As EDAC is a condition in which airways collapse during forced expiration and cough, treatment modalities for airway clearance that increase inspiratory volume or expiratory flow rate should not be considered.

We tried High Frequency Chest Wall Oscillator (HFCWO – Vest® System, Model 105 – Home Care, HillRom), an inflatable jacket (Vest) worn over the chest connected to an air pulse generator which rapidly inflates and deflates the vest producing oscillations to the chest wall.

The air pulse generator was set at pressure level 2 and frequency 14.

HCFWO alone caused worsening of dyspnea probably because it decreases End Expiratory Lung Volume below the Residual Functional Capacity[6] increasing airway resistance. Thus, as suggested by Perry et al.[7], we added a positive expiratory pressure using tPEP set at 8 cmH_2O to maintain airway opened during the oscillations.

While Perry[7] used a pressure-cycled ventilator to add a PEEP, we used a tPEP with a mouthpiece to make the treatment easier and more feasible at home for the patient and to facilitate secretions expulsion (which can be difficult when wearing a facial mask). This made the HFCWO very effective and JAT continued the treatment 3 times per day 15 min/sessions, achieving stable airway cleaning and improvement of dyspnea.

During the airway clearance program SpO_2 was monitored: it did not change from the basal values (i.e. 96–98%).

At discharge The Borg scale of dyspnea improved from 8 to 4 and she declared: ''I feel my lungs free of secretion, during the treatment I feel secretions moving upward'' The Quality of Life score (EQ-5D) improved from 50 to 85.

No modifications were observed in lung function. HFCWO and tPEP were prescribed at home 3 times per day every day. She kept to the treatment and after 48-month follow-up was still improved. JAT had no side effect from treatment, no more admissions for respiratory symptoms and no need for bronchoscopy.

Even if this tool is not a treatment for the disease, it ensured the patient kept her lungs clear from secretions and improved her dyspnea and quality of life.

We did not study the effects of HFCWO on respiratory system mechanics of this patient and, as EDAC is a rare disease, we could not conduct a randomized trial. Other studies and more patients are needed to verify the effectiveness of this intervention in EDAC.

However, we think that the association of HFCWO and tPEP should be taken into consideration when any other treatment fails with an EDAC patient. This is, in fact, a non-invasive and safe intervention that can be used at home in a self-care regime.

Conflicts of interest

The authors have no conflict of interest to declare.

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E. Bertella*, C. Simonelli, L. Bianchi

Respiratory Rehabilitation Division, Salvatore Maugeri Foundation, IRCCS – Institute of Lumezzane (Brescia), Italy

*Corresponding author.
E-mail address: enrica.bertella@fsm.it (E. Bertella).

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Systemic hypertension and the pathogenesis of diffuse alveolar haemorrhage

To the Editor,

Diffuse alveolar haemorrhage (DAH) is a rare, life-threatening syndrome, characterized by progressive alveolar infiltrates, anaemia and haemoptysis. DAH is a medical emergency, requiring prompt identification and treatment, but this may be hindered by an absence of haemoptysis or non-acute presentation. A comprehensive search for the underlying cause is imperative, as this will guide any effective treatment. It is important to note that different aetiologies are associated with specific pathogenesis. We describe the case of a patient with malignant

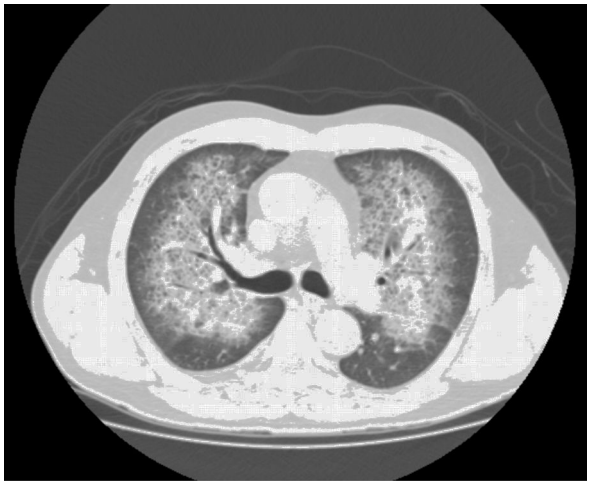

Figure 1 Non-contrast thoracic CT scan: Bilateral central parenchymal infiltrates with a ''crazy paving'' pattern.

hypertension (MH) presenting with DAH, we review previous reports on this association and discuss the implications on the pathogenesis and clinical management of DAH.

A 51-year-old male presented with a dry cough and mild dyspnoea which had lasted four days. He was a financial consultant, a heavy smoker and had a history of chronic sinusitis but without any prescribed medication. The physical examination revealed no signs of respiratory distress, a bilateral blood pressure (BP) of 220//130 mmHg, and left basal crackles. An echocardiography revealed signs of mild aortic, mitral and tricuspid regurgitation and a chest X-ray showed bilateral perihilar ''butterfly pattern'' alveolar infiltrates. Blood tests showed severe renal failure, hypokalaemia and normocytic anaemia (creatinine: 8.02 mg/dL (reference values (RV): 0.72–1.18), potassium: 2.0 mmol/L (RV: 3.5–5.1), haemoglobin: 11.2 g/dL (RV: 13.0–17.0)). Laboratory coagulation tests were normal (prothrombin time: 16.2 s (RV: 12.0–17.4), prothrombin 77% (RV: 70–120)). An arterial blood gas analysis showed respiratory alkalosis with a PaO2 level of 63 mmHg (RV 83–108). A non-contrast chest CT found bilateral central parenchymal infiltrates with a ''crazy paving'' pattern (Fig. 1) and a renal ultrasound found bilateral increased kidney echogenicity with preserved size. A bronchoscopy with bronchoalveolar lavage was performed, and progressively bloody fluid was retrieved. Urinalysis showed microscopic haematuria and proteinuria (+++) with a spot urine sample protein to creatinine ratio of 2380 mg/g (RV: <200). Fundoscopic examination identified signs of hypertensive optic neuropathy, and the patient was admitted for malignant hypertension, under aggressive anti-hypertensive therapy. He remained stable and the pulmonary infiltrates improved, but poor renal function and difficult hypertension continued. Further study revealed cytoplasmic iron deposits in alveolar macrophages with a moderate alveolar haemorrhage but no signs of pulmonary infection. The auto-immune panel was negative, including anti-nuclear antibodies, proteinase-3-antineutrophil cytoplasmic antibodies, myeloperoxidase-antineutrophil cytoplasmic antibodies, anti-glomerular basement membrane antibodies, cryoglobulins and rheumatoid factors. Complement levels were normal and viral serologies were negative. The plasma renin was 1090 μU/ml (RV: 7–76), aldosterone was 5660 pg/ml (RV: 30–310), and aldosterone to renin activity ratio was 6.3 ng/ng (RV: 14.5). An abdominal CT scan revealed a left adrenal nodule and a renal eco Doppler excluded renal artery stenosis. An ultrasound guided kidney biopsy was performed revealing hypertensive nephroangiosclerosis with chronic interstitial nephritis and arterio-sclerosis and arteriolo-sclerosis. There was no evidence of active necrotizing glomerulonephritis, crescent formation, immune deposits or vasculitis. The procedure was complicated by late bleeding and renal haematoma requiring embolization of the left renal artery. After the embolization, normalization of blood pressure and potassium levels were noted. A new evaluation of plasma renin and aldosterone levels showed normal results. The patient became asymptomatic; there was complete resolution of pulmonary infiltrates and hypoxaemia, but no recovery of renal function, requiring chronic haemodialysis. The final diagnosis was probable adrenal reninoma, leading to malignant hypertension, diffuse alveolar haemorrhage and chronic renal failure.

We report a patient with a probable reninoma leading to severe systemic hypertension and subsequent diffuse alveolar haemorrhage. Although we could not get histological confirmation of the reninoma, the left adrenal node, strikingly elevated levels of renin and prompt pressure response to the embolization procedure suggest that this was the cause of the hypertension. Malignant hypertension can be defined as extremely high blood pressure with associated papilledema. The pathogenesis of MH is poorly understood but disturbances in the renal release of vasoconstrictors and mechanical stress on vessels' walls appear to be critical. This is the seventh report on malignant hypertension-associated DAH. The main characteristics of this one and of the previous reports are described in Table 1. This possible new syndrome displays some striking characteristics: all patients are, relatively young, males, with 4 out of 7 were smokers. There was also a predominance of Asians. Since the incidence of malignant hypertension is not increased in males, there may be some relevance to this gender predominance.

These reports suggest that extreme hypertension can lead to pulmonary haemorrhage, similar to pulmonary renal syndrome caused by an immune-mediated mechanism. The mechanism of how high blood pressure can cause alveolar haemorrhage remains unclear, but humoral factors[1] might be involved in the alveolar capillaries. The damaged endothelium increases permeability and activates the coagulation cascade, including platelet activation and fibrin deposition. Red blood cells are destroyed within vessels,[2] which may explain why up to one-third of patients do not present with haemoptysis. Hida et al.,[1] argued that alveolar capillaries might be injured by malignant hypertension in the same way as systemic capillaries. However, Sato et al.[3] and Aithal et al.[4] concluded that left ventricular dysfunction resulting from systemic hypertension can cause pulmonary oedema, leading to haemorrhage. On the other hand, Dalal et al.[5] showed that smoking, platelet dysfunction due to underling uraemia and thrombocytopenia may also predispose to development of pulmonary haemorrhage. Sato et al.[3] and Dalal et al.[5] suggest that previous air travel may have contributed to the development of DAH, with an underlying mechanism similar to high altitude pulmonary oedema.

Table 1 Clinical characteristics of all reports of malignant hypertension with diffuse alveolar haemorrhage.

Case	Age	Gender	Nationality	Smoking	Previous HT	Family history of HT	Air travel	Ref.
1	34	Male	Japanese	NR	1 month	NR	NR	1
2	26	Male	North American	1–2 cig./day	No	Both parents	Yes	5
3	26	Male	Japanese	NR	3 years	NR	Yes	3
4	38	Male	Canadian	9 packs-year	3 months	NR	NR	4
5	32	Male	Japanese	NR	5 years	Both parents	NR	6
6	27	Male	Japanese	>30 pack-year	No	NR	NR	2
7	**51**	**Male**	**Portuguese**	**>30 pack-year**	**No**	**Mother**	**No**	**–**

HT: systemic hypertension; cig.: cigarette; NR: no reference; Ref.: reference.

However, it is worth noting that no other findings have supported this hypothesis, and this may be just a non-significant association. In conclusion, our and previous reports of malignant hypertension associated DAH suggest that high blood pressure may be a significant cause of alveolar haemorrhage. Clinicians caring for DAH patients should be aware of this association. We also propose large-scale studies on the association between systemic blood pressure and the risk of alveolar haemorrhage which could open the way to new treatment of DAH with BP lowering agent.

Disclosure

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Conflicts of interest

The authors have no conflicts of interest to declare.

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Joana Clemente Duarte[a,*], Rita Leal[b], Luís Escada[b], Tiago M. Alfaro[a]

[a] *Pneumology Unit A, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal*
[b] *Nephrology Unit, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal*

* Corresponding author.
E-mail address: Joana_Duarte_17@hotmail.com (J.C. Duarte).

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Mesothelial cyst of the pulmonary ligament: An uncommon lesion

Dear Editor,

The importance of a thorough knowledge of thoracic anatomy is timeless. Lung and pleural anatomy continues to be a subject for study nowadays, not only due to the greater spatial resolution of newer diagnostic studies, such as bronchoscopy, computed tomography and magnetic resonance, but also because of advances in thoracic surgery.

The pulmonary ligaments are among the lesser known structures of the thoracic anatomy. The radiological importance of the pulmonary ligament was first described in 1966 by Rabinowitz and Wolfe,[1] and only very few reports or series have been found which describe the radiological findings of pathologic processes within the pulmonary ligaments.[1–6] This ligament, just like any other thoracic anatomical structure can be affected by non-infectious or infectious inflammatory processes, tumors, congenital or developmental abnormalities and scarring.[2–6] There are only a few reports in the literature which mention this anatomical structure and different pathological process.

The following case description of a cystic lesion within the right pulmonary ligament illustrates the anatomy and pathology of this structure:

A 75-year-old woman diagnosed with a pharyngeal cancer was referred for magnetic resonance imaging of the neck and a computed tomography (CT) of the chest and