A Rare Cause of Cough

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A 65-year-old woman with a long history of nonproductive cough was referred to the Department of Pulmonary Diseases because at routine physical check-up her chest x-ray film (Fig 1) showed an abnormal bulge at the right heart border and a linear density at the base.

The past medical history was notable only for a thyroid operation 16 years previously and viral hepatitis at age 35. She did not smoke, use alcohol, or take any medications.

The physical examination findings were as follows: blood pressure, 120/80 mm Hg; pulse rate, 76 beats per minute; respiratory rate, 18/min; and temperature, 37.5°C. The anteroposterior diameter of the chest was slightly increased. Breath sounds were decreased over the right lung base. There were no adventitious sounds. Complete vision loss due to senile cataract was determined by an ophthalmologist. The remainder of the physical examination was unremarkable.

Hematologic studies revealed the following values: hematocrit, 39 percent; platelets, 270,000; and white blood cell count, 5,000/cu mm, with a normal differential. Routine blood biochemistry and urinalysis values were within normal limits. There was an electrocardiographic pattern of right bundle branch block. Echocardiographic findings were normal. The pulmonary function studies revealed a slight volume decrease. Arterial blood gas values were as follows: pH, 7.44; Pco₂, 42 mm Hg; and Po₂, 86 mm Hg.

Fiberoptic bronchoscopy revealed a narrowed superior segment bronchus of the right lower lobe with a pulsatile medial wall and intact mucosa.
Diagnosis: Segmental atelectasis due to compression by a right descending aorta

Computed tomography (CT) of the chest demonstrated a normal left aortic arch. Immediately beyond the aortic arch, the aorta crossed to the right behind the esophagus at the level of bifurcation (second arcus). It then descended on the right side of the vertebral column (Fig 2). The CT study also revealed that the right descending aorta was compressing the right lower lobe bronchus, causing atelectasis of the superior segment (Fig 3). An aortogram confirmed the CT findings.

Congenital abnormalities of the aortic arch have been well described. It is of interest that patients with vascular anomalies of the aortic arch may have considerable variation in the degree of symptoms, from severe airway obstruction and dysphagia to no symptoms. One such abnormality is a left aortic arch and right descending aorta; 16 cases have been described in the literature.1-3

In this anomaly, the ascending aorta arises normally from the left ventricle and courses in the usual manner in front of the trachea and over the left bronchus to form a left aortic arch. Instead of following a normal course to the left and posteriorly, the descending portion of the arch turns abruptly and then passes transversely behind the esophagus to the right of midline and continues as a right-sided descending aorta.4

The reason for this anomaly is uncertain. Shuford and Sybers4 suggested that when there is normal branching of the arch vessels, the anomaly may be the result of an interruption of the embryonic right arch of the hypothetical double aortic arch between the right subclavian artery and the descending aorta.

The majority of reported cases of left aortic arch and right descending aorta had associated cardiovascular and other congenital anomalies.3 Of the 16 cases reported previously, only four had no associated anomalies. When a left aortic arch with right descending aorta occurs together with a right ductus arteriosus or ligamentum arteriosum, they form a vascular ring. Clinical manifestations depend on the severity of compression of the airways or esophagus.1-3 In our case there was no vascular ring or any other cardiac anomaly.

Domínez and colleagues5 point out that the findings on plain chest radiograph and on barium swallow study are diagnostic and that aortography is not required for confirmation. The diagnostic criteria of a left aortic arch with right descending aorta on routine chest roentgenogram are tracheal deviation to the right and location of the descending aorta on the right side of the thoracic vertebral column.6 Obtaining a CT scan of the chest before aortography is valuable for the diagnosis.

In our case, the reason for the segmental atelectasis determined at bronchoscopy was the compression of the right lower lobe bronchus by the right descending aorta. We have not encountered a similar case in the literature.

REFERENCES
5. Domínez R, Oh KS, Dorst JP, Young LW. Left aortic arch with right descending aorta. AJR 1978; 130:917-20