Pulmonary Vascular Sling with Aberrant Right Upper Lobe Pulmonary Artery in A Child

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In the pulmonary vascular sling (PVS), the left pulmonary artery arises from the right, passes over the right bronchus, arches posteriorly above the tracheobronchial angle, and courses to the left lung between the trachea and the esophagus. Pulmonary vascular sling is generally associated with other cardiac defects such as ventricular and atrial septal defects. A history of atelectasis, emphysema, or pneumonia of the right lung may be found.

A 10-month-old male patient was referred to our clinic because of frequent lower respiratory tract infections. He had pneumonia with respiratory distress, tachypnea, and expiratory stridor. He had a pulse rate of 140/minute, blood pressure of 80/50 mmHg, normal heart sounds and grade II/VI holosystolic murmur at the apical region. Chest radiograph revealed right paracardiac pneumonic infiltration with normal cardiac silhouette. The patient underwent transthoracic echocardiography, which demonstrated a small defect in the interventricular septum with 64 mmHg systolic gradient, but the left pulmonary artery could not be visualized at the parasternal short axis position. Though patient did not undergo bronchoscopy clinically the diagnosis of laryngomalacia was suspected. Barium esophagography showed an anterior indentation at the level of the tracheal bifurcation (Fig 1). At cardiac catheterization, right ventriculography revealed left pulmonary artery arisen from the right pulmonary artery, which then itself gave rise to the right upper lobe artery (Fig 2). The patient was treated with suitable antibiotics and was sent for correcting operation of this anomaly. Patient underwent successful surgical treatment for the relief of tracheoesophageal compression by vascular anomalies at another institution, but died postoperatively.

The most common clinical presentations in PVS are respiratory symptoms, more prominent in the expiratory phase. Our patient was treated three times because of pneumonia, generally with complaints of expiratory stridor without dysphagia. Pulmonary vascular sling may be associated with other cardiac defects in more than half of all cases. The patient also had a small ventricular septal defect. Barium esophagography is probably the most useful noninvasive diagnostic tool for diagnosing of anterior indentation. But catheterization may be necessary to clarify this anatomy. In this case angiography successfully delineated the abnormal anatomy of the pulmonary vessels. The right upper pulmonary artery was arising from the right-sided left pulmonary artery. Associated intrinsic anomalies of the tracheobronchial tree as well as cardiovascular anomalies are commonly seen. Bronchoscopic examination may be needed, and extreme care should be done during this procedure. The results of surgical repair of PVS show high mortality rates. We believe that the worst result can be expected from this type.

We conclude that barium swallow and echocardiography are important diagnostic tools but angiography must be performed if PVS is suspected. In children with recurrent lower respiratory tract diseases and expiratory stridor without dysphagia, PVS may be considered in the differential diagnosis, even if there was previous diagnosis of laryngotracheomalacia.