Recurrence respiratory infections caused by a double aortic arch: The diagnostic role of spirometry

Cecilia Calabrese, Nadia Corcione, Valentina Di Spirito, Carmine Guarino, Giovanni Rossi, Gaetano Domenico Gargiulo, Alessandro Vatrella

Abstract

A young woman with a clinical history characterized by recurrent respiratory infections, occurring since early infancy, was referred to our hospital. When the patient was a young girl, she underwent sweat chloride test, serum analysis of immunoglobulins, and evaluation of blood lymphocyte subsets; all these diagnostic tests were normal, as well as chest X ray aside from pneumonia episodes. Skin prick tests were positive for several different allergens, and a diagnosis of allergic rhinitis was made. At the age of 11 years, she started to complain of gastroesophageal reflux disease (GERD) symptoms, and a gastroscopy detected a hiatal hernia with esophagitis. Despite pharmacologic treatments for allergic rhinitis and GERD, the patient continued to complain of chronic cough, associated with choking and recurrent respiratory infections treated with antibiotic therapy. For the first time in her life, we performed a spirometry that showed a flow-volume curve characterized by a plateau in the expiratory phase, suggestive of an central airway obstruction. Bronchoscopy demonstrated a compression of the distal portion of trachea. Computed tomography (CT) angiogram revealed a double aortic arch. Barium enhancement evidenced an esophageal compression. A surgical division of the smaller of the two arches was then performed. Therefore, we strongly suggest to perform lung function tests in all cases of unexplained respiratory complaints.

1. Introduction

Double aortic arch (DAA) is a congenital defect of embryonic aorta development, due to the persistence of the fourth right and left arches and dorsal aortas, resulting in the abnormal formation of complete vascular rings encircling trachea and esophagus. Therefore, DAA causes respiratory and digestive symptoms, whose severity and age of presentation depend on the degree of extrinsic compression. While respiratory complaints like cough, stridor, dyspnea and recurrent pneumonias are prevalent during early infancy, those due to esophageal compression such as dysphagia and choking occur later. The real prevalence of DAA in adult life is unknown, and 25 cases are cited in a comprehensive literature review. In adults, DAA is often misdiagnosed and confused with difficult-to-control asthma. Here we present a case of a young woman with a clinical history of recurrent respiratory infections and gastro-esophageal reflux symptoms, in whom spirometry guided the diagnosis of DAA. We emphasize the importance of an early execution of pulmonary function tests in every case of unexplained respiratory symptoms.

2. Case report

A 19-year-old woman was referred to our hospital for a clinical history mainly characterized, since early infancy, by recurrent mono- and bilateral pneumonias often requiring hospital admission. When the patient was a young girl, she underwent sweat chloride test, serum analysis of immunoglobulins, and evaluation of blood lymphocyte subsets; all these diagnostic tests were normal. Apart from the recurrent pneumonia episodes, chest X ray did not
show any significant abnormality. Skin prick tests were positive for house dust mite and parietaria, and a diagnosis of allergic rhinitis was made. Antihistamines and inhaled steroids were prescribed, but the patient continued to suffer from recurrent respiratory infections, requiring frequent courses of antibiotic therapy. At the age of 11 years, the young patient started to complain also of gastro-esophageal reflux disease (GERD) symptoms, and a gastroscopy detected a hiatal hernia with a second grade esophagitis. Despite pharmacologic treatment of GERD, she was frequently admitted to the emergency room for episodes of cough associated with choking and vomiting.

On admission, the patient complained of fever and cough with mucous sputum. Physical examination revealed the presence of fine, sparse inspiratory crackles at the level of the right lower third of chest. Laboratory tests were normal, except for a mild neutrophilic leukocytosis and an increase of C reactive protein (CRP) serum levels. Chest X ray was normal. Computed tomography (CT) scan of paranasal sinuses showed a hypertrophy of lower turbinates, and a surgical treatment was thus planned. For the first time in her life, the patient performed a spirometry: the flow-volume curve was characterized by a plateau in the expiratory phase, suggestive of a central airway obstruction (Fig. 1). In order to detect the underlying cause of such impaired ventilation, we carried out a bronchoscopy that showed a compression of the distal portion of trachea (Fig. 2). CT angiogram revealed a vascular ring, consisting of a double aortic arch with a right-arch dominance, compressing the trachea and esophagus (Fig. 3). The echocardiogram confirmed the presence of the vascular ring, not associated with any cardiac defect. Barium enhancement of the esophagus showed an esophageal compression (Fig. 4). The patient underwent surgery via thoracotomy and a division of the smaller, left arch was performed, at a site not affecting blood flow through common carotid artery. After surgery, her clinical conditions significantly improved. At follow-up evaluation performed one year after surgery, the patient resulted to be free of respiratory problems and pulmonary infections, although spirometry revealed the persistence of a mild, variable intrathoracic airway obstruction. She rarely complains of digestive symptoms.

3. Discussion

To the best of our knowledge, this is the first case reported in literature of an adult patient with a clinical history of recurrent respiratory infections caused by the presence of a DAA. Despite the early occurrence of pneumonia, later associated with upper digestive symptoms, the existence of a vascular ring was not suspected until a spirometry was performed at the age of 19 years. Overall, this report suggests that an impaired clearance of secretions at the level of an extrinsic airway obstruction can cause mucus accumulation, predisposing to bacterial growth that in turn produces inflammatory changes which worsen the obstructive respiratory
pattern, thus triggering a vicious circle between airway obstruction and lung infection.

Vascular rings are rare congenital vascular disorders, causing a complete or partial encirclement of trachea and esophagus. An early monograph from the Mayo Clinic divided vascular rings into 7 types, but over 95% of cases can be classified in 4 main categories: double aortic arch, right arch/left ligament, innominate artery compression and pulmonary artery sling. DAA is the most common form of complete vascular ring, which originates when the ascending aorta bifurcates into a right and a left arch, both usually patent; these arches encompass trachea and esophagus, and rejoin in the thoracic descending aorta. Symptoms such as stridor, persistent cough, wheezing, recurrent respiratory infections, vomiting and dysphagia usually lead to diagnosis and surgical treatment in early childhood. Indeed, 5-months is the median age of presentation. However, because of only a few published reports, the real prevalence of vascular rings in adults is unknown. Dysphagia is the prevalent presenting symptom, but also a clinical history of chronic cough and/or dyspnea is quite frequent; moreover, some asymptomatic cases can be incidentally diagnosed. Pediatricians and cardiologists are usually aware of this clinical condition, but this is not always true for pulmonologists and gastroenterologists managing adult patients. A delay in diagnosis and treatment can be responsible for several late complications, such as tracheomalacia or aortoesophageal fistula.

With regard to the present report, the typical spirometric pattern of a central airway obstruction led to the diagnosis of DAA. Similarly, available literature includes several cases of patients misdiagnosed as asthmatics until the execution of pulmonary function tests. Vascular rings may determine either a plateau on the expiratory limb of the flow-volume curve, indicative of a variable intrathoracic airway obstruction, or a flattening of both the

Fig. 3. Computed tomography angiogram with the three-dimensional reconstruction revealed a vascular ring consisting of a double aortic arch with right-arch dominance compressing the trachea and esophagus.

Fig. 4. The barium esophagography showed esophageal compression.
expiratory and inspiratory arms of the curve, consistent with a fixed intrathoracic airway obstruction. This condition does not usually lead to a decrease in FEV\(_1\) and/or VC, but PEF can be severely impaired, thus producing a ratio of FEV\(_1\) divided by PEF greater than 8 mL/L/min. Poor initial effort can also affect this ratio; therefore, it is of paramount importance to obtain the optimal patient collaboration during spirometry performance. Efforts must be maximal and repeatable in order to accurately evaluate flow-volume curve morphology, but of course it is particularly difficult to achieve this goal in the pediatric population. Current guidelines recommend that spirometry should be performed also in preschool children but, as reported in the present case, its execution is often delayed.\(^5\) Therefore, we strongly suggest to perform spirometry as soon as possible in all cases of unexplained respiratory symptoms, refractory to pharmacologic treatments. Persistent cough, stridor, wheezing, dyspnea, noisy breathing and recurrent respiratory infections can be related to abnormalities involving central airways, not always detectable by chest X ray. DAA may be suspected if a frontal chest radiograph shows the presence of either an aortic knob projecting over the right side of sternal manubrium, or a narrowing/deviation/indentation of tracheal shadow. However, as observed in our patient, chest radiogram can also be normal.\(^1,2\) In adults, DAA often causes upper digestive symptoms as choking with food, dysphagia and vomiting that, whether unexplained and associated with respiratory problems, could be also investigated by barium swallow.\(^3,9,10,21\) Echocardiography may be useful to diagnose vascular rings, but cardiologists are often consulted lately in the management of patients complaining of either respiratory or digestive symptoms. According to ATS guidelines, when flow-volume loops alert clinicians with regard to the possible existence of a central or upper airway obstruction, endoscopic and radiological approaches represent the next steps to further investigate this functional respiratory impairment.\(^1,9,10,21\) Laryngoscopy and bronchoscopy usually document the extrinsic compression, but chest CT scan represents the best diagnostic technique because it evidences the airway lumen as well as the anatomy of the vascular ring. When evaluation of blood flow is needed, especially in case of surgical planning, angiography and magnetic resonance imaging (MRI) can provide further useful information.

In patients with DAA, spirometry is usually performed only in those subjects complaining of persistent respiratory symptoms, despite the surgical treatment of vascular rings. These cases are mostly associated with a condition of tracheomalacia due to the prolonged tracheal compression caused by the vascular ring.\(^2,5,22–24\) Recently, in a group of infants with a prenatal echocardiographic diagnosis of aortic arch abnormalities, neonatal lung function tests were able to diagnose airway obstruction and to support the rational choice of a surgical therapeutic approach, before the occurrence of clinical manifestations.\(^22\) When compared with other diagnostic tests used to assess airway obstruction, including CT scan, MRI and bronchoscopy, spirometry results to be a sensitive, less expensive, non invasive and non radiating tool. Therefore, spirometry should be performed in all patients affected by DAA within the context of a pre-surgical setting aimed to evaluate the degree of pulmonary function impairment, as well as in follow-up post-surgical evaluations, in order to assess lung function recovery.

In conclusion, we strongly suggest to perform lung function tests in all patients complaining of unexplained respiratory symptoms, resistant to pharmacologic treatments.

Conflicts of interest

All authors, Calabrese Cecilia, Corcione Nadia, Di Spirito Valentina, Guarino Carmine, Rossi Giovanni, Gargiulo Gaetano Domenico, and Vatrella Alessandro, state that there are no conflicts of interest in connection with this article.

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