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Endovascular Treatment of Aberrant Systemic Arterial Supply to Normal Basilar Segments of the Right Lower Lobe: Case Report and Review of the Literature

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Abstract

We report the case of a 17-year-old man with acute chest pain due to a partial thrombosis of a pseudosequestration. Unlike a true sequestration, there was a normal bronchial distribution and the involved lung parenchyma was normal on CT scan. A therapeutic transarterial embolization of the aberrant systemic artery from the proximal abdominal aorta was performed successfully. The patient did not suffer from further chest pain during the follow-up of 12 months. A contrast-enhanced CT scan 4 months later demonstrated complete occlusion of the embolized aberrant artery. Our case represents an alternative treatment to surgery for this rare abnormality.

Key words: Cardiovascular system—Malformation—CT scan—Transarterial embolization

Pulmonary systemic arterialization without sequestration is the rarest form of congenital anomalous systemic arterial supply to the lungs [1–4]. In this rare abnormality, the arterial supply of one or more of the basal segments of the lower lobe comes from an aberrant artery arising from the aorta. The nonsequestrated lung parenchyma supplied by this aberrant artery has no other bronchial or arterial structural defect. Thoracotomy with resection of involved lung parenchyma is the usual treatment of choice. However, endovascular treatment could be an interesting alternative to surgery, as discussed in our case.

Case Report

A 17-year-old man was admitted to our institution in April 2000 with a new episode of an acute chest pain. It was known that he had a right basal asymptomatic variant of sequestration discovered fortuitously in 1995 on a chest radiograph. Past medical history included, in 1995, a clinical examination that showed only a right basal murmur, a contrast-enhanced CT scan, an angiogram (Fig. 1) and a transthoracic echocardiogram that confirmed the diagnosis. The CT scan showed an aneurysmal artery arising from the aorta above the cliac trunk supplying the basal segments of the right lower lobe without any parenchymal abnormality. Transthoracic echocardiography showed left ventricular dilation and hyperkinesia. Later, in March 1997, the patient had acute chest pain related to a partial thrombosis of the aberrant artery aneurysm as demonstrated by a contrast-enhanced CT scan and a probable lung infarction.

In April 2000, after a new episode of chest pain, the patient underwent a chest radiograph and CT scan. The clinical examination showed that the right basal murmur had disappeared. The chest radiograph showed a right basal opacity which corresponded to the pseudosequestration. The CT scan again showed the aneurysm of the aberrant artery, which was partially thrombosed (Fig. 2). There was neither associated mass effect nor structural change of the pulmonary parenchyma. There were normal pulmonary arteries and bronchial anatomy in the segments involved by the abnormality as demonstrated by pulmonary angiography and the CT scan.

We decided to treat this patient because he was symptomatic with recurrent chest pain and had poor cardiac tolerance of the left-to-left shunt. We proposed a transarterial embolization, which is less invasive than surgery. Selective angiography of the aberrant artery with a 5 Fr Cobra Catheter (Cordis Europa) showed a left-to-left shunt. It demonstrated the hypertrophied vessels arising from this aberrant artery and supplying the right basal segments. Venous drainage was into the left atrium by the right inferior pulmonary vein. The left-to-left shunt was considered significant owing to the results of the echocardiography. The aberrant artery was then embolized using various coils (William Cook Europe; 20 coils, diameter range 4–15 mm). The postembolization arteriogram showed complete occlusion of the feeding artery beyond the coils (Fig. 3). After the procedure, the patient suffered from a postembolization syndrome characterized by pain and fever that lasted 3 days and was treated conservatively by analgesics. Those symptoms can be explained by a pulmonary infarction following the vaso-occlusion. There was no other complication of the procedure. There was no change on the chest radiograph apart from the coils at the base of the right lower lobe. The patient was discharged 4 days after the procedure.

A follow-up dynamic CT scan 4 months later demonstrated complete occlusion of the aberrant artery by the coils (Fig. 4). Seven months later, the patient remained asymptomatic and echocardiography showed a decrease in the left ventricular dilation and hyperkinesia. The last follow-up at 12 months confirmed the patient is free from symptoms and there was normalization of measurements on echocardiography in terms of left ventricular end-diastolic diameter and left ventricular function expressed as the ejection

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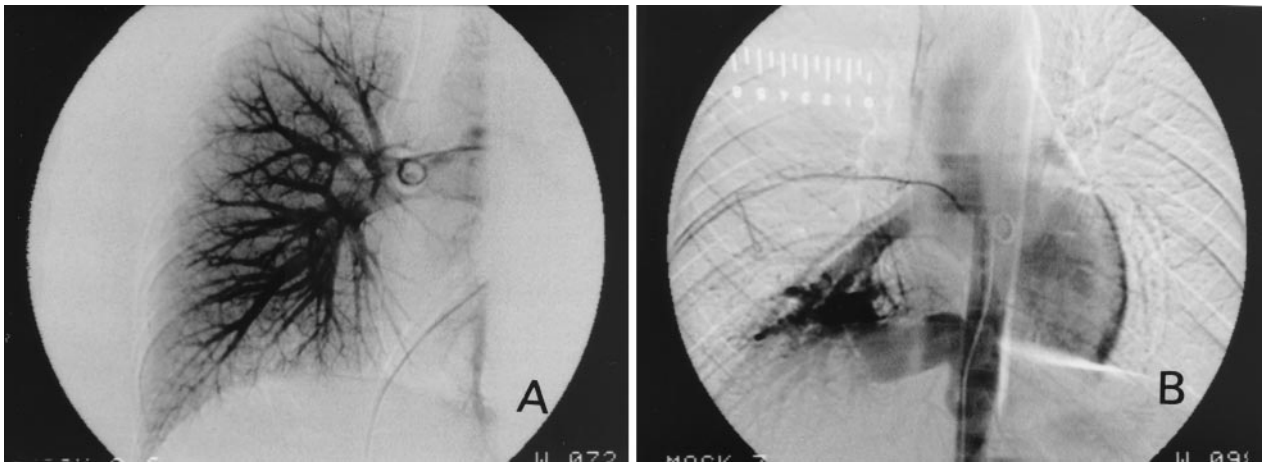

Fig. 1. **A** Anterior angiogram of the right pulmonary artery showing a normal distribution in the basal segments. **B** Anterior aortic angiogram showing an aneurysmal regular feeding artery arising from the upper abdominal aorta and venous drainage in the inferior right pulmonary vein.

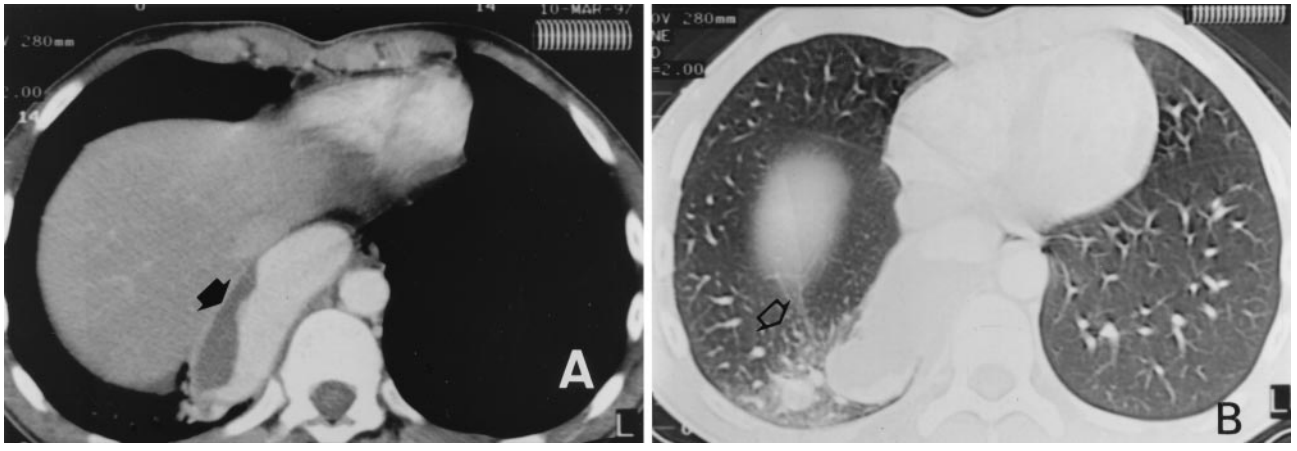

Fig. 2. **A** A 3 mm collimated contrast-enhanced CT image identifies an aneurysmal aberrant artery arising from the abdominal aorta above the celiac trunk and supplying the right lower lobe. This vascular malformation is partially thrombosed (arrow). **B** The parenchyma around the lesion appears normal apart from a limited area of ground-glass attenuation probably related to local parenchymal hemorrhage (arrow).

fraction. We decided to recommend regular lifetime follow-up owing to the age of the patient and the type of treatment used, and to depict any delayed aberrant artery recanalization.

Discussion

Pulmonary lesions with systemic arterial supply are not all sequestrations [1, 3]. Other congenital causes are arteriovenous fistula [5], pulmonary artery aplasia [6], and systemic arterial supply to normal lung [1, 6]. Acquired causes of systemic arterial supply to the lung include pulmonary suppuration [6], pleurectomy [6] and iatrogenic anastomosis [7]. Systemic arterialization of lung without sequestration corresponds to a variant of sequestration as described by Felker et al. [1]. The aberrant systemic artery may either be the only arterial supply to the lung [3], or may provide partial arterial supply in addition to normal pulmonary arterial supply, as in our case. Although the etiology of a systemic arterial supply to normal lung is unknown, the most likely theory is a persistent embryonic connection between aorta and pulmonary parenchyma [3].

Pulmonary sequestration is a rare congenital malformation characterized by a mass of nonfunctioning lung tissue separated from the normal bronchopulmonary tree and vascularized by an aberrant systemic artery [8–10]. It represents between 1% and 6% of all pulmonary malformations [11]. Two types of pulmonary sequestration are recognized depending on whether or not the malformation possesses its own pleural covering: intralobar sequestration is an abnormal region within the normal pulmonary parenchyma without its own pleural covering; extralobar sequestration corresponds to a true accessory lung, with its own pleural envelope.

Associated anomalies have not been described in systemic arterialization of lung, although intralobar and extralobar sequestration are associated with other congenital malformations in about 15% and 60% of cases respectively [11, 12]. Patients with systemic arterial supply may be totally asymptomatic [3]. The condition is usually asymptomatic in children but may be discovered following the incidental finding of a continuous chest murmur. However, left ventricular failure may develop from the left-to-left shunt. In adults,

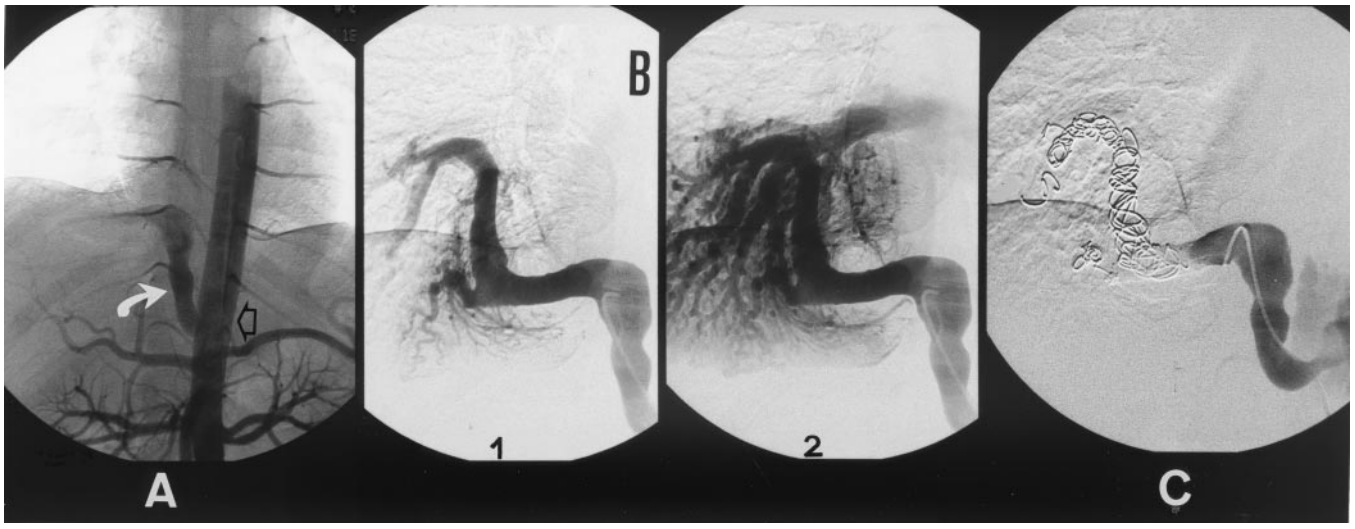

Fig. 3. **A** Aortic arteriogram showing an aneurysmal irregular aberrant artery arising from the aorta (open arrow) above the celiac trunk and supplying the right basal segments. The irregular margins (white arrow) are due to the partial thrombosis (compare with Fig. 1A). **B** Selective arteriogram of the aberrant artery showing hypervascularity (arterial phase: B1) and venous drainage by the inferior right pulmonary vein (venous phase: B2). **C** The postembolization arteriogram shows complete occlusion of the distal artery beyond the coils.

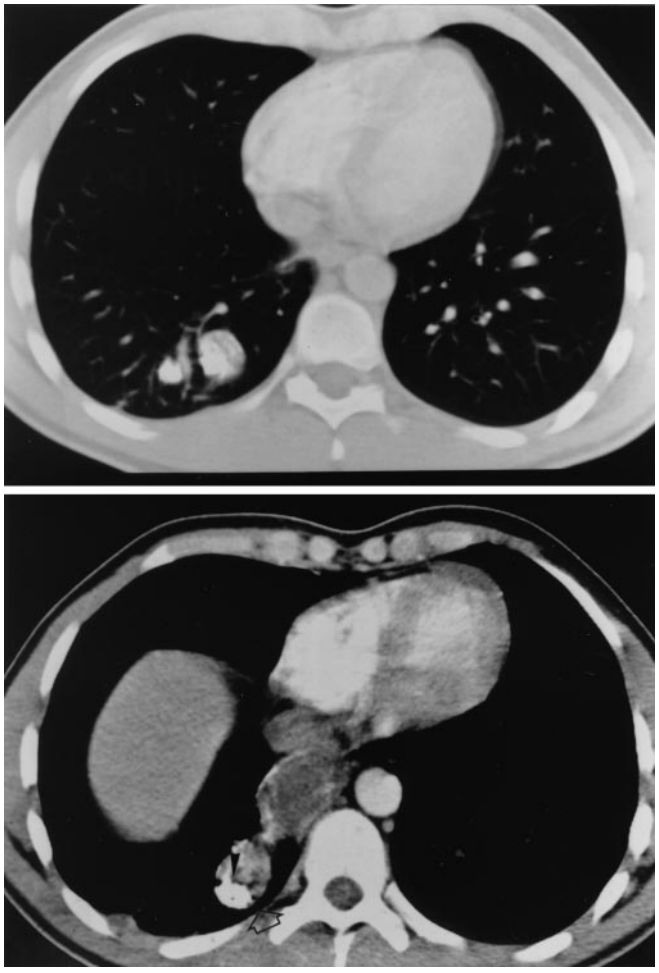

Fig. 4. A follow-up contrast-enhanced CT examination at 4 months demonstrates complete occlusion of the feeding artery (arrow) without parenchymal complication. Coils appear as an area of high attenuation (arrowhead).

it may also be asymptomatic or present as an incidental murmur or chest film abnormality. Massive hemoptysis is an unusual manifestation [13]. Treatment is recommended in symptomatic patients but also in asymptomatic patients owing to the risk of hemorrhagic complications.

The mainstay of treatment has always been surgical excision [12, 13]. It is believed by most authors that bronchopulmonary sequestration should be treated by ligation of the feeding artery and resection of the sequestered lung parenchyma because of its propensity for recurrent infections [11, 14]. In the absence of sequestered tissue, as in this case, ligation of the systemic vessel only may be sufficient [15, 16]. Kirks et al. [3] described two cases of systemic arterial supply to normal lung treated by surgery with ligation of the aberrant artery and lobectomy.

Transarterial embolization is well established in the management of hemoptysis [16]. Hayakawa et al. [17] and Gasparini et al. [18] have performed transarterial embolization in patients with massive hemoptysis due to sequestration; they obtained a partial result with gelatin sponge alone and a complete result with a combination of fibrin sponge and sclerosing agents. Only two cases of therapeutic embolization by coils of a systemic arterialization of normal lung have been described in the literature [1, 12]. In these two cases, there was no major complication and surgery was avoided.

Overall our case demonstrates the benefit of transarterial embolization in this rare abnormality: the treatment was selective and effective with a sufficient follow-up. Moreover this technique is noninvasive compared with surgery. There was no complication and the recovery time was short. Coils are probably the most effective method of occluding the aneurysmal artery, but in the presence of a smaller feeding artery, balloon embolotherapy could also be effective.

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