Pulmonary function test values due to a diaphragmatic hernia

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DESCRIPTION

A 67-year-old never-smoking man with a well-known congenital diaphragmatic hernia, verified on a 3-year-old chest X-ray (figure 1), consulted his general practitioner due to the detection of intermittent blood stool for 2 months, but emphasised also an increasing dyspnoea during this period corresponding to a medical research council dyspnoea scale 3. He was referred for colonoscopy, but prior to this a CT scan of the thorax and abdomen was performed showing progression of the left-sided diaphragmatic hernia with now almost the entire small intestine localised in the left hemithorax (figure 2). Despite the radiological findings, the patient had no symptoms from the gastrointestinal tract, and colonoscopy only revealed a non-malignant colon polyp as the most reliable cause of blood stool. The patient was subsequently referred for pulmonary evaluation where a pulmonary function test (PFT) including single-breath diffusion lung capacity for carbon monoxide showed a reduced diffusion capacity of 63% of predicted value (pred.), a hyper normal diffusion coefficient (KCO) corresponding to 160% pred., and a severe restriction in ventilation capacity with a forced expiratory volume in 1 s (FEV_1), a forced vital capacity (FVC) and a FEV_1/FVC corresponding to 39% pred., 38% pred. and 80% pred., respectively (figure 3).[1] No static lung function measurements were performed.

Figure 1 Chest X-ray in posterior to anterior projection performed 3 years earlier.

DLCO SB	[mmol/min/kPa]	8.10	4.29	53.0 %
DLCO/VA	[mmol/min/kPa/L]	1.29	1.74	134.4 %
VA	[L]	6.11	2.47	40.4 %
DLCOc SB	[mmol/min/kPa]	8.10	5.11	63.1 %
DLCOc/VA	[mmol/min/kPa/L]	1.29	2.07	160.0 %
Hb	[mmol/L]		6.20	
FEV 1	[L]	2.75	1.07	38.8 %
FVC	[L]	3.54	1.34	37.8 %
FEV 1 % FVC	[%]		79.61	

Figure 3 Spirometry data including forced expiratory ventilation in 1 s (FEV_1), forced vital capacity (FVC), single-breath (SB) haemoglobin corrected (c) diffusion lung capacity for carbon monoxide (DLCO) showing severe restriction in ventilation capacity and, when adjusting for alveolar volume (VA), a normal diffusion coefficient (DLCO/VA=KCO). Values in per cent correspond to per cent of predicted values.

Diaphragmatic hernia is a well-known cause of restrictive ventilation pattern on PFT which is also present in interstitial lung diseases (ILDs) and secondary to a pneumonectomy. However, a reduced KCO is expected in ILD unlike a normal and a sometimes even slighter increase in KCO in cases presenting with diaphragmatic hernia or pneumonectomy, respectively.[2 3] In such cases the specific

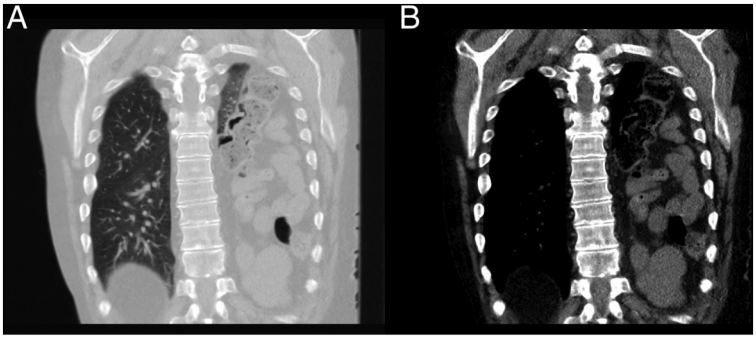

Figure 2 CT image of the thorax showing an enormously left-sided diaphragmatic hernia with almost the entire small intestine localised in the left hemithorax ((A) lung window; (B) chest–abdominal window).

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combination of ventilation and diffusion parameters from a PFT may indicate a potential underlying aetiology of the respiratory condition which may be uncovered and clarified by thoracic imaging.

Learning points

- Diaphragmatic hernias can lead to an intrathoracic space-occupying lesion causing a restrictive lung disease pattern with an oftentimes normal diffusion coefficient.
- An unexplained restrictive ventilation pattern should be followed up by thoracic imaging in order to uncover the potential underlying aetiology.

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