Monaldi Arch Chest Dis
2008; 69: 3, 137-141

CASE REPORT

Non-invasive diagnosis in a case of bronchopulmonary sequestration and proposal of diagnostic algorhythm

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ABSTRACT: ***Non-invasive diagnosis in a case of bronchopulmonary sequestration and proposal of diagnostic algorhythm. P. Caradonna, M. Bellia, F. Cannizzaro, S. Regio, M. Midiri, V. Bellia.***

The case of a 43-year-old woman with intralobar pulmonary sequestration, Pryce type one, is presented. The medical history was characterised by recurrent bronchopneumonia, productive cough with purulent sputum and hemoptysis in the last three years. Diagnosis was made by CT angiography: multiplanar, maximum intensity projection and volume rendering reconstructions were visualised. A volume reduction of middle and lower lobe with multiple cyst-like bronchiectasis was detected and no evident relationship with tracheobronchial tree was pointed out. Reconstructions aimed at evaluating bronchial structures demonstrated no patency of middle and lower lobar bronchi. The study carried out after contrast medium infusion in arterial phase showed a vascular disorder characterised by an accessory arterial branch arising from the upper portion of thoracic aorta which, after moving caudally to pulmonary hilus with a tortuous course, supplied the atelectatic parenchyma. No anomalous venous drainage was detected. The patient underwent surgery with resection of two pulmonary lobes. CT compares favourably with other alternative imaging technique for pulmonary sequestration as multiplanar reconstructions allow not only the detection of supplying vessel, but also the accurate description of heterogeneous characteristics of the mass and adjacent structures. Finally an imaging-based diagnostic algorhythm is proposed.

Monaldi Arch Chest Dis 2008; 69: 3, 137-141.

Keywords: Bronchopulmonary sequestration, CT angiography, Lung imaging.

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Introduction

Pulmonary sequestration is a rare malformation, characterised by a portion of abnormal lung tissue which has no/or partial bronchial connections, separated by the surrounding tissue of the lung and with an anomalous systemic blood supply. A medical history of recurrent pneumonia, hemoptysis, chest pain and productive cough with purulent sputum and fever, is a frequent feature of pulmonary sequestration. We report a case of a woman with intralobar sequestration Pryce's type I diagnosed by a non invasive method and propose an imaging-based diagnostic algorhythm.

Case Report

A 43-year-old woman was hospitalised as a result of recurring hemoptysis, fever (37.5-38°C), cough with purulent sputum and dumb chest pain on the right side. History included a right pleurisy of likely tuberculosis origin at the age 23 followed by a bronchographic diagnosis of cystic bronchiectasis. Three years before the admission she started suffering deep pain in the right hemithorax, cough with purulent, sometimes blood striped, smear, and episodic fever. Physical examination pointed out diminished breath sounds, with crackles and rales at the lower right lung field. Foul smelling breath unrelated to oral or oesophageal disorders was noticed.

Finger clubbing, but no cyanosis or shortness of breath, was noticed with leukocytosis (14.5 x 10^3/dl) and neutrophilia (89.3%).

Chest radiograph (figure 1) highlighted a paramediastinal parenchymal shadowing in the right basal zone with asymmetry of pulmonary fields due to volumetric reduction of right hemithorax. Chest CT scan was performed by a 40-slice multidetector computed tomography (MDCT) scanner (Brilliance 40, Philips Medical System, Cleveland, Ohio, USA). The examination was performed before and 25s after the administration of 100 ml of iodinated non-ionic contrast material (iomeprol, Iomeron 400, Bracco, Italy) at a flow rate of 3.5 ml/s, using the following scan parameters: collimation, 40 X 0.625 mm, gantry rotation time, 420 ms, slice thickness, 1,5 mm, slice increment, 0.7

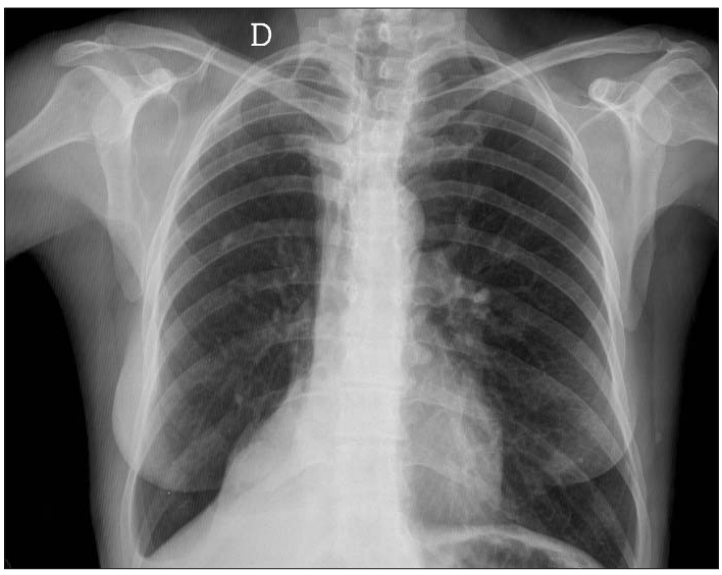

Fig 1. - Chest radiograph: paramediastinal parenchymal opacity may be noticed in the right basal zone with asymmetry of pulmonary fields due to volumetric reduction of right hemithorax.

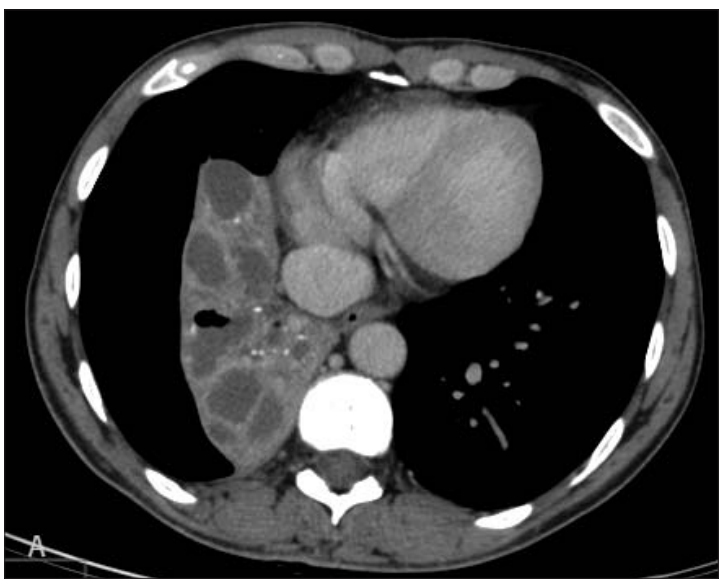

Fig. 2 - CT scan with contrast medium: on the right atelectasis of middle and lower lobes with multiple, gross, cyst-like bronchiectasis.

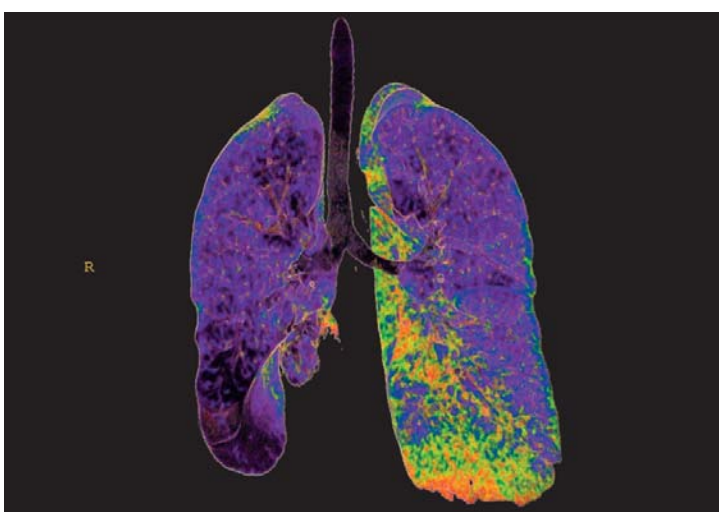

Fig. 3. - Bronchography-like volume rendering reconstructions: on the right complete obstruction of intermediate bronchus with absent ventilation of middle and lower lobes and hyperinflation of upper lobe.

mm, 120 Kv and 180 mAs. Data on the acquired volume were transferred to a dedicated workstation where, beside the axial images, multiplanar (MPR), maximum intensity projection (MIP) and volume rendering (VR) reconstructions were visualised. Axial and coronal scans, examined by both parenchymal and mediastinal windows, showed volume reduction of middle and lower lobe characterized by the evidence of multiple bronchiectasis with fluid content and cyst-like appearance and no evident relation with tracheobronchial tree (figure 2). Bronchography-like VR reconstructions on the right side showed complete obstruction of intermediate bronchus with absent ventilation of middle and lower lobes and hyperinflation of upper lobe (figure 3).

Reconstructions of bronchial structures demonstrated no patency of middle and lower lobar bronchi. The study carried out after contrast medium infusion in arterial phase showed a vascular disorder characterised by an accessory arterial branch arising from the upper portion of thoracic aorta, at D6 level, which, after moving caudally to pulmonary hilus with a tortuous course, provided blood supply to atelectatic lung (figure 4). No anomalous venous drainage was detected.

Fiberoptic bronchoscopic examination showed stenosis of right intermediate bronchus with a 2 mm wide residual lumen. Various bronchial biopsies led to the identification of fibrous tissue with chronic inflammatory signs. Cytology of sputum as well as direct, cultural and PCR search of M. tuberculosis were negative. Echocardiography showed a normal cardiac function with reduced excursions of cava vein during inspiration, due to compression by the mass. The pulmonary scintigraphic examination showed the lack of perfusion in the right lower lobe. On this basis the most likely diagnosis was bronchopulmonary sequestration. Then a right lateral thoracotomy was performed: a whitish mass was revealed in the lower portion of right hemithorax, supplied by an aberrant artery 6 mm in diameter originating from the aorta. This aberrant artery was ligated and truncated, and bilobectomy of middle and lower right lobe was performed.

Pathology of the resected lesion showed severe inflammation, as well as many bronchus-like dilated spaces lined by columnar epithelium and filled by purulent secretion (figure 5). The aberrant artery supplied the lesion and no communication with the pulmonary arterial system was found. Further clinical examination was uneventful.

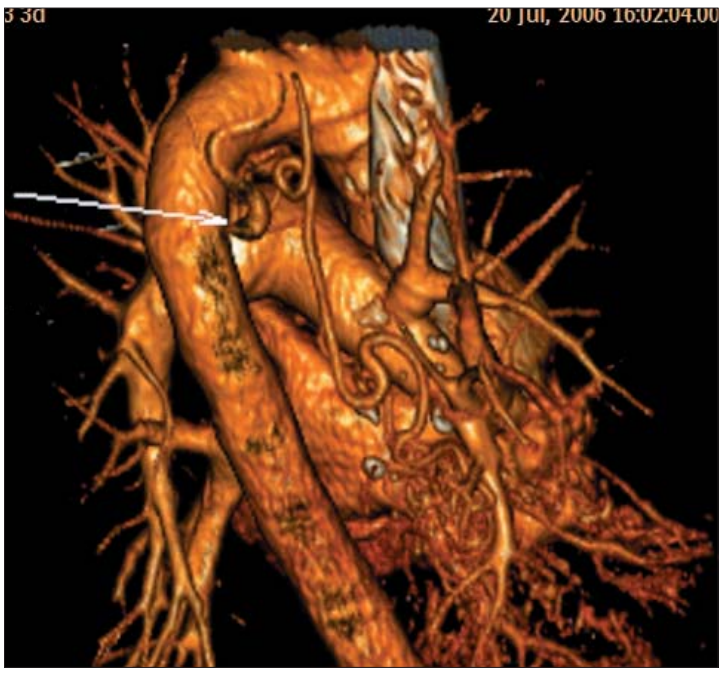

Fig. 4 - Angiography-like volume rendering reconstructions: accessory artery originating from descending aorta and moving to pulmonary hilus along a tortuous course to supply the sequestration.

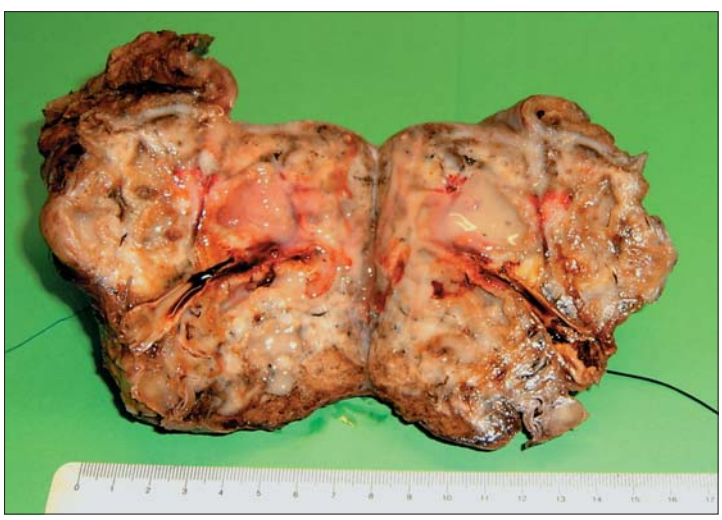

Fig. 5. - Macroscopic appearance of surgical specimen.

Discussion

Bronchopulmonary sequestration is characterised by a portion of pulmonary parenchyma that lacks a normal connection with tracheobronchial tree and with an abnormal arterial blood supply [1-3]. It accounts for 0.15%-6.4% of all congenital pulmonary malformations [2, 4].

There are two types of sequestrations, namely intralobar and extralobar [5, 6] ones. Intralobar sequestrations (80%) are contained within the normal visceral pleura and surrounded by the normal lung tissue. Extralobar sequestrations (20%) show an independent visceral pleura [7].

Broncopulmonary sequestrations are characterised by systemic blood supply by one or more abnormal artery vessel [8]. Usually, the most frequent sites of origin of abnormal arteries are thoracic aorta and aortic arch [4, 5]. More rarely they arise from abdominal aorta, celiac tree, renal arteries, and coronaric vessels [5, 9, 10].

Intralobar sequestrations show an equal gender distribution and often are diagnosed between the second and fourth decade of life [7]. Conversely the extralobar ones occur most frequently in males (75%-80%) and are usually diagnosed in infancy or childhood [5].

Most patients with pulmonary sequestration are asymptomatic for many years and the disease is often discovered accidentally [2].

Diagnostic approach to bronchopulmonary sequestration is aimed firstly at defining the presence of a sequestered lung mass, excluding alternative interpretations, and secondly at demonstrating the aberrant blood supply [11]. The latter objective is particularly important in the planning of operative approach to the lesion. In the past, imaging strategies for sequestrations included mainly bronchography and arteriography: the first procedure has long been abandoned for safety reasons, while invasive arteriography has represented the reference preoperative procedure [12, 13] since it allows the detection of the abnormal arterial supply [12, 14]. The advantage of angiography is that it provides detailed characteristic of abnormal artery (origin, course, diameter, tortuousness). The limitations of the method are poor display of topographic relationships with other anatomic structures, intrinsic invasions, risk of bleeding, of dissection, of embolism, pain in the injection site, risk of reaction to contrast medium. The intrinsic cost of the procedure is high while dedicated equipment and specifically trained personnel are necessary. Because of these limitations angiography is now restricted to patients in whom less invasive methods have failed to confirm the diagnosis [14].

More recently, less invasive or non-invasive alternatives have been proposed, including ultrasonography, magnetic resonance imaging (MRI) and helical CT imaging [15] that may be included into a rational diagnostic algorhythm (figure 6). Ultrasonographic imaging is ideally suited for evaluating the prenatal and perinatal thoracic malformations [16, 17]. Some authors have advocated its use as a screening method for sequestrations after chest radiography also in adult patients [15]. Assessment of aberrant feeding artery may be possible; colour and doppler ultrasonography may detect the pulsatile flow of the anomalous vessel [17]. The limitations of this imaging method are interposition of bony thorax, aerated lung parenchyma, or air containing cysts. Therefore the yield of this technique is closely related to the presence of consolidation providing the necessary acoustic window [15]. In addition the sonographic appearance of sequestrations is non specific and

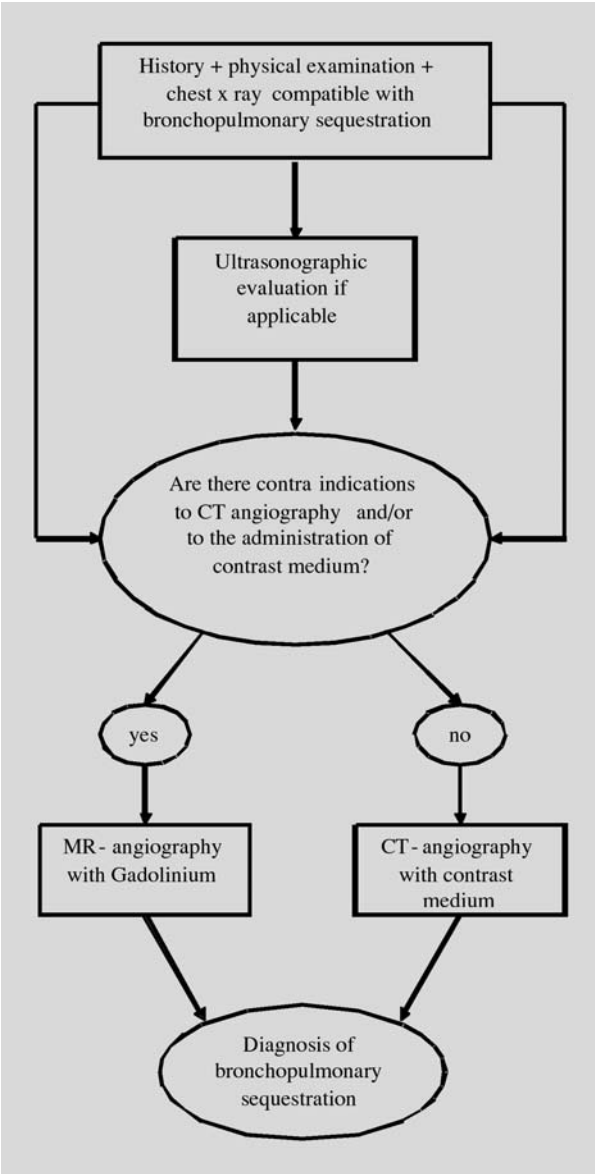

Fig. 6. - A rationale algorhythm for diagnosis of bronchopulmonary sequestration.

differential diagnosis with other causes of diaphragmatic and mediastinal masses must be made. The method does not allow to recognise minor aberrant systemic arteries that characterize the smaller lesions [18]. Therefore a trial of ultrasonography may be proposed as an intermediate diagnostic step in selected cases and always preceding a more reliable and comprehensive imaging evaluation such as MRI or CT.

MRI is a good diagnostic alternative for patients in whom angiography is contraindicated [19]. On MRI bronchopulmonary sequestration appears as a pulmonary segment hyperintense on both T1 weighted and T2 weighted images [20]. The MRI study obtained after a bolus injection of Gadolinium or through dedicated sequences allows the evaluation of thoracic aorta and pulmonary vessels. It defines the size, origin, and course of both the aberrant systemic artery and the venous drainage. The advantages of MRI are multiplanar and three-dimensional images, and the safety due to absence of exposure to ionizing radiations [20]. Notable limitations of MRI are the low spatial resolution in the vessels with turbulent flow, the inability to discriminate focal thin-walled cysts or the emphysematous changes of sequestration, the impact of respiratory artefacts [15, 20], the need of excluding patients with metallic devices and those affected by claustrophobia. Therefore the role of MRI is restricted to patients in whom an intolerance to contrast medium prevents the possibility of a CT angiography.

CT is recognised as the preferable imaging technique for pulmonary sequestrations [21, 22]: through multiplanar reconstructions [23] as it allows not only the detection of supplying vessel, but also the accurate description of heterogeneous characteristics of the mass and adjacent structures, including communications with the bronchial tree, gross lung anomalies, (eg. hypoplastic lung or emphysematous changes due to collateral air drift) and defects of the diaphragm [24]. In addition, through VR reconstructions the same set of scans allows virtual bronchographic images to be obtained, which may be helpful in the pre-operative assessment. Although implying the exposure of the patient to ionizing radiations, CT angiography compares favourably with other imaging techniques and may be considered the reference diagnostic method in all cases where administration of intravenous contrast material is not contraindicated.

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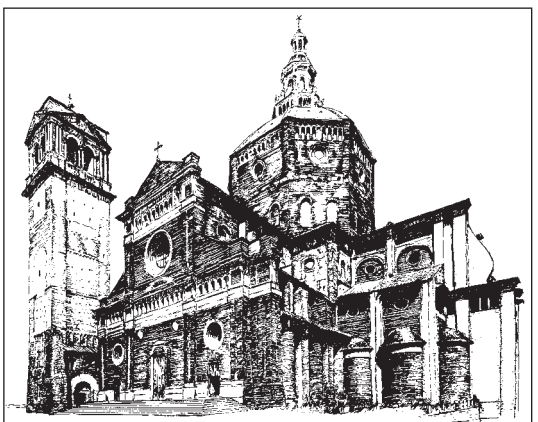

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