Radiological Case of the Month

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AGE 3 WEEKS, a full-term newborn developed recurrent cough and stridor without other signs of respiratory distress. She had no feeding problems and was thriving. At age 6 months, inspiratory and expiratory stridor became more pronounced, and she developed dyspnea. A chest radiograph was obtained to evaluate the airway, upper mediastinum, and esophagus (Figure 1). The vertebrae were normal. Magnetic resonance imaging followed (Figure 2). Surgery was performed, and the stridor and respiratory distress ceased immediately after surgery.

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Congenital Esophageal Duplication Cyst as a Rare Cause of Neonatal Progressive Stridor

A thoracotomy performed because of progressive stridor and dyspnea showed a fluid- and blood-filled mass adherent to the esophagus. It was completely resected. Histopathological examination showed esophageal mural structure with gastric mucosa (Figure 3). The diagnosis was an esophageal duplication cyst without connection to the lumen of the esophagus.

Esophageal duplications are congenital malformations resulting from aberration of ventral budding of the lung primordium from the embryonic foregut at 3 to 4 weeks’ gestation.1,2 They are tubular or cystic mediastinal masses with or without associated congenital anomalies (such as duplication of the small intestine, esophageal atresia, tracheoesophageal fistula, and spinal and/or vertebral anomalies). The overall incidence is estimated as 1 in 8200, with a predominance in males.3 Cystic duplications (or enteric cysts) are more rare and communicate with the esophageal lumen in 10% of patients.4

Although most cases are asymptomatic, swallowing difficulty, failure to thrive, or respiratory distress can be caused by displacement or compression of mediastinal structures. These and other clinical signs, including fever or cough, may be produced by enlargement of the cyst as a result of infection or bleeding, most likely to occur in duplication cysts, which contain gastric mucosa. There are few cases of enteric cysts as the cause of stridor in neonates or young infants.5,6

The differential diagnosis of masses in the upper posterior mediastinum includes bronchogenic cysts and neurogenic neoplasms. Bronchogenic cysts arise from the same embryonic esophageal bud and are more frequent than enteric cysts.7 Magnetic resonance imaging and computed tomographic scans clearly demonstrate esophageal duplication cysts.8 Barium esophagram may detect duplications of the esophagus, but in cases of a cystic mass of the upper mediastinum, it is not helpful because of the infrequent connection of enteric cysts to the esophageal lumen. Surgical resection of the cyst is the treatment of choice. Thoracoscopic surgery may be used as a method of less invasive treatment for this benign disorder.9

Our patient presented with congenital stridor and progressive dyspnea caused by an esophageal duplication cyst. Stridor is a relatively frequent symptom in newborns and young infants and is caused by a variety of diseases or abnormalities. A mass in the upper mediastinum causing inspiratory and expiratory stridor is a rare finding. Respiratory distress necessitated surgery, and the patient was cured by resection of the cyst.

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REFERENCES