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Vascular Lesions of the Respiratory Tract and Disorders of the Chest Wall and Diaphragm

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VASCULAR LESIONS OF THE RESPIRATORY TRACT

Associations of pulmonary isomerism, including abnormal visceral situs syndromes and situs inversus, with abnormal patterns of pulmonary arterial supply to the upper lung lobes; of pulmonary stenosis with absence of the tracheal pars membranacea and bridging bronchus with sling left pulmonary artery; of abnormal systemic artery supply and venous drainage of the right lung in scimitar syndrome; and of systemic artery supply to loci of pulmonary sequestration are discussed in Chapters 7 and 8.

Malinosculation is a term proposed for a category of lesions characterized by establishment of abnormal communications through small openings or anastomoses between already existing blood vessels or other tubular structures.[1] As applied to the respiratory tract, the term could include the proposed secondary junction of lung and foregut in congenital bronchopulmonary-foregut malformation (*i.e.*, Srikanth type 4) and the systemic arterial supply to a normal area of lung, as in scimitar syndrome.[2]

Pulmonary Hemangiomas and Arteriovenous Fistulas

Hemangiomas involving the chest wall or mediastinum can extend into the lung. The most important category of parenchymal pulmonary vascular malformation is arteriovenous fistula. It occurs in approximately 60% of patients with autosomal dominant Rendu-Osler-Weber telangiectasia. The lesions are often multiple. They decrease in size with the Valsalva maneuver and increase with the Müller maneuver (Fig. 10-1). Pneumothorax, hemoptysis, and brain abscess can occur, and dyspnea, cyanosis, digital clubbing, and polycythemia from arteriovenous shunt are relatively frequent findings. These lesions are rarely clinically significant before the second decade of life (see Chap. 11).

Congenital Alveolar Capillary Dysplasia

Congenital alveolar capillary dysplasia is a recently appreciated entity that may originally have been described by McMahon in 1948.[3] The term applies to lungs with deficient alveolar wall capillaries and thick muscular walls in intraacinar arterioles.[4–6] The clinical picture is that of neonatal persistent pulmonary hypertension. The association with the lesion of misalignment of the pulmonary vessels, in which the veins run centrally in the lobules with the pulmonary arteries rather than in the interlobular septa, has been described.[6] Some patients described with this term may have the anatomically more severe form of lung maldevelopment called congenital acinar dysplasia (see Chap. 7).

Hemitruncus and Absence of Pulmonary Arteries

Absence of the left pulmonary artery is frequently associated with tetralogy of Fallot, although absence of the right pulmonary artery occurs more frequently.[7–9] Hemitruncus (*i.e.*, origin of a pulmonary artery from the aorta) can cause heart failure in early life and

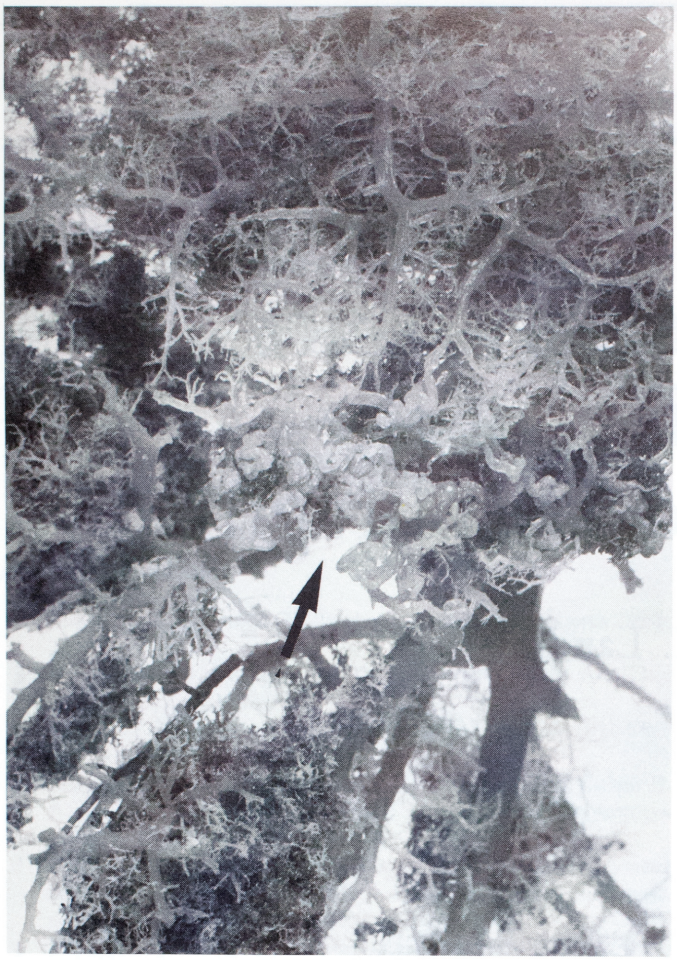

FIGURE 10-1. Plastic vascular cast of the lung in a 32-year-old patient with clinical and radiologic findings of arteriovenous fistulas. The lesion consists of a mass of thick, tortuous, anastomosing vessels (*arrow*). (Contributed by the editor.)

can lead to pulmonary vascular sclerosis of the affected lung like that seen bilaterally with patent ductus arteriosus.[10]

Azygos and Hemiazygos Veins

Not technically components of the respiratory tract, the azygos and hemiazygos veins are mentioned because of their relatively frequent association with the azygos lobe. The septum through the apical right lung can be seen radiologically as a line leading to the teardrop-shaped azygos vein.[11] The rare hemiazygos lobe must be differentiated from that of the left azygos lobe in situs inversus. In situations with azygos or hemiazygos continuation of the inferior vena cava, reported especially in cases of Ivemark asplenia and the polysplenia syndromes, the involved veins are enlarged.[12]

Arteriohepatic Dysplasia

Arteriohepatic dysplasia was a term originally applied to children with peripheral pulmonary artery stenoses and obstructive jaundice due to the paucity of intrahepatic bile ducts (*i.e.*, Alagille syndrome). Other features include vertebral abnormalities and distinctive facies. Diaplacental rubella can cause peripheral pulmonary artery stenosis and cholestatic liver disease. The nonsyndromic form of the Alagille paucity of intrahepatic bile ducts appears to have a poorer prognosis than the syndromic form.[13]

DISORDERS OF THE CHEST WALL AND DIAPHRAGM

The radiologic manifestations of many chest wall and diaphragmatic abnormalities (*e.g.*, the association of rib abnormalities, scoliosis, and vertebral anomalies with the basal cell nevus syndrome, which are autosomal dominant disorders of deformed, hypoplastic, or ribbonlike ribs with neurofibromatosis; micrognathia, multiple rib defects, and flail chest with cerebrocostomandibular syndrome) have been reviewed by Swischuck.[14]

Congenital Midline Cervical Cleft

Congenital midline cervical cleft is apparently sporadic, but it is more frequently seen in Caucasian females. It consists of an area of mucosal surface in the midline lower neck with a caudal sinus and a cephalad skin. The condition appears not to be related to thyroglossal duct or sinuses; it may be related to ectopic first branchial arch derivatives, and it has been associated with midline clefts of the tongue, lower lip, mandible, and sternum.[15] Epiglottic aplasia with secondary respiratory tract disease due to persistent aspiration of secretions or feedings has also been associated with median cleft of the mandible.[16]

Intrathoracic Rib

Intrathoracic rib is a rare lesion, with only 10 to 12 reported cases.[17] It radiologically appears as a smooth, ribbonlike, bony density arising from the posterior inferior face of another rib or from a vertebral body and coursing caudally, sometimes to the level of the diaphragm. It appears to cause no specific symptoms.

Diaphragmatic Eventration and Hernia

Probably the most common cause of eventration (*i.e.*, muscular deficiency with usually upward bulging of the affected hemidiaphragm) is denervation (*e.g.*, phrenic paralysis), but primary myopathy can also be a cause, and intrinsic failure of development of the diaphragmatic muscle cannot be excluded in some patients. Respiratory distress can occur if upward displacement of the diaphragm compromises lung expansion and if both hemidiaphragms are affected. Upward movement of the diaphragm on inspiration and downward movement on expiration interfere with ventilation.

Hernias through the clefts in the anterior diaphragm muscle at the sites of the inferior mammary arteries, the foramina of Morgagni, are usually small, and because they are covered by a membrane of fused peritoneum and pleura, upward herniation of abdominal viscera is restricted, and they are rarely clinically significant. They are most frequently associated with trisomy 18 and are seen as small, usually bilateral ear-shaped protrusions on the superior hepatic surface.

Comparable and usually small and membrane covered are the posterior hernias of the foramina of Bochdalek; adrenal and kidney can protrude upward through these hernias. The usually left-sided diaphragmatic hernia due to a defect of the central tendon of the diaphragm (*i.e.*, Bochdalek hernia) is not a hernia through a

Bochdalek foramen.[18] Typically, these hernias are large enough to permit significant upward herniation of abdominal viscera, including liver, stomach, small intestine, and spleen (Color Fig. 10-1; Fig. 10-2). Hypoplasia of the lung from reduced intrathoracic space occurs frequently (see Chap. 7). If the degree of visceral displacement is great enough, mediastinal shift and hypoplasia of the contralateral lung can occur. Bochdalek hernia and its sequelae are one of the less treatable major congenital malformations. The association of diaphragmatic defect of an eventration type with left extralobar sequestration of the lung is discussed in Chapter 8.

Accessory Diaphragm

The most common form of accessory diaphragm divides the right lower thoracic cavity by running laterally from the diaphragm anteriorly to the 3rd to 7th ribs posteriorly.[19] It is most frequently a feature of the scimitar syndrome.

Ectopia Cordis, Left Ventricular Diverticulum Syndrome, and Pentalogy of Cantrell

In ectopia cordis, the heart protrudes from the thorax through a sternal cleft and is exposed (*i.e.*, naked heart) or displaced into the abdomen, and the thoracic cavity is unusually small.[20,21] In addition to the sternal cleft, there are pericardial defect and diastasis recti or epigastric omphalocele, and the apex of the externalized heart points cephalad toward the chin. In thoracoabdominal ectopia cordis, there is a lower sternal cleft, defect of the septum transversum of the diaphragm and pericardium, and omphalocele or epigastric hernia into which the heart is displaced. Many types of congenital cardiac malformations affect ectopic hearts; tetralogy of Fallot is perhaps the most frequent. Pentalogy of Cantrell is a term that refers to thoracoabdominal ectopia cordis.[22–24] There is often a left ventricular diverticulum in the epigastrium (*i.e.*, left ventricular diverticulum syndrome).

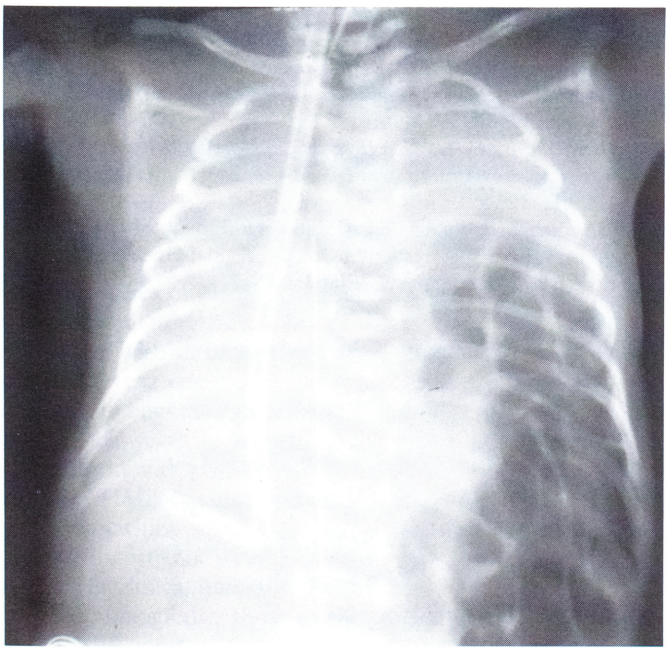

FIGURE 10-2. Radiologic appearance of a large, left diaphragmatic hernia that caused the death of a patient (see Color Fig. 10-1). (Courtesy of Juan Payser, M.D., Miami, FL.)

Amniotic Bands

Amniotic bands occasionally involve the thoracic wall. An example with deep grooves extending from the epigastrium bilaterally to the posterior thoracic midline plus constrictions and amputations of digits was given by Jones and Sinclair.[25] The researchers proposed that the lower body of the fetus had protruded through the site of amniotic rupture.

Intrathoracic Kidney

The presumed mechanism is protrusion of a kidney upward through an enlarged Bochdalek cleft or a muscular gap in the posterior lateral diaphragm. The clinical significance is primarily misdiagnosis of intrathoracic kidney as a posterior mediastinal or pulmonary tumor.[26]

Thoracoabdominal Enteric Duplication

Of 25 cases of gastrointestinal tract duplications extending from the abdomen into the thorax reviewed by Pokorny and Goldstein, 24 were on the right side.[27] They ascertained the sites of the gastrointestinal tract connections in these patients: jejunum, 9; duodenum, 4; ileum, 2; esophagus, 1; and not established, 9.

Poland Syndrome and Associated Disorders

The Poland syndrome of partial or total absence of the pectoralis major muscle with hand abnormalities on the affected side has been reported in association with pulmonary agenesis.[28] The condition can produce apparent radiologic hyperlucency (*i.e.*, transradiancy) of the lung on the affected side, an effect similar to that of mastectomy. Deshpande and colleagues reported a patient with absence of the left 4th and 5th ribs, fusion of the left 6th and 7th ribs, a supernumerary left breast and nipple, hypoplasia or absence of the upper left lung, high left scapula, scoliosis, and vertebral anomalies.[29]

Pectus Excavatum, Carinatum, and Other Chest Deformities

The principles of surgical correction of pectus excavatum, carinatum, and other chest deformities are presented by Garcia and associates[30] and by Shamberger and Welch.[31] Blickman and colleagues discuss the use of preoperative and postoperative scintigraphic pulmonary ventilation and perfusion studies to ascertain the benefit of the surgical procedures for lung function.[32] Although rare, such deformities can follow surgery for congenital heart disease or diaphragmatic hernia.[33,34] Huemmer and Willital presented a scheme of eight possible categories of chest wall deformities using frontal, sagittal, and horizontal plane measurements.[35]

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