Introduction

Systemic arterial supply to the lung parenchyma is demonstrated in several congenital and acquired diseases [1, 2], but the systemic arterial supply to normal lung parenchyma with regular bronchial tree, without any congenital heart and lung diseases is a rare disorder [1, 3]. Most of the patients with systemic arterial supply to the normal lung parenchyma have no respiratory symptoms and can be managed conservatively [3]. If clinical symptoms such as hemoptysis and congestive heart failure are present, operative methods must be concerned. Trans-arterial coil embolization of anomalous artery can improve symptoms and may bring about a satisfactory solution for this disease as a minimal invasive technique.

Case Report

A 20-year-old man was admitted to our hospital with hemoptysis lasting for 3 years. The laboratory tests and pulmonary function tests were normal. His physical examination revealed no significant abnormality. Echocardiography showed mild mitral valve regurgitation but left ventricular function was normal.

Posteroanterior chest X-ray showed increased pulmonary vascular opacities in the left lower lung field (Fig. 1). There was no evidence of pulmonary dysplasia. Computed tomography (CT) (Aquillion 64; Toshiba Medical Systems, Tokyo, Japan) revealed a 10-mm anomalous artery arising from the anterolateral aspect of the descending aorta at the level of T9 vertebra (Fig. 2). The artery was supplying the left lower lobe segments 7–10. The left lower lobe was diffusely high attenuated
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except superior segment because of hyperemia and congestion due to high pressure systemic supply to the lung. The vessels of the involved area were hypertrophied in comparison with those of the contralateral lung. There was a focal ground-glass area in the posterior basal segment that can resemble alveolar hemorrhage (Fig. 3). Left lower lobe pulmonary artery was small, and its branches were not distributed in the affected segments (Fig. 3). CT scan showed a normal bronchial tree ruling out lobar sequestration.

Angiography (Toshiba, Infinix, Japan) of descending thoracic aorta revealed anomalous artery coursing towards the left lung. Selective catheterization of the artery revealed that it was supplying nearly the whole left lower lobe. Late phase showed the pulmonary venous drainage via inferior pulmonary vein to the left atrium (Fig. 4). There was no direct communication between the abnormal systemic artery and the veins of the involved lung tissue. Pulmonary artery angiogram showed normal right and upper left pulmonary arteries. However, the inferior lobar branch of the left pulmonary artery was hypoplastic.

After inserting a 7-F short sheath introducer, a 7-F guiding catheter (Mach 1, Boston Scientific, Fremont, CA), via a 0.035-inch stiff guide wire (Amplatz TFE coated, William Cook Europe Aps, Bjaeverskov, Denmark), was advanced into the abnormal artery. A 0.035-inch embolization coil of 10 mm diameter and 8 cm long (MReye, William Cook Europe Aps, Bjaeverskov, Denmark) was delivered, and a safe and stable position was obtained. Two 0.035-inch coils (of 5 mm diameter — 8 cm long and of 5 mm diameter — 5 cm long, both MReye, William Cook Europe Aps, Bjaeverskov, Denmark) were released afterwards, and a firm coil package was obtained. Control angiogram showed complete occlusion of the artery (Fig. 5).

The post-procedure course was uneventful. The patient was discharged on the second day after the pro-

Fig. 2. | CT images of the aberrant artery and the left lower lobe. A: Volume-rendered CT scan of oblique coronal view of thorax shows the aberrant systemic artery (arrow) arising from anterolateral aspect of distal thoracic aorta. B: Coronal multiplanar reformatted CT image shows prominent increased vascularity in the left lower lobe comparing with the right side. C: Coronal multiplanar reformatted CT image shows decreased vascularity 1 month after coil embolization in the same region comparing with Fig. 2B

Fig. 3. | Axial CT images of the left lower lobe. A: A CT image obtained before coil embolization shows dilated pulmonary vessels in the left lower lobe and focal ground-glass density (arrow) in the posterobasal segment which can resemble alveolar hemorrhage. B: Focal ground-glass density totally disappeared and pulmonary vascularity significantly decreased becoming nearly similar to those of the right lung after coil embolization
There was no evidence of hemoptysis for 18 months after embolization. Aortic angiogram at 1 and 10 months after the embolization showed complete occlusion of collateral artery. One month CT revealed that the hypertrophied vessels were diminished and hyperattenuation of the parenchyma was significantly reduced due to marked improvement of pulmonary congestion. No parenchymal lesion was identified that may resemble pulmonary infarct. Left lower lobe pulmonary artery branches remained unchanged. Dilatation of the bronchial arteries was not observed.

Discussion

Systemic arterial supply to the lung parenchyma was categorized by Do et al. [1] in two groups as: (a) hypertrophied normal systemic arteries (b) aberrant systemic arteries. Chronic inflammatory lung diseases, chronic pulmonary artery obstruction, and pulmonary arteriovenous malformations often lead to formation of hypertrophic systemic arteries because of chronic hypoxia in adults. Many congenital heart and lung diseases can be described in the second group and usually diagnosed during early childhood [2, 4]. Decreased pulmonary blood flow and hypoxia in the early fetal life can act as the stimuli for the persistence of aberrant systemic arteries in the case of congenital heart diseases, bronchopulmonary dysplasia and some other congenital abnormalities [4]. The aortopulmonary collaterals in congenital heart diseases rarely need intervention because they usually regress after the treatment of primary disease [4]. Bronchopulmonary sequestration and congenital venolobar syndrome (scimitar syndrome) can also cause aberrant systemic arteries [1, 2]. There are helpful diagnostic features which facilitate the differentiation between bronchopulmonary sequestration, scimitar syndrome, and normal lung with systemic arterial supply [1]. Bronchopulmonary sequestration is characterized by a non-functioning lung area, with abnormal obstructed tracheobronchial tree. An anomalous systemic artery typically supplies the abnormal lung tissue [5, 6]. On CT, there is air or fluid containing cyst and soft tissue masses [3]. Scimitar syndrome is a rare condition presenting with severe congestive heart failure and pulmonary hypertension. The presence of large systemic-pulmonary collateral arteries may play a role to cause of these symptoms. Typically there is abnormal pulmonary venous drainage (scimitar vein) into the right atrium. Lung and pulmonary artery hypoplasia and bronchial tree abnormalities can also accompany the disease to some extent [7]. In our case, different from bronchopulmonary sequestration, the lung area that was supplied by the aberrant systemic artery had a normal connection to the bronchial tree, there were no characteristic CT findings of sequestrated abnormal lung tissue and there

Fig. 4. Digital subtraction angiography images before coil embolization. A: The aberrant artery selectively catheterized via thoracic aorta is shown to supply blood to the basal segments of the left lung. B: Selective left pulmonary artery angiogram shows normal left upper pulmonary arteries but the inferior lobar branch is significantly reduced in caliber. C: Late phase image of the aberrant artery angiogram shows the pulmonary venous drainage via inferior pulmonary vein to the left atrium. There was no direct communication between the aberrant artery and the veins of the involved lung tissue.

Fig. 5. Digital subtraction angiography images after coil embolization. A: Selective aberrant artery angiogram shows complete occlusion of the artery and cessation of the arterial blood flow to the basal segments. B: Selective left pulmonary artery angiogram shows the branches remained unchanged indicating hypoplasia.
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were no recurrent pulmonary infections. Normal pulmonary venous drainage and lung volume distinguish the situation from scimitar syndrome.

A systemic artery can supply a normal lung parenchyma without any congenital heart and lung diseases [1, 3]. Pryce [8] classified this entity as type 1 intralobar sequestration. Although the cause of systemic arterial supply to the normal lung is unknown, abnormal persistence of primitive aortic post branchial arches before development of pulmonary arteries may result this anomalous artery [3]. The aberrant artery commonly arises from the descending thoracic aorta and, in some cases, from the proximal abdominal aorta or the celiac artery [9]. The venous drainage is always via the normal pulmonary vein [10]. Basal segments especially in the left lung are most often involved [1, 3]. In most cases, pulmonary artery of the affected segment is absent [10, 11] but also may be normal [10]. CT is a useful technique for differential diagnosis [1, 3] and this entity also can be showed by magnetic resonance imaging and angiography [12]. Although most of the cases do not have any respiratory symptoms, some patients may present with hemoptysis, as in our case, heart murmur, and acute chest pain [3]. Pathological examination of these arteries showed that there is no muscular lamina which might explain hemoptysis [13]. Rarely, heart failure and respiratory distress caused by left-to-left shunt can be problematic [9]. Because of its potential risk for hemoptysis and heart failure, treatment is recommended also for asymptomatic patients [13]. Surgery has been the standard therapy for symptomatic patients, and lobectomy was performed in most of the cases. Other operational procedures are segmentectomy, anastomosis between the anomalous artery and pulmonary artery, and ligation of the anomalous artery [9]. Trans-arterial coil embolization may be a good alternative to standard surgery techniques as a minimal invasive intervention and first described by Brühlmann et al. [10] in 1998. Till then, 9 patients (including our patient) were reported in the English literature. Of these cases, one patient went to re-embolization because of recurrent flow [13]. Complete occlusion of the aberrant artery was achieved in the others, and no additional therapies were needed. In all cases, symptoms were improved successfully and there were no serious complications. The follow-up periods were between 6 months to 6 years. Saida et al. [14] reported an uneventful follow-up period for 6 years but other reports had relatively short follow-up periods.

We used coils as the embolic material to occlude the aberrant artery as in the other reported cases. Proximal occlusion with coils or vascular plugs should be preferred to distal arterial embolization with particular materials such as polyvinyl alcohol, because of increased risk of pulmonary ischemia. Brühlmann et al. [10] and Chabbert et al. [15] reported embolization of incomplete types of cases before. Then some cases of complete types as in our case, in which the normal pulmonary arterial supply to the affected segments is absent, have been achieved successfully. Even in complete cases, pulmonary infarcts were not detected except a tiny infarct reported by Izzillo et al. [16]. We also did not recognize any infarcts on control CT images. Mild chest pain reported after the intervention could be due to temporary pulmonary ischemia [14].

Conclusions

Trans-arterial coil embolization may be a safe and reasonable therapeutic option for systemic arteries that supply a normal lung parenchyma even in complete types. This technique – as definitive therapy – might be acceptable in those rare cases when real pulmonary sequestration can be safely excluded. In most cases, it is difficult to differentiate between congenital anomalies causing hemoptysis in young patients. Typical differential clues – normal bronchial tree, normal lung parenchyma, absence of recurrent infections in our case – must be considered in order to diagnose systemic arteries that supply a normal lung parenchyma.

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