Case report

An unusual spirometric shape that you must not forget

Case report

A 37-year-old woman, who is a mother of a 5-year-old girl, presented to our outpatient pulmonary clinic because of cough and yellowish expectorate for 20 days. She was a former smoker of 10 cigarettes a day for 20 years (10 pack-years) and worked as agricultural day labourer. Since the age of 25 years, the patient reported one or two episodes of lower respiratory tract infection (LTRI) during the winter months that resolved within a few days of antibiotic and mucolytic treatment. The persistence of cough and expectorate of the last LTRI let the general practitioner to seek a respiratory specialist respiratory consultation with spirometry.

She denied exertional dyspnoea and other symptoms. Moreover, skin prick tests for airborne allergens were moderately positive for Dermatophagoides pteronyssinus and Dermatophagoides farinae mites, but there was no history of rhinitis and of any other disease.

The physical examination of the chest was within normal limits, with no additional respiratory or cardiac sounds. Oxygen saturation by pulse oximetry was 96% on room air, blood pressure 120/80 mmHg and heart rate 65 beats per min with a regular rhythm.

Slow and forced ventilatory manoeuvres were executed without difficulty and with the patient’s full cooperation, despite her cough (figure 1).

Figure 1 Spirometry, flow–volume curve. Red arrow: volume–time curve; blue arrow: flow–volume loop; black circles: normal limits of the expiratory part of the flow–volume curve.

Task 1

The spirometric loop could be attributable to:

a. variable obstruction of extrathoracic airways
b. variable obstruction of intrathoracic airways
c. fixed airway obstruction
d. COPD

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An anomalous spirometric pattern can initiate a diagnostic path leading to the finding of a congenital vascular malformation. The flow–volume curve may hide a nonrespiratory pathology. Spirometry is not just asthma or COPD. http://bit.ly/30iHRCp
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Considering the clinical suspicion, we requested an ear, nose and throat (ENT) assessment with rhinolaryngoscopy, and a standard chest radiograph. Moreover, we asked the patient about previous chest radiography and ENT evaluations. She denied having undergone these examinations in the past. The endoscopic examination of the upper airways did not show any anatomical abnormalities. Figure 2 shows chest radiograph in posterior–anterior projection.

**Table 1 Spirometry: best and reference values**

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Best</th>
<th>Ref.</th>
<th>% ref.</th>
</tr>
</thead>
<tbody>
<tr>
<td>VC L</td>
<td>4.330</td>
<td>3.000</td>
<td>144%</td>
</tr>
<tr>
<td>FVC L</td>
<td>4.430</td>
<td>3.000</td>
<td>148%</td>
</tr>
<tr>
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<td>2.580</td>
<td>117%</td>
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<tr>
<td>FEV&lt;sub&gt;1&lt;/sub&gt;/FVC</td>
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<td></td>
<td></td>
</tr>
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<td>1.36</td>
<td>31%</td>
</tr>
<tr>
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<td></td>
<td></td>
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<td>6.300</td>
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<td>6.300</td>
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<td>4.000</td>
<td>67%</td>
</tr>
<tr>
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<td>4.000</td>
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<tr>
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</table>

VC: vital capacity; PIF: peak inspiratory flow; FEF<sub>50%</sub>: forced expiratory flow at 50% FVC; FIF<sub>50%</sub>: forced inspiratory flow at 50% FVC. Bold indicates reduced values.

**Task 2**

Which of the following findings can you observe on the chest radiograph?

a. Nothing abnormal
b. Stenosis of the lower third of the trachea, absence of the first left cardiac arch and right upper paratracheal abnormal mediastinal widening

c. Isolated tracheal stenosis

**Answer 2**

b. The chest radiograph evidences a stenosis of the lower third of trachea in addition to an upper right mediastinal widening and absence of the first arch of the left cardiac contour.

**Task 3**

What would be your further investigations for a patient with these features?

a. Echocardiogram
b. Chest computed tomography (CT) with contrast agent
c. Chest magnetic resonance imaging (MRI)
d. Fibreoptic bronchoscopy
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Chest CT with contrast was performed using a Toshiba Aquilion 64-slice scanner with the following parameters: 120 kV, 300 mA and rotation time 0.5 s. Figure 3 shows two mediastinal scans, an axial and a coronal one.

Contrast-enhanced CT was enough to make a diagnosis of fixed airway obstruction due to tracheal stenosis secondary to a fully closed vascular ring. The relationship between the vascular structures of the ring and the trachea is well visualised by the volume-rendering three-dimensional reconstruction (figure 5).

After this finding, the patient underwent colour Doppler echocardiography, which was normal, and biochemistry examinations, which only showed

**Figure 3** Contrast-enhanced chest CT (mediastinal window): a) axial scan; b) coronal scan. RAA: right aortic arch; LBCA: left brachiocephalic artery; Tr: trachea.

**Answer 3**
b. Chest CT with a contrast agent is the investigation of choice because it allows a proper and combined assessment of trachea and mediastinal structures.

Chest CT with contrast was performed using a Toshiba Aquilion 64-slice scanner with the following parameters: 120 kV, 300 mA and rotation time 0.5 s. Figure 3 shows two mediastinal scans, an axial and a coronal one.

**Task 4**
What do you see on the chest CT images?

a. Right aortic arch (RAA), mirror image branching of the arch vessels
b. Isolated narrowing of the trachea
c. Kommerell’s diverticulum at descendent portion of the aortic arch.
d. Aortic ring made of aortic arch at the front on the right, left brachiocephalic artery (LBCA) at the front on the left, Kommerell’s diverticulum at the back on the left; tracheal stenosis.

Contrast-enhanced CT was enough to make a diagnosis of fixed airway obstruction due to tracheal stenosis secondary to a fully closed vascular ring. The relationship between the vascular structures of the ring and the trachea is well visualised by the volume-rendering three-dimensional reconstruction (figure 5).

After this finding, the patient underwent colour Doppler echocardiography, which was normal, and biochemistry examinations, which only showed

**Figure 4** Contrast-enhanced chest CT (mediastinal window) chest: a) axial scan, diameter of diverticulum; b) coronal scan, tracheal diameter. KD: Kommerell’s diverticulum.

**Answer 4**
CT scan revealed a RAA, mirror image branching of the arch vessels, Kommerell’s diverticulum at the descendent portion of the aortic arch and narrowing of the trachea just 4 cm above the carina with triangular shape in the axial projection. The narrowing was due to an aortic ring made of the RAA at the front on the right, LBCA at the front on the left and Kommerell’s diverticulum at the back on the left (figure 4). This finding suggested a fully closed vascular ring.
increased total cholesterol (248 mg·dL−1), of low-density lipoprotein–cholesterol (163 mg·dL−1) and triglycerides (178 mg·dL−1).

The spirometric examination repeated 1 month later, when bronchitic symptoms had fully disappeared, confirmed the same pattern as the first observation (figure 6 and table 2).

Regarding therapeutic management, given the absence of disabling chronic symptoms due to the compression of trachea and oesophagus and, more specifically, the absence of wheezing, dyspnoea and dysphagia, we preferred a conservative approach with a multidisciplinary annual clinical–instrumental monitoring involving the pneumologist, cardiologist and radiologist.

### Discussion

Spirometry is not simply an examination to measure breath; a qualitative assessment of the shape of the flow–volume curve by itself can suggest diseases involving the respiratory system but originating from other organs [1, 2]. We describe a case of central airway obstruction due to a congenital malformation of the thoracic aorta.

Upper and central airway obstruction, i.e. between pharynx and main bronchi, is an infrequent but potentially life-threatening condition. Its course may be acute, with the risk of death in a short time, or chronic, often diagnosed only after a long time when reaching a significant degree of anatomical obstruction or in cases of respiratory tract infections, as in our case [3]. The diagnosis of upper airway obstruction with a chronic course may also be delayed due to the similar clinical presentation to asthma and COPD.

The first suspicion could come from a careful assessment of the flow–volume curve. At least three maximal and repeatable forced inspiratory and forced expiratory flow–volume curves are necessary to assess the presence of central or upper airway obstruction [4].

There are three types of spirometric pattern suggesting upper and central airway obstruction: the extrathoracic variable obstruction, the intrathoracic variable obstruction and the fixed airway obstruction [4, 5]. In the first type, there is only a flattening of the inspiratory part of the curve due to a constant flow reduction; in the second, we have only a flattening of the expiratory part of the curve; in the third one, a flattening of both inspiratory and expiratory portions of the loop can be detected, as in our case. The type of curve depends on two features:

- the behaviour of the lesion during inspiration and expiration, with the lumen narrowing.
only during one ventilatory phase (variable inspiratory or expiratory obstruction) or both (fixed obstruction).

- the intra- and extra-airway pressures fluctuation during the forced manoeuvre [5].

In addition to the qualitative evaluation of the loop, the presence of upper and central airway obstruction may be detected by several quantitative methods: FIV1/FEV1 ratio; FEV1/PEF ratio; MEF50%/MIF50% ratio; MIF50% <100 L·min⁻¹; and FEV1/forced expiratory volume in 0.5 s (FEV0.5) >1.5 [6–8].

The FIV1/FEV1 ratio is not seriously affected by a variability of effort, and is a satisfactory measurement to define upper and central airway obstruction. FIV1 and FEV1 are nearly the same in fixed airway obstruction, and therefore, the ratio is close to 1. In variable extrathoracic airway obstruction, FEV1 is considerably lower than the FIV1 and therefore, the ratio is <1. Conversely, in intrathoracic variable obstruction, FEV1 is considerably lower than the FIV1 and therefore the ratio is >1. FIV1/FEV1 >1 may be also seen in patients with COPD and chronic asthma.

A FEV1/PEF ratio >10 suggests central or upper airway obstruction may be present. According to Miller and Hyatt [7], MEF50%/MIF50% is helpful in recognising fixed (MEF50%/MIF50% ~1), variable extrathoracic (MEF50%/MIF50% >1) and variable intrathoracic (MEF50%/MIF50% <0.3) airway obstruction.

Moreover, upper and central airway obstruction may be suspected if MIF50% <100 L·min⁻¹ and FEV1/FEV0.5 >1.5.

Regarding airway resistance, the specific airway resistance loop has an S-deformation (closed) in patients with upper airway obstruction [9]. Moreover, impairment of respiration can be assumed if, in an adult, airway resistance during spontaneous breathing is >6 cmH2O·L⁻¹·s⁻¹ during inspiration and/or expiration [10].

However, to date no single “gold standard” technique is currently used in routine clinical practice, and visual and quantitative diagnostic criteria have low sensitivity; therefore, clinicians should refer suspected cases for further diagnostic evaluation even in the presence of normal spirometry [11, 12].

Malformations of the aortic arch account for 1–3% of congenital cardiopathies [13]. Among the most frequent are the double aortic arch and the RAA, with multiple branching subtypes of the main vessels and the eventual association with congenital cardiopathies [14, 15]. During embryogenesis, the RAA develops from the abnormal regression of the left aortic arch. However, in the event where both left and right arches persist, one has the abnormal double aortic arch. The incidence of the RAA in symptomatic patients, the first examination is chest radiography. If the vascular ring is complete, the trachea does not get dislocated on the opposite
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side but keeps its normal site because of the presence of the ligamentum arteriosum or remnant fibrous arch. On the lateral view, one can observe an increased density behind the trachea, a focal narrowing of the trachea or an anterior curvature of the lower portion of trachea, rather than of the normal posterior curvature. The definition of the anatomy of the vascular ring and the relationship with the surrounding structures requires CT or contrast-enhanced MRI, but CT is preferable given that it allows a more accurate characterisation of the Airways [15].

In the presence of respiratory symptoms, the patient should be directed to the pneumologist for spirometry. The pattern observable in the event of a complete vascular ring is that of a fixed obstruction, as shown in our case. Echocardiography could be used for the assessment of the aorta and the exclusion of simultaneous congenital cardiac malformations. Additionally, in some cases, it could be useful to perform fibreoptic bronchoscopy for a better assessment of the degree of tracheal stenosis.

Available treatment is based on surgery and it is indicated only in clinically significative cases. It consists of cutting the ligamentum arteriosum, which compresses the trachea and the oesophagus, while the Kommerell’s diverticulum has to be resected at its origin [27–29].

Conclusions

Physicians involved in the management of spirometry should be aware of vascular anomalies that could cause tracheal stenosis. In our case report, we have described a tracheal stenosis with fixed airway obstruction on spirometry due to a complete vascular ring, composed of a RAA with a mirror image of the originating arterial branches (type I of Edwards’ classification) on the front and concomitant type III Kommerell’s diverticulum on the back. Vice versa, it is advised that the echocardiographic finding of a congenital aortic malformation by a cardiologist should be followed by spirometric evaluation.

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Conflict of interest

G. Castellana has nothing to disclose. R. Castellana has nothing to disclose. R. Castellana has nothing to disclose. C. Castellana has nothing to disclose. P. Verde has nothing to disclose. G. Castellana has nothing to disclose.

References

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