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CASE REPORT

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# Anthracofibrosis mimicking chronic thromboembolic pulmonary hypertension

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## Abstract

We present the case of a 78-year-old female undergoing pulmonary endarterectomy (PEA) because of suspected chronic thromboembolic pulmonary hypertension (CTEPH). During surgery firm black masses were encountered in the aortopulmonary window and on the cranial part of the right pulmonary artery (PA). After PA arteriotomy we visualized intraluminal black firm stenosing plaques at the orifices of the three right and of the left lingular and lower lobar branches. Since no dissection plane could be obtained the procedure was discontinued. Subsequent bronchoscopy visualized a submucosal dark black-blue discoloration in both main bronchi. Pathological analysis revealed anthracofibrosis, which could be explained by biomass smoke exposure in the past. We are the first to provide intravascular pictures and pathologic images of this very rare entity. Moreover, we report stenoses at the orifices of the three right-sided lobar and of the left-sided lingular and lower lobe arteries, in contrast to three previous reports that report on single locations caused by extrinsic PA compression from lymphadenopathy. Our case, however, suggests extension of fibrosis with anthracotic pigment into the PA wall. We conclude that in the absence of a clear history of exposure to carbon smoke and with consequently no diagnostic bronchoscopy, anthracofibrosis of the lungs may mimic CTEPH not only by external compression but also by extension into pulmonary vascular structures. PEA-surgery should not be attempted in these cases.

**KEYWORDS**
anthracofibrosis, anthracosis, CTEPH, pulmonary endarterectomy

## CASE-REPORT

A 70-year-old female North-African migrant with a possible history of tuberculosis presented with exertional dyspnea (New York Heart Association (NYHA) Class II) and signs of pulmonary hypertension (PH) on echocardiography. Ventilation/perfusion (V/Q)-scan showed decreased perfusion in multiple subsegmental zones and in large zones of the left lower and right upper lobes (Figure S1). At right heart catheterization pulmonary artery pressure (PAP) was 38/17/24 mmHg (systolic/diastolic/mean), cardiac index 2.25 L/min.m$^2$, wedge pressure 7 mmHg, and pulmonary vascular resistance (PVR) 4.2 Wood units (WU). Computed tomography (CT) scan showed some consolidations and adherent wall material in both pulmonary arteries (PAs), with right PA stenosis. Lifelong coumarin therapy was started. Six months later she remained in NYHA Class II with a systolic PAP of 57 mmHg on echocardiography. Chronic thromboembolic pulmonary hypertension (CTEPH) was diagnosed and pulmonary endarterectomy (PEA) was proposed, but refused by the patient.

Aged 78, she presented again with progressive dyspnea requiring supplemental oxygen therapy (NYHA Class III). Except for an increased diameter of the pulmonary trunk (from 28 to 35 mm) CT scan showed similar findings as 8 years before (Figure 1a) with bilateral mosaic attenuation of the lung parenchyma and hypertrophic bronchial arteries (Figures S2 and S3). At right heart catheterization PAP was 62/24/37 mmHg, cardiac index 2.04 L/min.m$^2$, wedge pressure 13 mmHg, and PVR 7.1 WU. Pulmonary angiography showed dilated main PAs, significant stenotic right upper and right middle lobe branches with delayed staining of multiple segmental branches (Figure 1b), delayed staining of multiple left upper lobe and of the A6 segmental lower lobe arteries, significant stenosis of the left main trunk and complete occlusion of the left lingular and lower lobe pulmonary arteries (Figure S4). Respiratory function test showed normal volumes with a trend to obstruction and a severe decrease in diffusion capacity of the lungs for carbon monoxide (44%) (Figure S5). Given her progressive clinical deterioration the patient and her family consented to PEA.

During surgery, firm black masses were encountered in the aortopulmonary window and on the cranial part of the right PA (Figure 1c). After right PA arteriotomy, we visualized intraluminal black firm stenosing plaques at the orifices of the upper, middle, and lower lobe PAs (Figure 1d). Attempts to safely create a dissection plane were unsuccessful as the firm black mass seemed to be in continuity with the mass on the external part of the arterial wall. In the left PA firm black plaques at the orifices of the lingular and lower lobar branches were seen. Multiple tissue biopsies were taken. During weaning from cardiopulmonary bypass, blood was evacuated from the endotracheal tube with simultaneous oxygenation problems. During bronchoscopy, the bleeding appeared to have stopped but submucosal dark black-blue discolorations were seen on the ventral side of both right and left main bronchi (Figure 1e).

On postoperative days 1 and 3 PAP was 56/24/35 and 60/29/39 mmHg, CI was 1.85 and 2.03, wedge pressure was 14 and 21 mmHg, and PVR 7.0 and 5.5 WU, respectively. Anticoagulant and supplemental oxygen therapies were continued, and the patient remained in NYHA Class III.

Histopathological analysis of the tissue outside the PA showed densely ordered fibrous connective tissue with the presence of histiocytes and substantial deposition of anthracosilicotic pigment. Using polarized light fine silicotic fibers were detected in this connective tissue. The specimens from the inside of the PA consisted of intima, media, and adventitia. In the adventitia between the accompanying neurovascular structures, similar observations were made as seen outside the PA (Figure S6).

Postoperatively, thorough anamnesis of family members revealed a life and work time in a hardly ventilated kitchen with a cooking stove fueled by coal, for more than 30 years in the country of origin.

## DISCUSSION

Anthracosis of the lungs is black discoloration of bronchial mucosa that can occlude the bronchial lumen and is associated with bronchial fibrosis and anthracotic pigment-laden histiocytes.[1] It usually presents with a chronic course of dyspnea and/or cough in an elderly nonsmoking patient. Concurrent exposure to carbon smoke from the combustion of fuels and inorganic compounds that contain limestone and alumina-silicates has been postulated as the cause.[2,3] Bronchoscopy is the gold standard for diagnosis.[1] Pathologic findings suggest that anthracofibrosis might be caused by anthracotic lymph nodes compressing adjacent bronchi, leading to fibrosis and resulting in airway stenosis.[4–6] Anthracofibrosis causing PA stenosis or occlusion is however very rare and is, to the best of our knowledge, limited to three reported cases in the literature.[5,7,8] Extrinsic PA compression from lymphadenopathy is the suspected mechanism, but associated PH was limited to two cases.[7,8] By adding a fourth case, we are the first to provide intravascular pictures and pathologic images. Also, bronchial anthracofibrosis owing to silicosis has

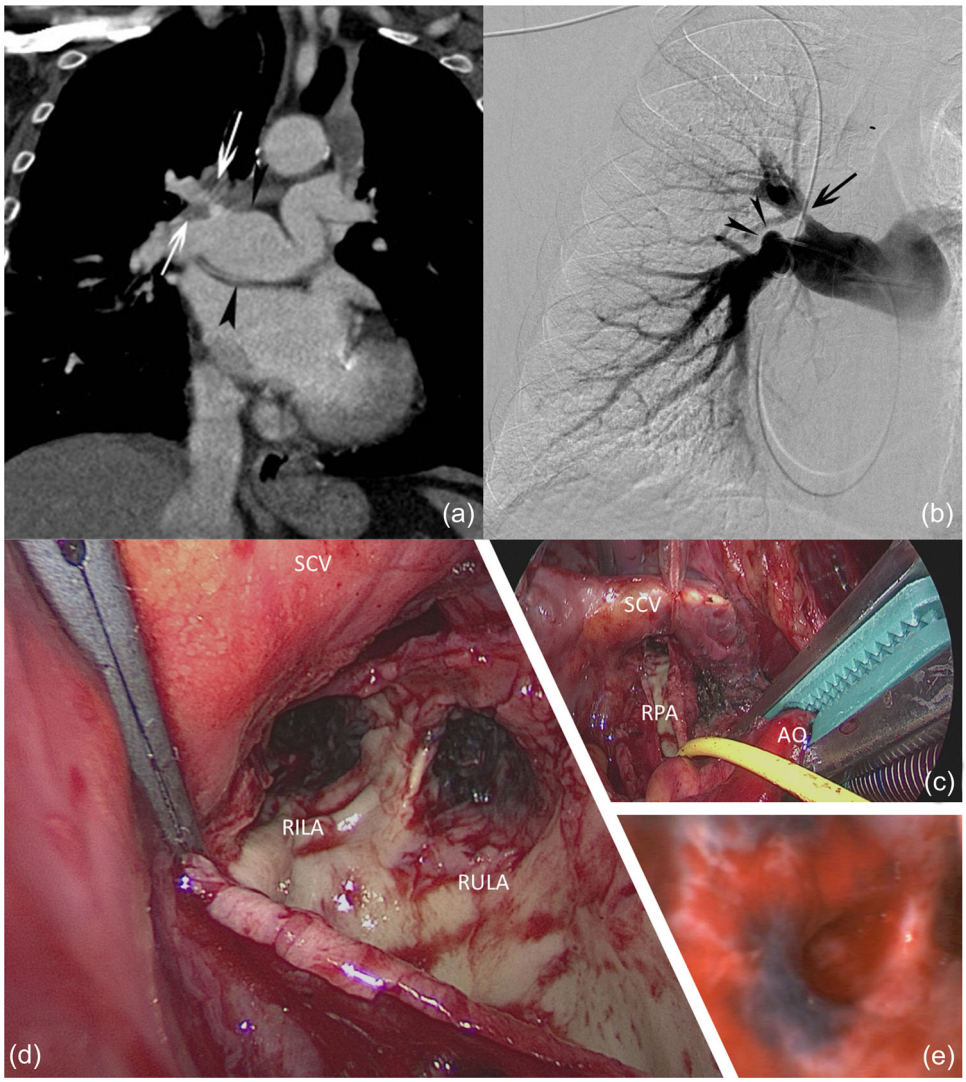


**FIGURE 1** (a) Preoperative coronal CT image of the chest shows an increased. diameter of the right pulmonary artery (black arrowheads) and stenosis of the right superior lobar pulmonary artery (white arrows) with bilateral material adherent to the vascular wall. (b) Preoperative pulmonary angiography shows a significant stenosis in the right upper lobe branch with delayed staining of multiple mediobasal segmental branches in the upper lobe and a significant stenosis of the right middle lobe branch. (c) Peroperative picture shows black firm plaques on the cranial part of the right PA. (d) Peroperative picture shows black firm stenosing intralumaninal plaques at the orifices of the upper, middle and lower lobe pulmonary artery branches. (e) Flexible bronchoscopy performed during surgery showing submucosal dark black-blue discoloration on the ventral side of the right main bronchus. AO, aorta; CT, computed tomography; RILA, right interlobar artery; RPA, right pulmonary artery; RULA, right upper lobe artery; SCV, superior caval vein.

been described.[7] Our case, however, suggests not only extrinsic compression but also extension of fibrosis with anthracotic pigment into the PA wall as the anthracosilicotic pigment was found in the adventitia. The location of anthracotic material at the inlet of lobar arteries coincides with the reported most frequently involved right upper and middle lobe bronchial inlets.[1,9] In contrast to previous reports in which anthracosis was limited to one location in the PA tree, we report stenoses at the origins of the three right-sided lobar arteries and occlusions the left-sided lingular and lower lobar artery.

Over the 8-year interval, there was no clear progression of the intravascular lesions visible on imaging modalities. Symptoms, however, worsened and hemodynamics dramatically deteriorated. We speculate this suggests occurrence of distal microvasculopathy like in CTEPH patients, and therefore common pathophysiological pathways and rationale for medical therapy.

Conditions that may mimic CTEPH on imaging tests and all-cause abnormal V/Q-scans are in situ thrombosis, pulmonary artery sarcoma or other malignancies, fibrosing mediastinitis, pulmonary vein stenosis or occlusion, pulmonary vasculitis, sarcoidosis, and congenital anomalies of

the pulmonary arteries. All have however their distinct radiological features on CT to differentiate them from CTEPH.[10] With this report we add anthracofibrosis to the list of CTEPH mimickers. The mediastinal and hilar lymphadenopathy on CT-scan and the presumed history of tuberculosis, which is the most common disease associated with bronchial anthracofibrosis, were, in retrospect, limited directions for anthracosis.[1] The key imaging findings of anthracofibrosis on CT are multifocal non-continuous stenosis of the bronchial tree, calcified enlarged mediastinal or hilar lymph nodes, and secondary lung parenchymal changes that can differentiate it from more common conditions such as TB and bronchogenic carcinoma.[11] However, as this patient at primary presentation did not report exposure to carbon smoke and already showed signs of PH with adherent wall material in the PAs, the consolidations on CT were erroneously diagnosed as residual lesions from previous TB infection and the other findings diagnosed as CTEPH. Bronchoscopy is not routinely performed in the diagnostic work-up of CTEPH.[12]

In retrospect, surgery, although providing us biopsies and pictures, had no curative consequences. Creating a dissection plane was impossible and resection of the firm black masses would have resulted in devastating damage to the PA at the level of the lobar junctions. A pulmonary artery bypass graft has been described very rarely and only in cases of Takayasu arteritis of for a neoplasm, bypassing the main pulmonary arteries.[13,14] In our case however, given the location of the disease, there would have been the need to place the distal anastomosis on the lobar arteries or even beyond. This would require extensive dissection to separate the pulmonary parenchyma from the blood vessels. The final diagnosis of anthracofibrosis was only made after surgery by pathological examination. Therefore the option to perform a pulmonary artery bypass graft was not considered during surgery because: (1) a lack of experience with this technique in our team, (2) the uncertainty of the diagnosis, and (3) the required distal location and the number of presumed distal anastomoses (at least 4 required bypasses: 2 in the right lung, 2 in the left lung). Moreover, as the imaging remained stable over the course of 8 years but the resistance increased, also the effect of pulmonary artery bypass surgery would be uncertain. Hence our (unconfirmed) speculation was that the increased resistance was caused by distal vasculopathy.

Stenting of the pulmonary arteries after the surprising surgical findings would also have been an option, although this option could cause damage to the pulmonary arterial wall because of the very firm appearance of the intravascular black masses. However, after surgery, the patient refused any additional treatment, and therefore this option was not further explored.

We conclude that in the absence of a clear history of exposure to carbon smoke and with consequently no diagnostic bronchoscopy, anthracofibrosis of the lungs may mimic CTEPH not only by external compression but also by extension into pulmonary vascular structures. Key imaging findings of this entity on CT should alert clinicians. PEA-surgery should not be attempted in these cases.

## AUTHOR CONTRIBUTIONS

Silke Van Genechten assisted during surgery and wrote the first draft of the manuscript. Bart Meyns gave advise during surgery, participated in the multidisciplinary meeting discussing this patients' case, and reviewed and revised the manuscript. Laurent Godinas participated in the multidisciplinary meeting discussing this patients' case, was involved in the postoperative care and reviewed and revised the manuscript. Geert Maleux conducted all imaging modalities, participated in the multidisciplinary meeting discussing this patients' case, and reviewed and revised the manuscript. Stephanie Everaerts performed the bronchoscopy, suggested the diagnosis of anthracosis, and reviewed and revised the manuscript. Dieter Van Beersel performed anesthesia and immediate postoperative care of the patient, and reviewed and revised the manuscript. Catharina Belge diagnosed the patient with CTEPH, participated in the multidisciplinary meeting discussing this patients' case, was involved in the postoperative care and reviewed and revised the manuscript. Birgit Weynand performed the anatomopathological research, confirmed the diagnosis of anthracosilicosis with extension into the adventitia, provided pathological images and reviewed and revised the manuscript. Marion Delcroix participated and coordinated the multidisciplinary meeting discussing this patients' case, was involved in the postoperative care and reviewed and revised the manuscript. Tom Verbelen participated in the multidisciplinary meeting discussing this patients' case, performed the surgery, contributed to the second draft of the manuscript, and critically reviewed and revised the manuscript.

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## CONFLICTS OF INTEREST STATEMENT

The authors declare no conflicts of interest.

## DATA AVAILABILITY STATEMENT

The data that supports the findings of this study are available in the supplementary material of this article.

## ETHICS STATEMENT

Written consent was delivered to publish patient information and images by the patient and her children.

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## SUPPORTING INFORMATION

Additional supporting information can be found online in the Supporting Information section at the end of this article.

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