Bronchial Obstruction Due to Pulmonary Artery Anomalies

II. Pulmonary Artery Aneurysm

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In a previous publication attention was drawn to the occurrence of pulmonary artery anomalies as conditions causing tracheobronchial obstruction. In the present report 2 infants are presented in whom a large aneurysm of the pulmonary artery pressed upon the walls of the left bronchus, producing severe respiratory symptoms.

During the past decade vascular malformations of the superior mediastinum have been the object of intense clinical interest. However, the occurrence of anomalies of the pulmonary artery as a cause of tracheobronchial obstruction has gone almost unrecognized. We previously reported 3 cases in which significant respiratory obstruction was produced by a left pulmonary artery coursing anomalously above the right bronchus and behind the trachea, thus producing a "vascular sling." 

The present report is concerned with the description of 2 patients in whom disabling respiratory difficulties resulted from pressure on the left bronchus by a large pulmonary artery aneurysm, a hitherto undescribed condition.

Case Reports

Case I. D. M., a 3-month-old white boy was admitted to Children's Memorial Hospital because of "congenital heart disease and dextrocardia." He was the product of an uneventful gestation and delivery. At birth an imperforate anus was found with a tiny fistulous tract, terminating just lateral to the median raphe. This was repaired and subsequently dilated with satisfactory resumption of bowel function. Cyanosis, especially on crying, rapid and labored respirations, and a systolic murmur were noted shortly after birth.

On admission the infant appeared well nourished with mild cyanosis of the lips and extremities. The heart tones were best heard over the right chest and the cardiac impulse was palpable in the right anterior axillary line. A rather harsh systolic murmur was heard over the entire chest. In the second and third left intercostal space was a loud to-and-fro murmur. The left chest was hyperresonant and rales were heard at the left base. The liver edge was felt 1 inch below the right costal margin. The electrocardiogram revealed upright P waves in the standard limb leads with shift of the transition zone to the right in the chest leads. Roentgenogram (fig. 1) and fluoroscopy of the chest showed a marked emphysema of the left lung with shift to the right of the heart and mediastinal structures. In addition, a large pulsating mass was present in the region of the pulmonary artery segment. A venous angiogram revealed opacification of the left atrium from

Fig. 1. Case I. Roentgenogram in the frontal projection showing marked emphysema of the left lung with displacement of the heart and mediastinum to the right and a large mass in the region of the left pulmonary artery.
the right atrium and of the aorta from the right ventricle, and the presence of a huge aneurysm of the pulmonary artery (fig. 2).

Because of the severity of the respiratory symptoms, surgical relief was attempted. Through a left thoracotomy a huge aneurysmal dilatation of the left pulmonary artery and similar but less pronounced changes of the right pulmonary artery were found. The left bronchus was collapsed and remained so when the aneurysmal left pulmonary artery was elevated. Although it was realized that pneumonectomy in this age group is almost uniformly fatal, it was thought that the severe mediastinal shift could be corrected in no other way. The left pulmonary artery was ligated and the emphysematous left lung was removed. The infant died 10 hours later.

At autopsy a tetralogy of Fallot with a widely patent foramen ovale was found. The main pulmonary artery measured 1.5 cm. in diameter at its origin from the right ventricle; the left pulmonary artery, which was removed at surgery, 3 cm. at about 0.5 cm. from the bifurcation; the right pulmonary artery was also dilated and became bulbous at the right hilum.

Case 2. I. T., a white baby girl, born at full term of an uncomplicated pregnancy, was seen at the Children's Memorial Hospital at the age of 13 days because of rapid shallow respirations and cyanosis of the lips and fingernails. On admission the infant appeared well nourished, tachypneic, with minimal retractions of the suprasternal notch and mild cyanosis. The heart tones were best heard over the right precordium and the point of maximal pulsation was felt in the fourth right intercostal space and nipple line. A loud to-and-fro murmur with a continuous thrill was present over the second and third left intercostal space parasternally. The lungs were clear. The liver edge was felt just below the right costal margin. Roentgenogram (fig. 3) and fluoroscopy of the chest showed the heart and mediastinal structures markedly shifted to the right with a large pulsating density at the level of the pulmonary artery segment. The left lung was emphysematous. A venous angiocardiogram (fig. 4) revealed a huge aneurysmal dilatation of the main and left pulmonary artery with no definite evidence of shunt in either direction.

The infant's hospital course was marked by recurring episodes of dyspnea and cyanosis. Death occurred during one such attack.

Postmortem examination revealed tetralogy of Fallot, aneurysm of the main and left pulmonary arteries, and obstructive emphysema of the left lung.
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FIG. 3 Left. Case 2. Roentgenogram in the frontal projection showing emphysema of the left lung, shift of the heart and mediastinal structures to the right, and a large round density in the region of the pulmonary artery segment.

FIG. 4 Right. Case 2. Film obtained late in the cycle with contrast material outlining the right and left cardiac chambers, and a hugely dilated main and left pulmonary artery. Aortic filling followed opacification of the left chambers.

DISCUSSION

Aneurysm of the pulmonary artery is one of the rare lesions of the cardiovascular system, occurring at a rate of approximately 1 in 14,000 autopsies. Exceedingly uncommon are those forms of congenital aneurysm that cannot be accounted for by any known cause of pulmonary artery dilatation, viz., atrial septal defect, ventricular septal defect, patent ductus arteriosus, valvular pulmonic stenosis, and primary pulmonary hypertension.

The symptoms and findings of this lesion were comprehensively reviewed by Boyd and McGavack and by Deterling and Clagett. These include dyspnea, cough, chest pain, hemoptysis, and hilar mass with expansile pulsations. However, a thorough search of the literature failed to reveal any case in which a pulmonary artery aneurysm encroached upon the bronchial tree, producing serious obstructive symptoms.

The 2 cases herein presented are strikingly similar. In both a huge aneurysmal dilatation of the pulmonary artery impinged upon the walls of the left bronchus, leading to obstructive emphysema of the left lung and mediastinal displacement to the right. Unlike other forms of tracheobronchial obstruction of vascular nature, this lesion does not appear to be surgically correctible inasmuch as irreversible changes of the bronchial walls necessitate pneumonectomy, which is usually fatal in the first few months of life.

SUMMARY

Two infants with severe respiratory embarrassment secondary to compression of the left bronchus by a large aneurysmal dilatation of the pulmonary artery are presented. Review of the literature failed to reveal any similar case.

SUMMARIO IN INTERLINGUA

Es presentate le casos de duo infantes con sever embarrasso respiratori secundari a com-
pression del broncio sinistro per un grande
dilatazione aneurysmica della arteria pulmonar.
La revista della letteratura non produceva ulle
simile caso.

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A simple safe test for the detection of atrial septal defects which is of value in the
symptomless patient is described. This test is based upon the rise in the right atrial pres-
sure above the resting level that occurs immediately after performance of the Valsalva
maneuver. With an atrial septal defect or a patent foramen ovale the sudden rise in net
right atrial pressure during the first few seconds after the end of the Valsalva maneuver
causes a right-to-left pressure gradient across the defect with desaturated blood going
into the left atrium. The resulting change in arterial oxygen saturation may be detected
by the ear oxymeter method. In 32 patients with suspected atrial septal defects study
by means of this test proved to be of value. In 12 in whom the diagnosis was proved
by cardiac catheterization or thoracotomy, characteristic and reproducible changes in
arterial oxygen saturation were found. In 14 further patients with positive oxymeter
responses to the Valsalva test, there was catheter evidence of a left-to-right shunt at the
atrial level but definite anatomic proof of an atrial septal defect was lacking. Five
patients had a negative response. Three subsequently were found not to have atrial septal
defects and 2 had congestive cardiac failure associated with atrial septal defects. In
these latter 2 patients, Muller's maneuver or elevation of the legs caused a fall in systemic
arterial oxygen saturation, enabling the correct diagnosis to be made.
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