Case report - Thoracic general

Isolated absence of the right pulmonary artery as a cause of massive hemoptysis

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Abstract

Isolated absence of a pulmonary artery is a rare cause of massive hemoptysis. We report a case of unilateral absence of the right pulmonary artery in an adult without any other cardiovascular anomalies. The patient presented massive hemoptysis, which was successfully treated with a right pneumonectomy.

Keywords: Isolated unilateral absent pulmonary artery; Massive hemoptysis; Pneumonectomy

1. Introduction

Patients with isolated unilateral absent pulmonary artery (isolated UPAA) usually survive into adulthood experiencing only minor, if any, symptoms. While about 20% of the patients with isolated UPAA develop inconsequential hemoptysis, massive hemoptysis is a very rare complication of this anomaly [3, 4]. The following case report is about a 30-year-old man with isolated absence of the right pulmonary artery who presented massive hemoptysis, which was successfully treated with a pneumonectomy of the affected side.

2. Case report

A 30-year-old man was admitted due to massive hemoptysis. Upon admission, his body temperature was 36.6 °C, pulse rate of 115 beats/min, respiration rate of 20/min, and blood pressure of 100/70 mmHg. No clinical signs of cyanosis, edema, or clubbing of the fingers were found. Cardiac auscultation was normal. Chest auscultation revealed ronchus in the right lung. A chest radiograph revealed cardiac and mediastinal displacement to the right, right hemi-diaphragm elevation and a smaller right hemithorax (Fig. 1). A CT-scan revealed the absence of the right main pulmonary artery. Blood was supplied to the right lung by systemic arterial branches originating from the right internal thoracic artery, right axillary and intercostal branches and diaphragmatic artery, with a consequent increase of the bore of those vessels (Fig. 2a–h).

Abnormally calibrous, tortuous vessels were detected at the middle lobe and the anterior segment of the upper right lobe. A focal vascular dilation with approximately 10 mm of diameter, possibly representing an aneurysmatic formation or an arteriovenous fistula, was also detected at the latter topography. The electrocardiography revealed sinus tachycardia, while no structural cardiac changes were detected by the echocardiogram. The pressures in the cardiac chambers were normal. Since the situation was life-threatening, the patient was conducted to the surgery room to be subjected to an emergency pneumonectomy. A bronchoscopy was performed immediately after anesthetic induction and revealed a great amount of blood and blood clots emerging from the main right bronchus.

Upon opening of the chest, countless vessels were detected between the thoracic wall and the lung surface, most of them on the anterior surface of the lung and between the lower lobe and the diaphragm. They were sectioned either by electrocautery or spring-loaded clips. After the liberation of the lung, a calibrous arterial branch was detected originating from the inner thoracic artery and penetrating the lung at the middle lobe. It was dissected and sectioned with double-ligature. Next, two more calibrous arterial branches were detected penetrating the base of the lower lobe after crossing the diaphragm. They were doubly ligated and sectioned. Calibrous bronchial arteries were dissected and sectioned after double ligation. Pulmonary veins were isolated and also sectioned after double ligation. The right main stem bronchus was dissected, sectioned and sutured. The surgery was conducted without any complications and the patient released from the hospital on the sixth postoperative day.

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Fig. 1. PA chest radiograph revealing cardiac and mediastinal displacement to the right, right hemi-diaphragm elevation and a smaller right hemithorax.

Fig. 2. (a) Axial image showing the absence of the right main pulmonary artery. (b) Right inner thoracic artery with increased bore. (c) Prominent bronchial and intercostals arteries on the right. (d) Calibrous right axillary artery. (e) Anomalous, tortuous calibrous vessels penetrating the middle lobe. (f) Focal vascular dilation on anterior segment of the upper right lobe, possibly representing an aneurysm or arteriovenous fistula. (g) Image on coronal plane showing the contrast between the sizes of both lungs. (h) 3D image showing the absence of the right main pulmonary artery, the prominence of the inner thoracic artery and the anomalous vascularization with tortuous arteries on the right lung.

3. Discussion

The unilateral absence of a pulmonary artery is a rare congenital anomaly and is frequently associated with other cardiovascular anomalies, such as tetralogy of Fallot, septal defects, right aortic arch and persistence of ductus arteriosus [1, 5, 6, 8]. Most cases are diagnosed and surgically treated in the patient’s first year of life [7]. Nevertheless, many patients with isolated UPAA have a benign clinical course and a diagnosis may not be made until they reach adulthood [1]. When symptomatic, patients with isolated UPAA may experience recurrent respiratory infections, dyspnea on exertion, high-altitude pulmonary edema, pulmonary hypertension in the contralateral lung or hemoptysis [1, 2].

Hemoptysis occurs in about 20% of cases of UPAA and can be self-limiting for many years, but may also lead to massive pulmonary hemorrhage and death [3].

Hemoptysis in patients with isolated UPAA has been attributed to the extensive systemic collateral circulation on the affected lung. The systemic collaterals usually come from the bronchial, intercostals, subclavian or subdiaphragmatic arteries [3]. Massive hemoptysis can only be treated with selective embolization of the bronchial and non-bronchial systemic arteries [9] or pneumonectomy of the affected side [4, 10].

In the presented case, the patient remained asymptomatic until adulthood. The clinical presentation began with hemoptysis that eventually became life-threatening. The patient’s plain chest radiograph had characteristic findings, consisting of ipsilateral cardiac and mediastinal displacement, smaller right hemithorax, ipsilateral hemidiaphragm elevation, highly diminished pulmonary vascular markings, contralateral lung hyperinflation and herniation beyond the midline [1]. The CT-scan permitted the detection of the absence of the right main pulmonary artery, as well as of the presence of extensive collateral circulation originating from the right internal thoracic artery, right axillary and intercostals branches and diaphragmatic artery.

Selective embolization of the bronchial and non-bronchial systemic arteries is an indicated procedure for the management of massive hemoptysis in patients who, due to poor pulmonary reserve and other co-morbid conditions, are not surgical candidates [9]. However, in the presented case,
the endovenous approach was not possible inasmuch as the hospital lacked technical conditions for the realization of selective embolization. Therefore, since surgery was not contraindicated by the patient’s clinical condition and considering the urgency of the situation, the team decided for the pneumonectomy.

Supposing that selective embolization of the systemic collaterals was technically feasible in the presented case, on the one hand the risk of submitting the patient to a surgery would have been avoided. On the other hand, factors such as the serious technical difficulty in locating the precise embolization sites for the safe release of coils (which could demand more time and eventually compromise even more the patient’s clinical condition), the relatively high long-term recurrence rates and the possibility of occurrence of several complications related to selective embolization[9] would make a careful evaluation necessary before deciding to treat the patient with this procedure in this specific case.

References