Extralobar Pulmonary Sequestration Presenting as an Anterior Mediastinal Tumor in an Adult*

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Extralobar pulmonary sequestration (EPS) is a rather uncommon congenital anomaly. Most patients are diagnosed in their early life or during the first decade because of the early appearance of symptoms, including feeding difficulty, cyanosis, and dyspnea, or because of symptoms arising from the associated congenital abnormalities. Extralobar pulmonary sequestration is more often found between the lower lobe and the diaphragm and is usually associated with other congenital abnormalities, but the case reported herein differs in these respects. In this report, an incidental finding of EPS is described in a 30-year-old Chinese man, where EPS presents as an anterior mediastinal mass roentgenographically. The mass was attached to the right suprahilar region by a fibrovascular pedicle which contained a small elastic artery, veins, and nerve bundles.

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| EPS = extralobar pulmonary sequestration; IPS = intralobar pulmonary sequestration; PS = pulmonary sequestration |

Pulmonary sequestration (PS) is an uncommon congenital anomaly consisting of a mass of dysplastic lung tissue that does not have a normal connection with the tracheobronchial tree and the pulmonary vascular supply. A constant feature of this abnormality is an anomalous systemic blood supply to the sequestrated segment from the thoracic aorta or abdominal aorta, often through the celiac axis.

There are two types of PS. Intralobar pulmonary sequestration (IPS) is contained within the visceral pleura of a pulmonary lobe, and venous drainage is usually through the pulmonary venous system. Extralobar pulmonary sequestration (EPS) is contained within its own pleura, and venous drainage is usually through the systemic venous system.

Intralobar pulmonary sequestration is almost always located posterobasally in the lower lobe, with a left-side preponderance; cases in the upper lobe are rare. Extralobar pulmonary sequestration is more often found between the lower lobe and the diaphragm but may be located in a variety of locations, including within the substance of the diaphragm, within the lung, in the pleural or pericardial space, or in the retroperitoneum. Occasionally, EPS may be located in the mediastinum, particularly in the posterior mediastinum. Extralobar pulmonary sequestration is rarely found in the anterior mediastinum.

The diagnosis of PS before surgery relies upon the identification of an abnormal systemic vessel supplying the PS. This vessel has usually been identified by arteriography, but more recently by other modalities, such as computed tomography (CT) and ultrasound.

The case presented herein is an incidental finding of EPS presenting as an anterior mediastinal mass roentgenographically in a male adult. Because of its unusual location and the failure of thoracic CT to demonstrate the aberrant arterial supply, EPS was not diagnosed before surgery.

CASE REPORT

A 30-year-old man was referred to our hospital because of an incidental finding of an anterior mediastinal mass on his chest roentgenograms obtained for a physical checkup in August 1991. The patient had been rather well in the past. The physical examination at the time of admission did not disclose any abnormality. A posteroanterior chest x-ray film revealed a mass located in the anterior mediastinum (Fig 1). Bronchoscopic findings were normal. A thyroid scan showed no abnormal uptake in the mediastinum. Thoracic CT revealed an anterior mediastinal mass located in the retrocaval region (Fig 2). After injection of contrast medium, the mass showed moderate enhancement. No aberrant arterial supply could be demonstrated (Fig 3). With the impression of an anterior mediastinal mass, probably a thymoma or teratoma, the

![Figure 1. Posteroanterior chest x-ray film evidences anterior mediastinal tumor.](http://example.com/figure1)

![Figure 2. Thoracic CT shows retrocaval mass with rather homogeneous density.](http://example.com/figure2)
By far the majority of PSs are located in the lower lobe. Like IPS, EPS is usually found in the lower hemithorax. It is more often found between the lower lobe and the diaphragm but may also be found in a variety of locations, including within the substance of the diaphragm, in the mediastinum, within the lung, in the pleural or pericardial space, or in the retroperitoneum; however, EPS is rarely found in the anterior mediastinum. To our knowledge, only 1 case of EPS (a 3-year-old boy) presenting roentgenographically as an anterior mediastinal polycystic mass proven at autopsy has been reported. 10

Because identifying the systemic arterial supply is the critical aspect of roentgenographic diagnosis for PS, aortography has been considered essential; however, CT has been able to demonstrate systemic arteries in most cases. 4,5 In this regard, CT may be of certain value in the diagnosis and evaluation of PS; however, thoracic CT showed no benefit to our patient because no aberrant systemic artery could be shown. There is no clear explanation for why CT did not show anomalous vessels in this case. Likely explanations are the small size of the artery and an unfavorable orientation with respect to the scanning plane. In the report of Ikezoe and coworkers, 6 the aberrant vessels could not be demonstrated in 7 of 23 cases with PS.

Because of its unusual location and the failure of thoracic CT to demonstrate the small aberrant artery, EPS was not suspected or diagnosed before surgery in the case reported herein. In summary, PS is a relatively rare, albeit important, cause of pulmonary disease in the adult. Although chest roentgenography and arteriography are useful in detecting this condition, the most crucial element in diagnosis is a high index of suspicion. Based on this case, PS may be added into the list for differential diagnosis of an anterior mediastinal tumor, even in adults.

REFERENCES

10 Williams AO, Enumah FI. Extralobar pulmonary sequestration. Thorax 1968; 23:220-23