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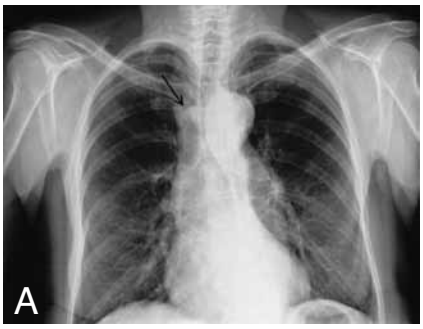
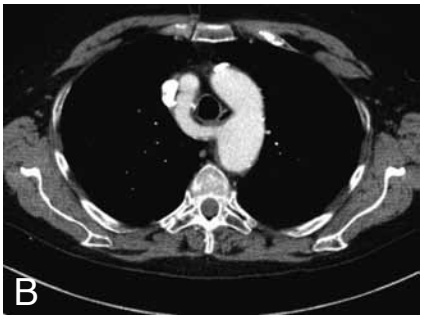
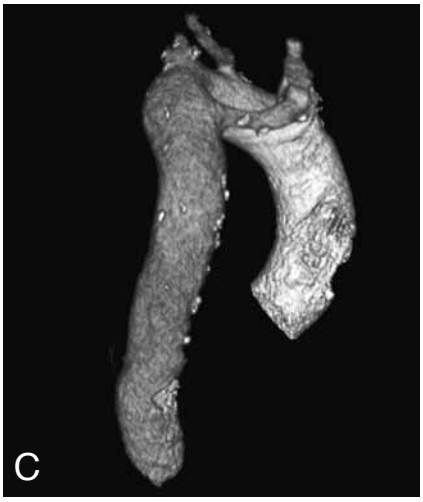
IMAGES IN CLINICAL RADIOLOGY







Silent double aortic arch in an elderly patient

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A 63-year-old woman with COPD presented to our hospital with complaints of breathlessness on exertion, productive cough and weight loss. Chest X-ray showed a bright silhouette extending from the aortic knob in the right paratracheal area on the PA-view and a posterior indentation of the tracheal air column on the lateral view. No other abnormalities were observed. Further work-out consisted of a contrast enhanced CT of the chest which revealed bilateral aortic arches, both of which arising from the ascending aorta anterior to the trachea. Each arch gave origin to the common carotid artery and the subclavian artery, and joined the right posterior ward to form the descending aorta. The left arch was the larger of the two arches. The trachea and esophagus were placed in the middle of the aortic ring. No abnormal compression of the trachea or the esophagus was observed.

Comment

Congenital anomalies of the aortic arch represent less than 1% of all congenital cardiac defects. Double aortic arch is a splitting of the ascending aorta into two segments, which pass on either side of the esophagus and trachea and join together as a single descending aorta. Double aortic arch is the most common form of a complete vascular ring, a class of congenital anomalies of the aortic arch system in which the trachea and esophagus are completely encircled by connected segments of the aortic arch and its branches. In the most common variation of double aortic arch, both aortic arch segments are patent, the right segment is larger than the left segment and the ductus arteriosus and descending aorta are left sided (75% of cases). A left dominant double aortic arch with a right descending aorta, as was observed in our patient, is less common and constitutes 18% of cases. In the codominant or balanced type, seen in 7% of cases, both arches are of equal diameter. In an uncommon variation of the double aortic arch, one of both arches may be atretic and may be represented by only a cord.

Double aortic arch is a congenital anomaly which arises because of persistence of the fourth right pharyngeal arch artery. Embryologically, the ventral and dorsal aortas are connected by aortic arches which persist or involute to give rise to the normal aortic arch, its branches and the minor arteries of the head. The right fourth pharyngeal arch normally involutes at about 36 to 38 days. The remnant of the right pharyngeal arch becomes the right innominate artery. The left fourth pharyngeal arch persists and gives rise to the normal left aortic arch. Failure of dissolution of the right fourth pharyngeal arch artery and persistence of the left pharyngeal arch produces a vascular ring consisting of two aortic arches and encircling the trachea and esophagus. Although the defect is most frequently seen as an isolated defect in an otherwise normal individual, it has occasionally been associated with tetralogy of Fallot, transposition of the great arteries, pulmonary atresia defects, ventricular septal defects and 22q11.2 deletion syndrome.

In most cases, this abnormality is diagnosed in infancy and patients present with symptoms related to esophageal or tracheal obstruction such as stridor, respiratory distress, recurrent upper respiratory tract infection or pneumonia, dysphagia or feeding difficulties. The severity and onset of symptoms depends on the space between the aortic segments. Sporadically, patients have been reported in whom diagnosis was delayed until childhood or even adulthood. Presentation in these patients usually consists of progressive or persistent dyspnea, dysphagia or asthma-like symptoms. The symptoms in adult patients can be atypical and misleading, as illustrated by case reports of double aortic arches detected in adulthood after long-term treatment for assumed reactive airway-disease. Case reports of elderly patients are rare, especially symptom-free cases.

In our patient a double aortic arch was discovered incidentally when imaging studies were conducted for ruling out pulmonary infection and/or malignancy. As the trachea and esophagus were placed in the middle of the aortic ring and no compression of the trachea could be observed, our patients complaints of breathlessness were attributed to a non-infectious COPD exacerbation and she quickly improved under optimal inhalation therapy and corticosteroids. No satisfactory explanation for the weight loss was found.

Reference

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