Roentgenogram of the Month

Recurrent Bibasilar Infiltrates*

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A 62-year-old man was admitted with fever, weight loss, and worsening of his chronic cough, productive of sputum. There was no history of alcoholism, loss of consciousness, or dysphagia. He was treated for recurrent bacterial pneumonia during the past eight years. He had smoked cigars for 40 years.

Physical examination showed a cachectic man with a respiratory rate of 32/min, temperature 38.5°C, pulse 120/min. He had clubbing of the fingers. There were bilateral wheezes and bibasilar crackles. His white blood cell count was 12,200/cu mm with 73 percent neutrophils and 8 percent bands.

Sputum cytology and cultures for bacteria, mycobacteria, and fungi were nondiagnostic. A variety of laboratory tests were negative, including cold agglutinin, antinuclear antibodies, rheumatoid factor, and complement fixation for fungi. Esophagogram was normal. The chest roentgenogram is shown in Figure 1. A tomogram of the neck and upper thorax is shown in Figure 2.

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Figure 1

Figure 2

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**Diagnosis: Tracheobronchiomegaly (Mounier-Kuhn syndrome)**

The PA chest roentgenogram (Fig 1) shows patchy bibasilar consolidation with shaggy confluent opacities more on the right than left. The trachea is abnormally widened. Tomogram of the neck (Fig 2) confirms the marked dilatation of the trachea, which measures 40 mm in its largest diameter (Fig 3). Fiberoptic bronchoscopy performed after the patient swallowed 60 ml of methylene blue solution, revealed tracheobronchiomegaly with collapse of airways on expiration, tracheobronchitis, and pooling of purulent secretions in the lower lobes. Methylene blue was not seen in the airways. Tracheobronchial biopsy of the basal infiltrate showed pneumonia.

Tracheobronchiomegaly, first described clinically by Mounier-Kuhn1 in 1932, is a rare disease characterized by extremely large central airways and recurrent bouts of bronchitis and pneumonitis. Katz et al2 defined the normal dimensions of the trachea and main stem bronchi and compared them with the dimensions seen in these patients. They labelled the syndrome "tracheobronchiomegaly." Himalstein and Gallagher3 found at least five cases in approximately 500 bronchograms of adults, an incidence of 1 percent. This should remind those caring for patients with pulmonary disease to watch for this relatively rare and unappreciated cause for recurring pneumonitis.

The diagnosis may be suspected from the routine posteroanterior roentgenogram of the chest when the air column of the trachea appears widened, especially when its right border is visible to the right of its normal position in the superior mediastinum. Bronchoscopy and bronchography are useful to confirm the diagnosis, but are not essential. Bronchograms may provide the accurate measurements that confirm or refute the diagnosis. There is often marked collapse of the trachea and large bronchi on expiration.

The cause of this disease is unknown. One of the mechanisms that has been proposed to explain the airway dilatation is segmental myenteric plexus deficiency. However, Martinez-Catinchi et al4 showed hyperactivity of the airways to methacholine. This is in contrast to the theory proposed by Diaz5 and confirms the report of Ratliff et al6 who observed neural elements in tracheal biopsies. Two documented cases within the same family led Johnson and Green7 to suggest a familial etiology. The present theory is that the condition is caused by congenital sparsity of elastic and muscular elements within the walls of trachea and bronchi that promote the retention of secretions, which eventually become contaminated and lead to repeated pulmonary infections, parenchymal destruction, and disability.

Therapy should include prophylaxis against upper respiratory tract infections, more prolonged antibiotic therapy than the usual case of bronchitis or pneumonitis, and postural drainage.

**REFERENCES**

5. Diaz CJ. Un caso de megartagusa idiopatica con truquemalacia. Rev Clin Esp 1940; 1:432