotease imbalance, and relative arterial hypoperfusion, remain subject to debate.¹⁰

In UAPA, a few patients may be asymptomatic. However, the common presentations in the published cases were recurrent pulmonary infections, haemoptysis,

exertional dyspnoea and pulmonary hypertension. In cases with large collateral arteries leading to pulmonary hypertension or haemoptysis, embolisation techniques or pneumonectomy can be done.^{1 4 11} Recently, de Mello Junior *et al* described a patient with UAPA who presented with massive haemoptysis. He was successfully treated with a right pneumonectomy.¹²

The distal part of the absent right pulmonary artery is usually refilled by systemic collaterals from bronchial, intercostals, subclavian or subdiaphragmatic arteries).^{13 14}

In July 2009 Devulapally *et al* published online a case of 36-year-old woman with dextroversion, absence of right pulmonary artery and hypoplastic right lung. Her presenting symptom was exertional dyspnoea, but there was no evidence of cardiac ischaemia.¹⁵

In this case the high resolution CT missed the anomaly because high-resolution CT can depict in detail the gross anatomy and pathology of the lungs and airways. It has been shown to be useful in the evaluation of patients with known or suspected diffuse lung disease.¹⁶ However, CTPA is useful in delineating the anatomic detail of the pulmonary vasculature.¹⁷

To date, there are only four case reports published describing a UAPA and coronary collaterals supplying the affected lung.^{18–21} Two of them reported absence of the right pulmonary artery,^{19 21} while the other two reported

Figure 2 CT coronary angiogram showing absent right pulmonary artery, normal pulmonary trunk and normal left pulmonary artery. With right multiple tortuous enhanced structures posterior to superior vena cava indicate arterial collaterals fillings.

absence of the left pulmonary artery.^{18–20} In three of the four published cases there was no myocardial perfusion defect.^{18–20} However, in the fourth case impairment of the myocardium perfusion and evidence of ischaemia was reported in a 65-year-old female patient.²¹

Recently, in France a new case was published, which described a 44-year-old female who presented a congenital right pulmonary artery agenesis associated with a hypotrophic and multi-cystic right lung complicated with recurrent bronchitis. This patient had a coronary syndrome for which the coronary artery imaging showed a coronary-to-bronchial artery collateralisation with an aneurysm at this level. It gives rise to a coronary syndrome by coronary steal.²²

Our patient is a young man who presented with compressing retrosternal chest pain, exertional dyspnoea, new inverted T wave in the infrolateral leads in the ECG, minor troponin leak and myocardial perfusion defect by the myocardial perfusion isotope scan. This resembled the acute coronary syndrome.

To our knowledge the only two cases reported the UAPA with impairment of the coronary perfusion was published by Kadi *et al*, and this was in a 65-year-old woman,²¹ and De Dominicis *et al* when they described a 44-year-old female who presented a congenital right pulmonary artery agenesis associated a coronary-to-bronchial artery collateralisation with an aneurysm at this level. It gives rise to a coronary syndrome by coronary steal.²² In our case, the patient is male, young and no aneurysm.

Theoretically, the shunt between coronary arteries and the pulmonary collaterals results in steal phenomena which affect the coronary arteries, causing myocardial hypoperfusion, and chronic ischaemia. This was evident in this case and was substantiated by the findings of low normal ejection fraction and the mild global hypokinesia seen in the transthoracic echocardiogram. The positive findings

Figure 3 (A, B) CT coronary angiogram reconstructions (volume-rendering technique and angiography) are showing multiple tortuous collaterals from coronary arteries and right internal mammary artery replacing absent right pulmonary artery.

in the adenosine stress Tc-99m estamibi SPECT and the abnormalities in the ECG findings were also contributory to the diagnosis. All of the former results were consistent with the findings, on the CT coronary angiogram, of prominent collaterals arising from mid part of the left circumflex artery and the conus branch of the right coronary artery.

We also believe that the choice for adenosine might have been beneficial in this case as it augments the degree of steal phenomena. This could certainly have improved the value of the scintigram.

We noticed that in this patient the use of B blockers improved his cardiac symptoms, mainly, chest pain. It is assumed that the improvement in symptoms is due to the beneficiary actions of B blockers, on the myocardium by reducing its oxygen demand. Their effect also improves the coronary filling by increasing the diastolic time effect. This support what was published by Chih *et al* was regarding the medical treatment of coronary artery fistula.²³

CONCLUSION

Congenital right pulmonary artery absence can present in young patients with acute coronary syndrome (type 2 myocardial infarction). This is due to the coronary arteries collaterals supplying the distal part of the absent right pulmonary artery resulting possibly in a steal phenomenon and hence, coronary hypoperfusion and ischaemia. The diagnosis of such cases can be achieved with non-invasive modalities like adenosine stress Tc-99m estamibi SPECT and CT angiogram. The use of B blockers has a role in improvement of the chest pain.

Learning points

- ▶ Cardiac ischaemia resembling an acute coronary syndrome can be caused by coronary steal.
- ▶ CT angiography is a preferred imaging modality in patients with chest pain and a low likelihood for atherosclerosis.
- ▶ β -blocker treatment did relieve symptoms in our patient with coronary steal caused by an unilateral absence of the right pulmonary artery.

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Competing interests None.

Patient consent Obtained.

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