Anomalous systemic arterial supply to the lungs has been reported in some cases of congenital heart disease, congenital lung diseases such as bronchopulmonary sequestration and hypogenetic lung syndrome, and in apparently normal heart and lung [1, 2]. The latter has been described as the rarest form of the condition [3]. The basal segments of the left lower lobe are most frequently involved, and this anomaly is therefore referred to as the systemic arterial supply to normal basal segments of the left lower lobe [3, 4]. We report a case of this anomaly, describing its characteristic radiographic and operative findings.

Case Report

A five-year-old girl whose growth and development were normal was admitted to hospital due to intermittent periumbilical pain. Physical examination was unremarkable except for the presence of a grade 3/6 continuous systolic murmur along the left lower sternal border. The murmur was first noted at 7 months of age, but no further investigation was performed. Laboratory findings were normal, though an electrocardiogram demonstrated bilateral ventricular hypertrophy.

Posteroanterior chest radiography revealed retrocardiac nodular and tubular densities, partially obliterating the descending thoracic aorta [Fig. 1A]. The radiographic findings were initially misinterpreted as pneumonia, but further careful review of these and comparison with the patient’s clinical information suggested possible systemic arterial supply to the normal lung. To confirm or refute this, chest CT scanning was performed: a contrast-enhanced spiral scan showed an anomalous systemic artery arising from the descending thoracic aorta and branching into the normal pulmonary parenchyma of the left lower lobe, and the dilated, tortuous left inferior pulmonary vein draining into the left atrium, partially obliterating the descending thoracic aorta [Figs. 1B, C]. At lung window setting, the left interlobar pulmonary artery distal to the origin of the superior seg-
mental artery was absent, but the bronchial tree was normal [Fig. 1D]. A CT scan also depicted crowding of dilated vessels, with ground-glass attenuation in the left basal lung [Fig. 1E]. Chest MR imaging was performed as preoperative work-up: a contrast-enhanced MR angiogram, like the CT image described above, showed an anomalous systemic artery arising from the descending thoracic aorta [Fig. 1F], and a dilated left inferior pulmonary vein draining into the left atrium.

At thoracostomy, an anomalous systemic artery was found to originate from the descending thoracic aorta, supplying the common basal segments of the left lower lobe via the pulmonary ligament. The veins of the left basal segments drained into the left atrium via the large inferior pulmonary vein. Two small branches of the left pulmonary artery were seen to supply the superior segment of the left lower lobe, but no branch of the left pulmonary artery supplying the basal segments was found.
A left lower lobectomy was performed, and subsequent pathologic examination showed that the bronchial trees and pulmonary parenchyma were normal.

**Discussion**

Anomalous systemic arterial supply to the normal basal segments of the lower lobe of the lung is a rare congenital abnormality, and is regarded by some authors as part of the broad spectrum of bronchopulmonary sequestration (1-5). Some, on the other hand, believe this entity is a congenital vascular anomaly that is distinct from bronchopulmonary sequestration (2). This, defined as abnormal lung tissue that has no normal continuity with the tracheobronchial tree and is supplied by an anomalous systemic artery, is of two types, intra- or extralobar, a classification which depends on its pleural covering and venous drainage system (6). In contrast, anomalous systemic arterial supply to the normal basal segments of the lower lobe of the lung has a normal connection to the bronchial tree and the normal pulmonary parenchyma (5-7). The etiology of systemic arterial supply to the normal lung is unknown, but it is thought that the persistence of an embryonic connection between the aorta and the pulmonary parenchyma leads to this anomaly (1-4, 7).

Most patients are asymptomatic, and the anomaly is discovered at chest radiography for the evaluation of cardiac murmur, as in our case. Occasionally, blood flow through the aberrant vessel is enough to result in significant volume overload of the left-sided cardiac chambers, and congestive heart failure (8).

The anomalous systemic artery supplying the basal segments of the left lower lobe and the large inferior pulmonary vein appear at chest radiography as an ill-defined area of nodular and tubular opacity, and confusion with pneumonia or pulmonary vascular malformation such as arteriovenous malformation may arise. Some authors have claimed that the origin or tortuosity of the anomalous artery is responsible for the focal obscurity of the descending aorta (2-4, 9). In our case, it was attributed to the dilated inferior pulmonary vein adjacent to the descending thoracic aorta.

Contrast-enhanced spiral CT of the chest depicted the dilated and tortuous vascular structures, with ground-glass attenuation, in the left lower lobe, and the anomalous systemic artery originated from the descending thoracic aorta. The ground-glass attenuation in the involved lung is thought to represent increased pulmonary perfusion. CT also provided information about the normal bronchial tree and pulmonary parenchyma, which was histologically confirmed. Another important
CT finding was the absence of the interlobar pulmonary artery distal to the origin of the superior segmental artery. These CT findings facilitate distinction from classic bronchopulmonary sequestration (2, 4, 6, 7).

Although MR imaging can also demonstrate an anomalous systemic artery, it is not easy to determine confidently whether the lung tissue supplied by a systemic artery is normal. We therefore believe, as reported by others (2, 4, 6, 7) that contrast-enhanced spiral CT of the chest is the mainstay for evaluation of systemic arterial supply to the normal basal segments.

If clinical symptoms such as hemoptysis and congestive heart failure are not present, treatment is conservative, though for the correction of left-to-right shunt, surgery is always imperative. An anomalous systemic artery may simply be ligated if there is pulmonary supply to the involved segments of the lung. If, on the other hand, the aberrant artery represents the sole source of blood flow, lobectomy is required (2, 5, 8). In our case, contrast-enhanced CT demonstrated that an anomalous systemic artery arising from the descending aorta provided the only blood flow to the normal basal segments of the left lower lobe, and left lower lobectomy was therefore planned and performed on the basis of the CT findings.

In conclusion, systemic arterial supply to the normal basal segments of the left lower lobe should be included in the differential diagnosis when a chest radiograph depicts retrocardiac nodular density with focal obscurity of the descending thoracic aorta. We believe that contrast-enhanced CT is indispensable for correct diagnosis and proper treatment.

References
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