

PULMONARY SEQUESTRATION SUPPLIED BY THE CIRCUMFLEX ARTERY – A RARE CASE REPORT

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Abstract

Pulmonary sequestration (PS) is a rare congenital malformation, even more when its arterial supply is a coronary artery. We present a case of a 68-year-old man admitted in the emergency room with an acute coronary syndrome and no evidence of significant coronary disease. Instead, he had an abnormal branch from the circumflex coronary artery nourishing a mass in the left lower pulmonary lobe. A coronary steal phenomenon was proposed to explain the clinical presentation. An anterior left thoracotomy with ligation of the abnormal branch and atypical resection of the lung segment comprising the sequestration was performed.

INTRODUCTION

Pulmonary sequestration (PS) is a rare congenital lung malformation in which a non-functioning mass of lung tissue develops with abnormal connection to the tracheobronchial tree and abnormal arterial supply.¹ Abnormal arterial supply from a coronary artery is extremely rare², being the term “coronary lung sequestration” proposed by some authors for these cases.³

Here, we present a case of a coronary lung sequestration supplied by the circumflex coronary artery.

CASE REPORT

A 68-year-old man was admitted to the emergency room of his local Hospital with the diagnosis of acute non-ST elevation myocardial infarction. Physical examination was unremarkable, the electrocardiography (ECG) revealed sinus rhythm and 1 mm ST depression from V3-V5. The echocardiogram showed mild left atrium (LA) dilation and normal ventricular function, without any regional wall motion abnormality. The chest X-ray suggested an abnormal shadow on left hilum and the computed tomography diagnosed a just-cisural solid lesion in the left lower lobe (Figure 1). After the initial treatment the patient remained asymptomatic. Maximum troponin I (hs) was 2825 ng/ml.

He had a past medical history of hypertension,



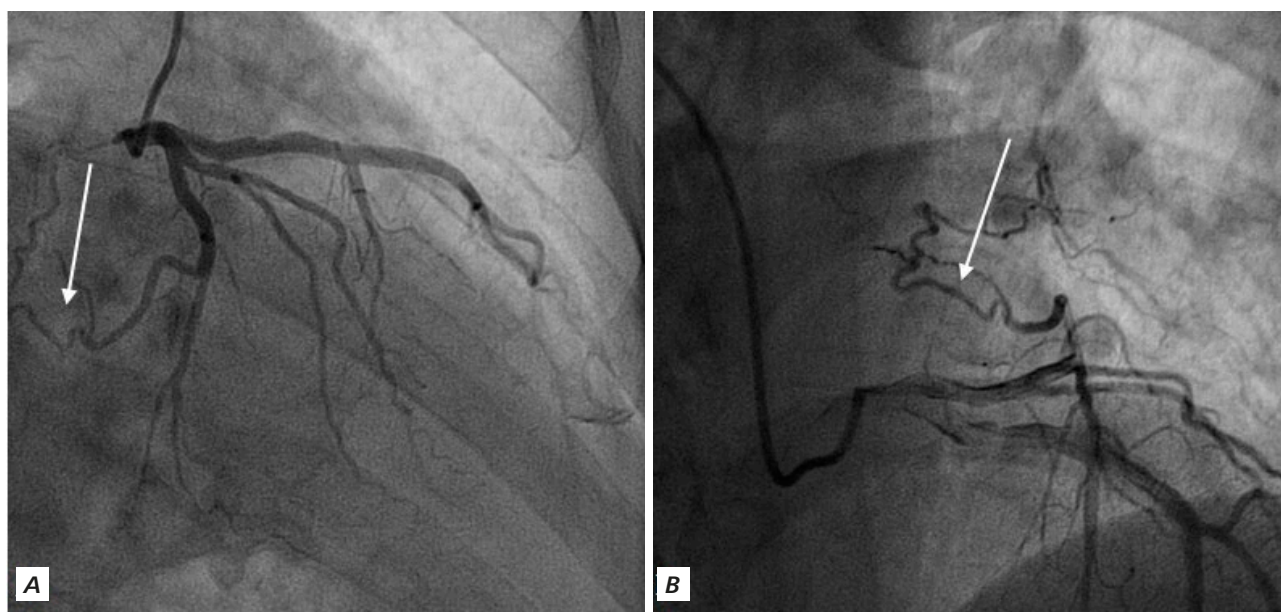
Figure 1

Lesion on the antero-superior segment of the left lower lobe, irrigated by the circumflex artery (arrow).

dyslipidemia and stable angina, with increasing symptoms in the three days before admission. He had no recall of recurrent chest infections.

Coronary angiography revealed absence of significant coronary lesions and an abnormal early large branch arising from the proximal third of the circumflex artery supplying the lesion of the left lung (Figure 2).

In the absence of significant angiographic coronary

**Figure 2**

A and B – Branch from the circumflex coronary artery irrigating the sequestration (arrow).

lesions, a coronary steal phenomenon was proposed to explain the patient's acute coronary syndrome. After Heart Team discussion, he was transferred to our department and submitted to a limited anterior left thoracotomy, with ligation of the abnormal branch and excision of the anterior-superior segment of the left lower lobe, comprising the sequestration (Figure 3 and 4). Pathological examination confirmed an intra-lobar sequestration. The patient was discharged 5 days after surgery, uneventfully. One year after surgery, he remains asymptomatic, with normal left ventricular perfusion, volume and function on a myocardial perfusion scintigraphy.

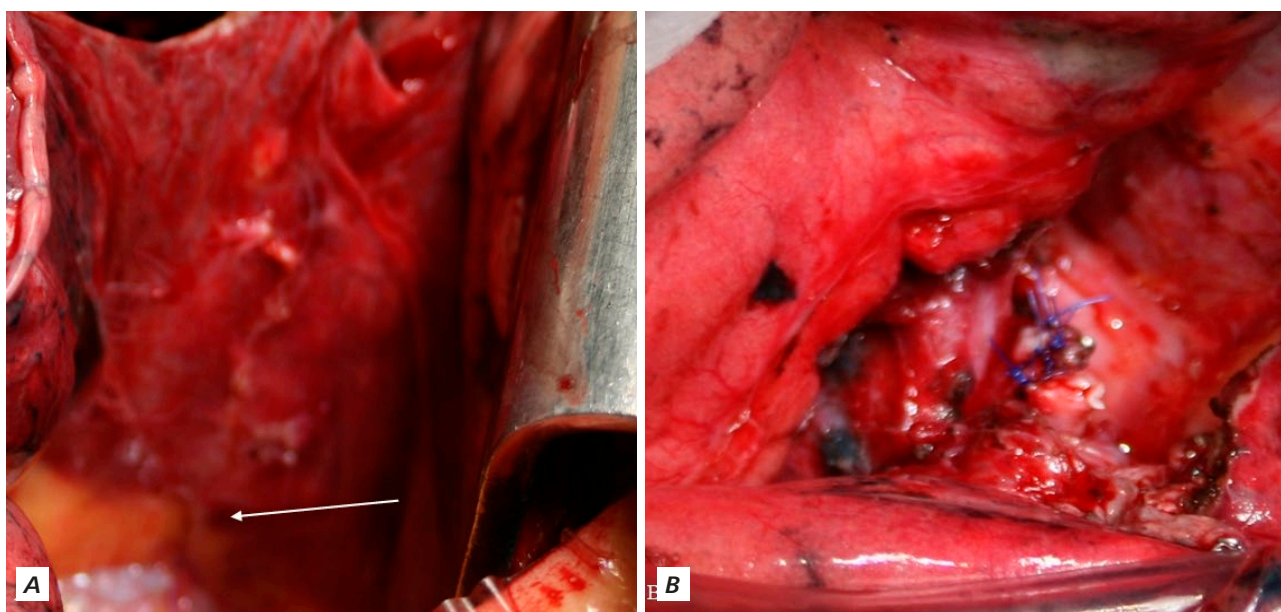
DISCUSSION

Pulmonary sequestration (PS) accounts for only 0.15-6.4% of all congenital pulmonary abnormalities.¹

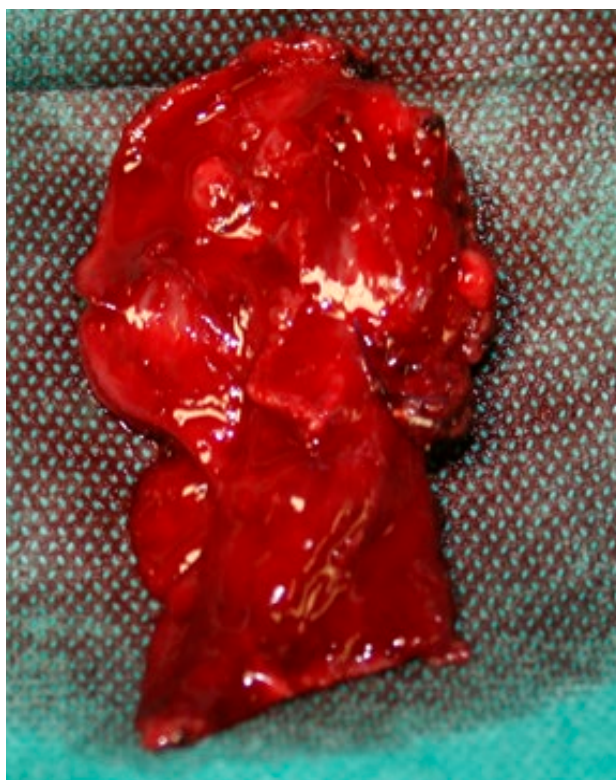
There are two forms of PS: intra-lobar pulmonary sequestration (ILS), representing 84% of all cases^{2,4}, and extra-lobar pulmonary sequestration (ELS).

ILS is surrounded by normal lung parenchyma, almost always in the lower lobes and generally on the left side. ELS is separated from normal lung and has its own visceral pleura.⁵

Most PS receives its arterial supply from a systemic

**Figure 3**

A – Feeding artery to the sequestration; B – After resection of the sequestration with 3 prolene ligatures for vessels.

**Figure 4**

Sequestration. There was no airway connection to the lesion.

artery, mainly from the thoracic aorta (74%), the abdominal aorta (18,7%) and the intercostal arteries (3,2%).^{1,2} Arterial supply of an ILS from the coronary circulation is extremely rare (0,1%)², but when it does occur, the left circumflex artery is the most frequently involved.^{3,6}

Traditionally, the gold standard for the diagnosis of PS was arterial angiography, as it explicitly reveals the aberrant arterial blood supply. Nowadays, with the advent of non-invasive techniques, such as computed tomographic angiography (CTA) and magnetic resonance angiography (MRA), conventional angiography has been replaced.⁷ Both CTA and MRA are reliable imaging modalities for demonstrating the anomalous artery supplying the PS.⁸

The natural history of sequestration supplied by a coronary artery remains unknown because of its rare incidence. Several complications related to PS include recurrent pulmonary infections, unpredictable fatal hemoptysis and heart failure from persistent left-to-right shunt.⁶ The reported case underlies myocardial ischemia due to coronary steal phenomenon as a potential complication of this rare pulmonary sequestrations supplied by a coronary artery, as previously reported by Nakayama.⁹ In this group of patients with an acute coronary event in the absence of native coronary artery disease there is no doubt that a surgical approach is needed.

The discussion arises in the asymptomatic patients with incidental finding of a pulmonary sequestration, with some authors advocating that owing to the potentially severe complications all diagnosed lesions should be removed⁴, and others supporting an expectant approach.^{3,5,10}

Currently, the most consensus treatment for PS is surgical resection. We performed a limited conventional thoracotomy so that we could easily and safely identify the abnormal arterial branch and the pulmonary sequestration.

In centers experienced in video-assisted thoracic surgery (VATS) it would be a feasible alternative method for PS resection and abnormal vessel ligation.¹¹

Preoperative imaging evaluation is essential to identify the location of the arterial supply and pattern of venous drainage, especially when a minimally invasive approach is to be considered.^{2,12} In PS with arterial supply from a coronary artery, beside the usual imaging tests, a coronary angiography is also required both for diagnosis and to exclude hemodynamically significant atherosclerotic disease.

Recent studies have suggested interventional therapy, such transcatheter endovascular embolization and thoracic endovascular stent-graft exclusion, for treatment of PS.¹²

CONCLUSION

Although rare, coronary steal phenomenon from a pulmonary sequestration may cause myocardial ischemia and acute coronary events. It underlies the importance of early recognition and correct diagnosis of this entity to assure the best clinical management.

Several surgical and interventional approaches have been proposed, but no treatment guidelines have yet been established.

Therefore, it is essential to report all cases of lung sequestration with coronary arterial supply both to better understand its natural history and define its better clinical approach.

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