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Case report

Therapeutic embolization of a systemic arterialization of lung without sequestration

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Abstract. We report the case of a 51-year-old man with massive haemoptysis due to a systemic arterialization of lung without sequestration. Unlike bronchopulmonary sequestration there was a normal bronchial distribution and the involved lung parenchyma was normal. Therefore a therapeutic transarterial embolization of the aberrant systemic vessel from the distal thoracic aorta was performed. The embolization was successful and the patient did not suffer from further haemoptysis during the subsequent follow-up of ten months. A postembolization aortogram 6 months later demonstrated a complete occlusion of the embolized aberrant artery; in the lung perfusion scan there was only a small perfusion defect, but normal ventilation in the embolized basal part of the left lower lobe. Our case represents an alternative treatment to surgery for this rare anomaly.

Key words: Lung – Pulmonary artery abnormalities – Therapeutic embolization – Haemoptysis – Lung sequestration

Introduction

Systemic arterialization of lung without sequestration is the rarest form of congenital anomalous systemic arterial supply to the lungs [1, 2]. In this rare anomaly the arterial supply of one or more of the basal segments of the lower lobe derives from an aberrant vessel arising from the aorta. The non-sequestrated lung parenchyma, which is supplied by the aberrant artery, has no parenchymal or bronchial abnormalities and there is a normal connection with the bronchial tree.

All cases of this rare abnormality which have been described in the literature up to now [1–13] have been treated by thoracotomy with ligation of the abnormal artery and/or resection of the involved lung parenchyma.

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Case report

A 51-year-old man was admitted with a first-episode of massive haemoptysis. His past history and his family history were unremarkable. The physical and laboratory findings were within normal limits. A chest radiograph demonstrated a patchy area of consolidation in the left basal lower lobe. Bronchoscopy showed bleeding from the left lower lobe, but no tumour or stenosis was found. The tracheobronchial tree was reported as normal in size, configuration and position. Spiral CT with bolus contrast technique showed a large anomalous artery arising from the lower thoracic aorta and supplying basal segments of the left lower lobe. Venous drainage from the involved segments was towards the inferior left pulmonary vein. There were no bronchial or parenchymal abnormalities except a hypervascularity in the basal segments of the lower lobe (Fig. 1). The suspected diagnosis of a systemic arterialization of lung was confirmed by angiography. The pulmonary angiogram revealed normal branching of the pulmonary artery into the left lower lobe. But compared with the opposite side, the calibre of all the segmental arteries of the left lower lobe was smaller (Fig. 2). Corresponding to CT findings, aortography and selective angiography demonstrated a large arterial branch (11 mm in diameter) arising from the distal thoracic aorta and supplying the basal segments of the left lower lobe. Venous drainage was into the left atrium by the left inferior pulmonary vein (Figs. 3, 4 a). Thus, the same vein drained both the pulmonary circulation and the aberrant artery.

After selective angiography of the aberrant artery with a 5-F Cobra Catheter (Cook Europe, Denmark) a coaxially placed microcatheter (FasTracker 18, Target Therapeutics, Fremont, Calif.) was positioned proximal to its branching. The embolization was carried out with platinum microcoils (interlocking detachable coils, complex helicoidal fibred platinum coils; Target Therapeutics, Fremont, Calif.) and Tornado coils (Cook Europe, Denmark). After 5 min, there was no detectable flow in the embolized vessel.

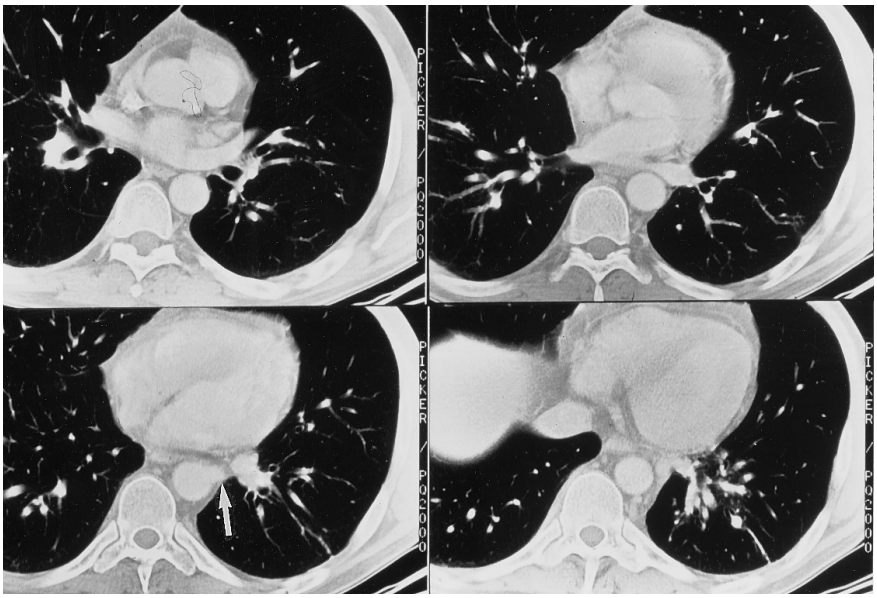


Fig. 1. Axial CT sections through the lower thorax at different levels of the lower lobe. No bronchial or parenchymal abnormalities are visible in the posterobasal segment of left lower lobe. The only abnormality in the left lower lobe (besides a fine linear atelectasis) is hypervascularity. The origin of the systemic artery to the lung is marked with an *arrow*

Six months later, the patient was without symptoms and a follow-up angiogram demonstrated a total occlusion of the aberrant systemic artery (Fig. 4 b). Ventilation-perfusion lung scans showed only a small perfusion defect in a part of the posterobasal segment of the left lower lobe, which was much smaller than the area of the lung supplied by the aberrant artery.

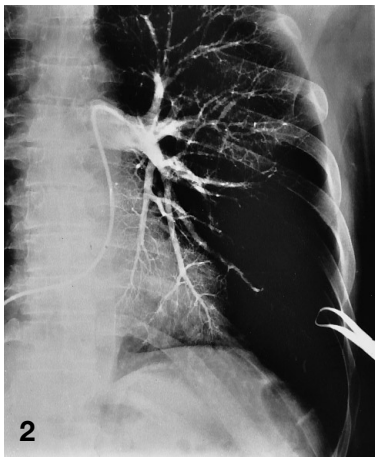
There were no postembolization complications and haemoptysis has not recurred during the follow-up period of 10 months after embolization.

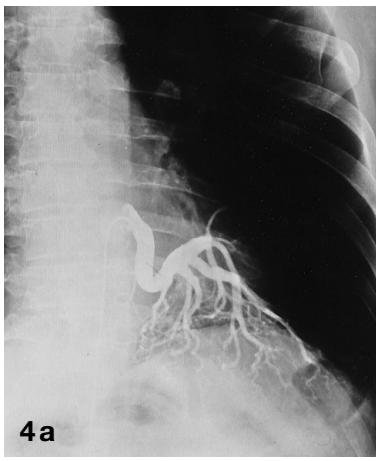
Discussion

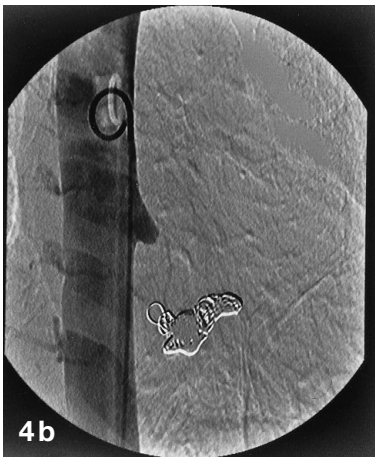
Anomalous systemic arterial supply to the lungs has been described in bronchopulmonary sequestration, the scimitar syndrome and in systemic arterialization of normal lung. In the latter, which Pernot et al. [3] also called a systemic artery–pulmonary vein fistula without sequestration and normal lung parenchyma with no structural or bronchial abnormalities, is supplied by a systemic artery. This vessel always supplies one of the lower lobes, most frequently one of the basal segments [1–13]. The absence of parenchymal abnormalities and a normal bronchial supply clearly distinguish systemic arterial supply without sequestration from true sequestration [4]. This malformation must also be distinguished from pulmonary arteriovenous fistula, where there is an abnormal communication between pulmonary arteries and pulmonary veins producing a right-to-left shunt. The most frequent clinical symptoms are dyspnoea, haemoptysis and central nervous system complications, namely strokes and abscesses [14]. The feeding vessel in systemic arterial supply of normal lung without sequestration arises, as in our case, from the distal part of the thoracic descending aorta [4–9], from the proximal abdominal aorta or from the coeliac axis [2, 4, 6–8, 10–13]. The systemic artery can measure up to 1 cm in diameter [10]. In most cases an atresia of the corresponding pulmonary artery is present [2–6, 9–13]. There are also some cases [7, 8] with normal branching of the pulmonary artery in the involved segment as in our case. The venous return is always via the normal inferior pulmonary vein [2–13]. In effect, the anomaly is a left-to-left shunt.

Systemic arterialization of lung without sequestration corresponds to type I of the sequestration complex, which was first described by Pryce et al. [1]. The sequestration complex comprises several overlapping clinical, radiographic and pathological entities whose common denominator is an anomalous systemic arterial supply to the lungs. The lesion is probably related to bronchopulmonary sequestration which is defined as an area of nonfunctioning lung tissue often containing cysts, which receives its blood supply from a systemic artery and characteristically has no connection with the tracheobronchial tree [15]. Because of the absence of abnormalities of lung parenchyma in the involved segments and a normal bronchial distribution, systemic arterialization of lung without sequestration is distinct from true sequestration [4]. Moreover, accompanying anomalies have not been described in systemic arterialization of lung, whereas 14 % of cases with intralobar sequestration and 59 % of cases with extralobar sequestration are associated with other congenital malformations [15].

Symptoms of systemic arterialization without sequestration are variable. In most patients, especially in children, systemic arterialization of lung is asymptomatic and is discovered following the incidental finding of a continuous murmur best heard over the left anterior chest and the back [3]. However, left ventricular failure may result from the left-to-left shunt [8]. In adults it is often asymptomatic presenting as an incidental chest-film finding [2]. Massive haemoptysis seems to be an unusual manifestation. In our literature survey we found only 3 of 18 patients with systemic arterialization to normal lung who had haemoptysis [2, 5, 10].







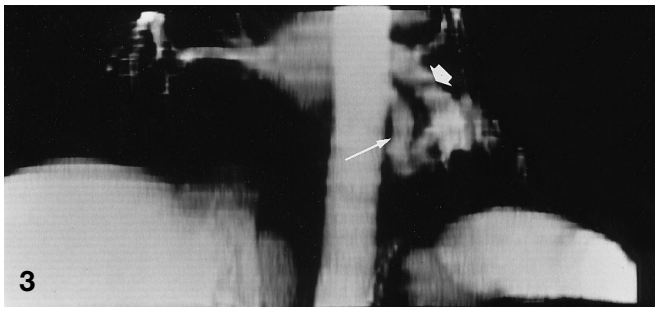


Fig. 2. Left pulmonary angiogram. Normal branching of the left pulmonary artery with small calibre of its segmental arteries

Fig. 3. Reconstructed CT angiogram shows an abnormal artery from the distal thoracic aorta *(long arrow)* descending to the left lower lobe. Venous drainage is by the left inferior pulmonary vein *(short arrow)*

Fig. 4. **a** Selective angiogram of aberrant artery before embolization. **b** Follow-up aortogram after 6 months shows complete blockage of the aberrant artery by multiple platinum microcoils and thrombosis

The treatment of all patients with this rare anomaly up to now has been surgical. In most cases the anomalous vessel was ligated and the involved lung parenchyma was resected [2–6, 8–12]. If the parenchyma is normal, a ligation of the systemic vessel only may be sufficient [3, 7]. This is in contrast to bronchopulmonary sequestration, where most authors agree that this anomaly should be treated by ligation of the systemic vessel and resection of the sequestred lung parenchyma because of its propensity for recurrent infection [15, 16]. Campbell et al. [7] described a 14-month-old male with systemic arterialization of normal lung who was treated by surgical ligation of the aberrant vessel only. The patient did well during the subsequent follow-up of 1 year. A similar case of a 7-year-old boy is reported by Varma and Clarke [13]. In this case too a surgical ligation of the systemic vessel only was performed.

Transarterial embolization has been well established in the management of haemoptysis [17]. Hayakawa et al. [18] presented a case of a 68-year-old woman with massive haemoptysis due to bronchopulmonary sequestration. Initially, the diagnosis of sequestration was not considered and bronchial artery embolization was performed for management of haemoptysis from the left lower lobe. Four days later, after one more episode of pulmonary haemorrhage, the feeding artery was detected and was embolized with a gelatin sponge. Complete haemostasis resulted until elective surgical ligation 2 months later. A similar case is reported by Gasparini [19]. The systemic artery of intralobar sequestration was embolized with a fibrin sponge and sclerosing agents. No surgical ligation or resection was done and the patient remained free of symptoms during the subsequent follow-up of 32 months.

Our case is different from these reported cases because it was not a bronchopulmonary sequestration and embolization was used as the only treatment. We decided to embolize the aberrant systemic artery, because the involved lung parenchyma had no abnormalities, normal segmental branching and was also supplied by segmental pulmonary arteries. Due to this double arterial supply of the involved lung parenchyma, a significant perfusion defect was absent in lung scintigraphy after embolization.

To our knowledge, we describe the first transarterial therapeutic embolization of systemic arterialization of lung without sequestration. Based on the favourable result on follow-up in our patient, this treatement may be considered an alternative to thoracotomy in patients with systemic arterialization of lung without seqestration. However, bronchial and pulmonary arterial supply to the segment involved must first be verified by bronchography or CT, and pulmonary angiography, respectively.

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Book review

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Farr, R. F., Allisy-Roberts, P. J.: Physics for Medical Imaging. London: W. B. Saunders 1997. 276 pp., 130 figs., (ISBN 0-7020-1770-1), £ 22.50.

The book *Physics for Medical Imaging* by R. F. Farr and P. J. Allisy-Roberts with a contribution from J. Weir follows the revised medical physics curriculum for Part I of the Fellowship of the Royal College of Radiologists (FRCR) examination and is intended primarily as a review book for trainee radiologists preparing for this examination. The text is divided into eight chapters covering all aspects of modern medical imaging from basic radiation physics and conventional film-screen radiography to digital imaging, computed tomography, ultrasound and magnetic resonance imaging. Each chapter concludes with several review questions which are cross-referenced at the end of the book with the pages covering this specific topic.

Trying to comply with the space limitations of a small paperback book, the text is written in a very concise style with some chapters just giving short definitions of important key words. This may be ideal for examination preparation, but makes the book hard to read for someone who is interested in a broader context. This drawback is compensated for in part by a large number of easy-to-understand line drawings and tables supporting the text.

The strength of the book certainly lies in the first three chapters dealing with the physics of conventional X-ray imaging. Written for physicians and not medical physicists, it succeeds in explaining the underlying physical principles in a straightforward manner avoiding complex mathematical formulae. Compared with the first three chapters, the chapter dealing with digital imaging is more than disappointing. Less than one page is devoted to new digital receptors and picture archiving and communication systems (PACS). Storage phophor technology is mentioned just briefly in a paragraph entitled 'silverless radiography'; newer techniques such as those based on selenium are not mentioned at all. The book does not fail to incorporate a chapter on nuclear medicine, which also includes a section covering the newer tomographic techniques SPECT and PET. The first part of the book dealing with imaging modalities based on ionizing radiation is concluded by a chapter on radiation hazards and protection. This chapter covers general principles of radiation protection as well as specific aspects of UK radiation protection legislation.

To complete the spectrum of modern medical imaging, the last two chapters deal with the physics of ultrasound and magnetic resonance imaging. Since it would be beyond the scope of this book to cover all aspects of magnetic resonance imaging physics, the authors concentrate on the three basic pulse sequences, gradient fields and safety aspects.

The book appears well suited for its intended use as a preparation tool for the FRCR examination, but it will not be the right choice for someone who is looking for a thorough review of medical physics.

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