Case report - Thoracic non-oncologic

Right pulmonary artery agenesis and coronary-to-bronchial artery aneurysm

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Received 11 August 2010; received in revised form 29 November 2010; accepted 2 December 2010

Abstract

Isolated unilateral pulmonary artery agenesis is a rare congenital anomaly that may be complicated with hemoptysis, recurrent pulmonary infections or pulmonary hypertension. To our knowledge the occurrence of a coronary syndrome associated with a coronary-to-bronchial artery saccular aneurysmal collateralization has never been described before. A 44-year-old female presented a congenital right pulmonary artery agenesis associated with a hypotrophic and multicystic right lung complicated with recurrent bronchitis. This patient had a coronary syndrome for which the coronary artery imaging showed a coronary-to-bronchial artery collateralization with an aneurysm at this level. It gives rise to a coronary syndrome by coronary steal. Two bronchial collaterals arising from a diaphragmatic artery and the subclavian artery were also found on the computed tomography (CT)-scan. This last collateral also showed another saccular aneurysm. We first performed an embolization of those two aneurysms in order to decrease the risk of hemorrhage and coronary steal, before performing a right pneumonectomy. In this case, the surgery was indicated because of the pathological lung and the risk of postembolization ischaemia. The postoperative course was uneventful and the patient was doing well six months later.

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Keywords: Aneurysm; Pneumonectomy; Agenesis; Pulmonary artery

1. Introduction

Isolated unilateral pulmonary artery agenesis is a rare congenital anomaly that may be complicated with hemoptysis, recurrent pulmonary infections or pulmonary hypertension. The occurrence of a coronary syndrome associated with a coronary-to-bronchial artery saccular aneurysmal collateralization is exceptional.

2. Case report

We report the case of a 44-year-old female, a mother of eight children, with a medical history of hypertension, asthma, obesity (BMI = 37), and recurrent bronchitis secondary to a congenital right pulmonary artery agenesis (Fig. 1a), associated with a hypotrophic and multicystic right lung (Fig. 1b and c). She had presented at the emergency department complaining of chest pain. The explorations found a ST-segment depression of at least 3 mm on the echocardiogram (ECG) on the inferior leads (II, III and aVF) and on lateral leads (V5 and V6), the blood test did not show cardiac enzymes modifications, but diabetes mellitus was incidentally discovered.

The emergency coronary artery imaging showed a spasm on the left anterior descending artery, which regressed after administration of a nitrate, as well as a coronary-to-bronchial artery collateral arising from the right postero-lateral coronary artery, with a 6-mm saccular aneurysm at this level (Video 1). This suggested a coronary syndrome by coronary steal. Two bronchial collaterals arising, respectively from a diaphragmatic artery and the subclavian artery were also found on the computed tomography (CT)-scan. The latter also showed another aneurysm. We did not find any other cerebral or abdominal aneurysm. The indication for an embolization of those aneurysms, followed by a right pneumonectomy was planned. Pulmonary function tests found a forced expiratory volume (FEV) at 1.9 l (69% of the theoretical value) and blood gas were normal. The lung scan showed 31% of ventilation and 0% of perfusion on the right side. The ECG showed a left ventricular ejection fraction of 60%.

We first performed an embolization of these two aneurysms. It was done by a retrograde femoral artery approach and Tornado coils were dropped into the saccular aneurysm from the bronchial artery collateral arising from the right coronary artery (Video 2) and into the one from the subclavian artery. Immediate angiographic control confirmed the aneurysm exclusion and thrombosis. During the procedure, the patient experienced a chest pain and an
Six hours after the operation the patient had a new chest pain, without electrical modification and as the troponinemia was decreasing. Therefore, this pain was probably related to the ischemia of the pulmonary embolized areas. Three days later, when the troponinemia was normal, a right intrapericardial pneumonectomy was performed. The pneumonectomy included a ligation of all the collaterals coming from diaphragmatic, coronary and subclavian arteries (Fig. 2a and b). The bronchial stump was reinforced by a pericardial flap pedicled on the phrenic artery and a pericardial prosthesis was fixed to prevent cardiac luxation. The main postoperative complications were an atrial fibrillation and a hypoxemia, which both resolved after medical treatment. The postoperative echocardiography was unchanged. After six months, the patient was asymptomatic, without chest pain and the CT-scan control showed a normal right pneumonectomy cavity, without any new aneurysms.

3. Discussion

Isolated unilateral pulmonary artery agenesis is a rare congenital anomaly [1]. It can remain asymptomatic or become complicated with hemoptysis [2], recurrent pulmonary infections or a pulmonary hypertension [1]. On the affected side, there are either persistence of embryonic arteries (from the aorta or its branches), hyperplasia of
normal bronchial arteries and/or abnormal acquired collateralization arising from the bronchial, subclavian, intercostal or diaphragmatic arteries.

The coronary-to-bronchial artery collateralization is also a rare anomaly, which is often described in association with a localized or more diffuse bronchiectasis [3, 4] but is only rarely the site of an aneurysm [5]. It may be responsible for coronary steal [6, 7] or hemoptysis [8]. In each case, a treatment by embolization is most often described [6], or more rarely found by surgery when another procedure is necessary or in case of embolization failure [8].

The association of a unilateral pulmonary artery agenesis and coronary-to-bronchial artery collateralization has been described a few times in the literature [9, 10]. One reports a case complicated with a coronary steal and a myocardial ischemia in an elderly patient [10].

The case we are reporting is the association of these two anomalies with an aneurysm on the coronary-to-bronchial collateral. The presence of this aneurysm and of a coronary syndrome on the coronary steal motivated a more aggressive treatment.

A two-stage treatment, with at first an embolization of the aneurysms on the bronchial arteries vascularising the right lung being performed. Indeed, we thought that surgical ligation into the mediastinum by a posterolateral thoracotomy approach might have a risk of hemorrhage and therefore, of myocardial ischemia.

Performing a pneumonectomy in a second stage was necessary because of a multicystic lung responsible for chronic infections, but also because of a necrosis and risk of abscessing due to the embolization of an important part of the right lung vascularize.

For our patient, the multicystic lung should have been removed years before, when the bronchitis began, this delay probably enabled the development of bronchial arteries aneurysms.

4. Conclusion

In the case of an isolated unilateral pulmonary artery agenesis, the supplying vessels often consist of aberrant bronchial arteries, which may lead to complications, such as hemorrhage. The collateralization of a bronchial artery with a coronary vessel is exceptional and its symptomatology and management are very specific. The beginning of such may lead to the development of aneurysms, most often in the case of an association with a bronchiectasis. In this case, the combination of interventional radiology and only surgery achieved a satisfactory outcome.

References


eComment: Congenital isolated unilateral absence of pulmonary artery and variants of collateral blood supply of the ipsilateral lung

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doi:10.1510/icvts.2010.250795A
We compliment the authors on their description of a unique combination of rare congenital anomalies and the choice of an optimal strategy for surgical treatment of this pathology [1]. We would like to comment on several problems related to this case.
We found 352 cases of congenital unilateral absence of pulmonary artery (UAPA) in the world literature; an additional 67 cases were diagnosed in the Bakoulev Scientific Center for Cardiovascular Surgery. In 237 out of 419 cases, UAPA was associated with congenital heart defects. Patients with absent left pulmonary artery (LPA) prevailed in this group (75%, 177/237). Conversely, among 182 patients with isolated pulmonary artery (PA) agenesis, the right pulmonary artery (RPA) was absent in 60% of cases (109/182).
Several variants of collateral blood supply to the ipsilateral lung were described in 46% of cases (83/182). Only in eight cases (4.4%, 8/182) was isolated UAPA associated with coronary–bronchial communication or an anomalous vessel between the coronary artery and ipsilateral lung vessels. The rate of anomalous coronary collateral communication is similar in isolated absence of the left and right PA: 4.1% (3/73) with absence of LPA and 4.5% (5/109) with absence of RPA. Among five cases of isolated absence of the RPA there were two cases with an anomalous vessel arising from the circumflex branch of the left coronary artery to the right lung, two cases with an anomalous vessel to the right lung arising from the right coronary artery, and the fifth patient described in this paper had a communication between the right coronary and the bronchial arteries. Thus, the presented association of isolated absence of the RPA with the communication between the right coronary and the bronchial arteries is the first one to be described in the literature.
Despite the presence of anomalous communications between the coronary arteries on the one hand, and bronchial or pulmonary arteries on the other hand, coronary steal syndrome was revealed in only two out of eight patients. Doppler echocardiography and myocardial perfusion scintigraphy data suggested undisturbed myocardial perfusion [2]. According to the authors, the blood flow in the circumflex branch of the left coronary artery occurred mainly during diastole, while the blood flow in the collateral vessel occurred mainly in systole.
We consider the staged approach to be justified in the presented case. A similar approach was described in 1987 for a patient with absence of the LPA and peribronchial ‘arteriovenous malformation’ [3]. It is necessary to note that the presence of the hilar artery in the ipsilateral lung makes it possible to perform the operation aimed at the restoration of antegrade blood flow in the affected lung [4]. Such surgical intervention performed during childhood provides normal development of the pulmonary vessels and prevents the development of collateral blood flow.
flow in the ipsilateral lung, as well as preventing pulmonary hypertension in
the contralateral lung.

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