A 35-year-old female patient was presented to our hospital with complaints of wheezing and dyspnea on efforts of three years duration. She did not have any personal history of atopy and never smoked. Her physical examination was normal. The chest X-ray demonstrated an abnormality in the right paratracheal region with absence of the aortic knuckle on the left side (Figure 1). It was reported by the spirometry technician that the patient underwent a presyncopal episode after the spirometry test, so the symptoms related to the presyncopal episode were improved. It was therefore suspected that the patient underwent an inspiratory flattening before the fluid treatment, it showed both inspiratory and expiratory flattening after the fluid treatment (Figure 2a and 2b). The diagnosis was confirmed by computed tomography (CT) scan (Figure 3).

What is your diagnosis?
See the next page
CT scan of the thorax showed a right-sided aortic arch, which explained the findings (Figure 3).

Conclusion

The aortic arch anomalies are rarely seen in adults and are commonly asymptomatic. Patients can present to hospital with exercise dyspnea, wheezing, stridor, recurrent respiratory infections, cough, and dysphagia. The progression from asymptomatic to symptomatic form depends on the tracheal compression caused by the aorta. In addition, chronic tracheal compression caused by the aorta can lead to softening of the tracheal wall (called as tracheomalacia); therefore, the trachea becomes more susceptible to collapse.

The patient’s initial symptoms included dyspnea and wheezing during exercise, which depended on dynamic changes induced by exercise in the aorta. The symptoms disappeared due to the fact that the diameter of aorta reverts to normal after exercise. In other words, at rest, the compression did not produce narrowing of the airway sufficient to cause symptoms. However, the fluid-treatment given due to the presyncopal episode rendered the tracheal compression sufficient to cause symptoms at rest too.

Obstructive lesions of the central airways can be divided functionally into variable and fixed types. If the airway obstruction, caused by any pathology, shows variability during the respiratory cycle, it is called as variable type. The reverse applies to the fixed type. In the spirometric examination, flow-volume curve gives important information about the central airways. While variable intrathoracic CAO results in flattening of only the expiratory limb of the flow-volume curve, fixed intrathoracic CAO results in flattening of both the expiratory and inspiratory limbs of the flow-volume curve. Respiratory physiology is mainly the determining factor concerning the functional discrimination. In intrathoracic airways, during inspiration, extratracheal pressure (approximately equal to intrapleural pressure) is more negative relative to intratracheal pressure and this negative pressure gradient enlarges the tracheal lumen, whereas during expiration, extratracheal pressure is more positive relative to intratracheal pressure and this positive pressure gradient narrows the tracheal lumen. In addition, the narrowing in the tracheal lumen leads to an increase in both velocity of the airflow, which is also called as convective acceleration, according to Bernoulli’s principle, decreases gas pressure in the constricted region. Frictional resistance caused by turbulent flow also contributes to the pressure loss. Constricted segment therefore further decreases the intratracheal pressure in the distal side of constricted segment, which results in further contraction in the airway.

In this patient, changes of the flow-volume curve were interpreted in the following way. During forced expiration, the tracheal compression was strengthened by the expiratory mechanisms mentioned above, thereby resulting in flattening of the expiratory limb of the flow-volume curve in both before and after fluid loading. During forced inspiration, because the negative pressure gradient compensates the compression caused by the aorta before fluid loading, the inspiratory limb of the flow-volume curve was not affected. In contrast, extra-pressure caused by the aortic dilatation after fluid loading cannot be compensated by forced inspiration anymore, and therefore the flow-volume curve also was showing flattening of the inspiratory limb.

Flow-volume curve examination is specific but not sensitive for the diagnosis of CAO. It cannot detect CAO, unless tracheal narrowing becomes more than 80%. Therefore, if there is a sign of flattening in the flow-volume curve, it indicates that tracheal narrowing is clinically significant. Vascular anomalies in adults are often mistaken for asthma. Delayed diagnosis can result in unnecessary investigations and periods of ineffective treatment. Flow-volume curve is a helpful instrument in the diagnosis of CAO and a right aortic arch anomaly. This report underscores the importance of spirometry in the evaluation of patients with dyspnea.

References