De novo Circumscribed Pulmonary Lobar Cystic Lymphatic Anomaly in a Young Boy

A Possible Sequela of Bronchopulmonary Dysplasia

Nelly Karmazin, M.D.; Howard B. Panitch, M.D.; Rohinton K. Balsara, M.D., F.C.C.P.; Eric N. Faerber, M.D.; and Jean-Pierre de Chadarevian, M.D.

This report describes a massive pulmonary lymphatic cystic anomaly affecting the right lower lobe of a nine-year-old boy. A year earlier, only an ill-defined small infiltrate could be seen in the affected lobe radiologically. The pathogenesis of this highly unusual lesion is discussed, taking into consideration the possible role of three months of mechanical ventilation in the neonatal period.

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Diffusely anomalous pulmonary lymphatic drainage in children is seen in the neonatal period. It is referred to as lymphangiectasis and tends to be uniformly fatal soon after birth. It may be isolated or associated with cardiovascular malformations and, in particular, an anomalous pulmonary venous return. Occasionally, it may be part of a generalized lymphatic dysplasia affecting many sites.1-3

Apparently primary but localized pulmonary lymphatic anomalies have been reported on rare occasions in adults.4,5 They have been called lymphangiomas or lymphangiectasis. Such lesions, to our knowledge, are exceptional in children,6,7 which makes the lesions unlikely to be included in the differential diagnosis of circumscribed cystic pulmonary lesions. This report proposes to document such a case and discuss its pathogenesis.

Case Report

A nine-year-old boy was admitted to St. Christopher's Hospital for Children with the chief complaint of fever and hemoptysis. Born 12 weeks prematurely, he had required mechanical ventilation for the first three months of life and had developed necrotizing enterocolitis, which was treated surgically. At eight years of age, he was incidentally found to have a small infiltrate in his right lower lobe, but was lost to follow-up until he developed low-grade fever and hemoptysis a year later. At this time, his PPD test and cultures of sputum were negative and his chest x-ray film and CT showed a round, well-defined, large right lower lobe mass (Fig 1). Contrast medium was administered during the study and failed to demonstrate any arteriovenous connection. A right lower lobectomy was performed.

Pathologic Study

At the posterior basal segment of the resected lobe, a spherical mass extended from the pleura into the pulmonary parenchyma (Fig 2). The visceral pleural surface covering the mass contained multiple dark red cysts measuring 0.2 to 0.5 cm in diameter. The cut surface of the mass showed it to be 4.5 cm in diameter, round and well demarcated. It had a spongy consistency and was made of numerous cystic spaces filled with blood-stained fluid.

Histologic examination revealed a network of channels, most of which centered around the bronchioles. Some were present in the septa and in the subpleural region. There was no capsule separating the lesion from the surrounding parenchyma. The channels varied in size, and their walls were of variable thickness. The inner surface of the channels was lined by flattened endothelial-looking cells (Fig 3). The walls were composed of connective tissue in which, in some areas, groups of smooth muscle fibers were present. Focal aggregates of lymphocytes could be seen in the walls of the channels and in the parenchyma adjacent to the lesion. Some channels were filled with red blood cells; others contained eosinophilic fluid. The surrounding pulmonary tissue was the site of a mild peribronchiolar lymphocytic infiltration. Acute intra-alveolar hemorrhage was prominent in a few areas.

Discussion

Diffuse or localized dysplastic development and increased intralymphatic pressure are essentially the two pathogenetic mechanisms traditionally invoked to explain the presence of lymphatic anomalies in children.

*From the Departments of Anatomical Pathology (Drs. Karmazin and de Chadarevian), Surgery (Dr. Balsara), Radiology (Dr. Faerber), and Pediatrics (Drs. Panitch and de Chadarevian), St. Christopher's Hospital for Children and Temple University School of Medicine, Philadelphia.

Reprint requests: Dr. de Chadarevian, St. Christopher's Hospital, 2600 North Lawrence, Philadelphia 19133
In the case of this patient, it is not possible to categorically establish the pathogenetic mechanism involved in the development of the pulmonary lesion. Its first detection at eight years of age and gradual enlargement on sequential chest roentgenograms is not necessarily an indication in favor of or against the diagnosis of lymphatic hamartoma (lymphangioma). It is well established that vascular hamartomas may be inconspicuous at birth. They enlarge through opening of collapsed channels, in addition to their natural growth which is coordinated with that of the rest of the body. Therefore, this lesion, in the absence of detectable morphologic cause for its development, could be construed to be a lymphangioma, similar to the lesions reported ten years ago in three boys aged 13, 16, and 19 years. In two of those cases, there was associated mediastinal involvement, and two lobes were affected. On the other hand, one cannot easily dismiss the possible role played by pulmonary scarring which may have developed following the stormy perinatal period this child went through, and which required mechanical ventilation for the first three months of life. Lymphangiectasis does develop in the course of bronchopulmonary dysplasia; and although at nine years of age, there is no evidence of residual pulmonary damage, it is not inconceivable that this might have played a role in the pathogenesis of the present lesion. Unfortunately, at this stage, we know of no way to answer the question, although a significant future increase in the number of cases of late-onset localized pulmonary lymphatic anomalies would definitely favor such an explanation.

REFERENCES
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