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CASE REPORT

Unilateral Pulmonary Oedema secondary to Anterior Mitral Valve Leaflet Prolapse

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Cardiogenic unilateral pulmonary oedema (UPE) is an uncommon clinical condition which is often mistaken for lower respiratory tract infection (LRTI). It is typically caused by asymmetric systolic flow reversal in the right pulmonary veins due to severe mitral valve regurgitation (MR). We describe the case of a 41-year-old woman with a history of undifferentiated chronic mixed connective tissue disease, Raynaud's disease and pituitary adenoma who presented with acute shortness of breath (SOB), chest tightness, orthopnoea and paroxysmal nocturnal dyspnoea. A computerized tomography pulmonary angiogram (CTPA) showed signs consistent with pulmonary oedema secondary to congestive heart failure. This prompted urgent echocardiography which revealed a flail and prolapsing A2 leaflet of the mitral valve with a posteriorly-directed MR jet, resulting in systolic flow reversal in the pulmonary veins which is consistent with severe MR. The patient underwent an urgent mitral valve (MV) replacement and optimization of medical therapy, resulting in clinical and radiographic improvement.

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Cardiogenic unilateral pulmonary oedema (UPE) is an uncommon clinical condition which is often misdiagnosed for presumed lower respiratory tract infection (LRTI). Diagnostic challenges carry an increased risk of morbidity and mortality. UPE is commonly due to asymmetric systolic flow reversal in the right pulmonary veins caused by severe mitral valve regurgitation.

CASE PRESENTATION

We describe the case of a 41-year-old Caucasian woman who is a known case of undifferentiated chronic mixed connective tissue disease, Raynaud's disease and pituitary adenoma. She presented to the emergency department with acute worsening of shortness of breath (SOB) that had started a few days prior. This was worse at night and on exertion. She also complained of occasional chest tightness, orthopnoea and paroxysmal nocturnal dyspnoea (PND).

Salient clinical findings on examination included pallor, lethargy, restlessness and SOB. The patient had a blood pressure of 109/75 mmHg, regular pulse of 110 bpm, afebrile body temperature and oxygen saturations of 88% on room air. Lung auscultation revealed bibasal coarse crepitations which were more prominent over the right mid zone. Heart sounds were unremarkable. Chest X-ray showed bilateral airspace shadowing, more prominent on the right (Figure 1). Laboratory investigations showed a white cell count (WCC) of 10.64 x10^9/L with neutrophils of 7.71 x10^9/L, haemoglobin of 11.9 g/dL, creatinine of 46 µmol/L, C-Reactive Protein (CRP) of 32.4 mg/L, B-natriuretic peptide of 2867 pg/mL and no acute troponin rise from 11 to 7 ng/L. Serum electrolytes were within normal limits.

In the context of these findings, the patient was admitted to hospital and started on intravenous Co-Amoxiclav and Clarithromycin to treat a suspected pneumonia. On day two of admission, the patient complained of worsening SOB and pleuritic chest pain. A D-dimer was taken which was found to be 1373 ng/mL. Subsequently, a computerized tomography pulmonary angiogram (CTPA) was performed. This showed a well-defined area of ground glass opacification in the right lower lung zone, small bilateral pleural effusions and cardiomegaly which all demonstrate signs consistent with pulmonary oedema secondary to congestive heart failure (Figure 2). A transthoracic echocardiogram was then performed which revealed acute mitral regurgitation (MR) secondary to anterior mitral valve prolapse. In order to better visualise the anatomy, a transoesophageal echocardiogram (TOE)

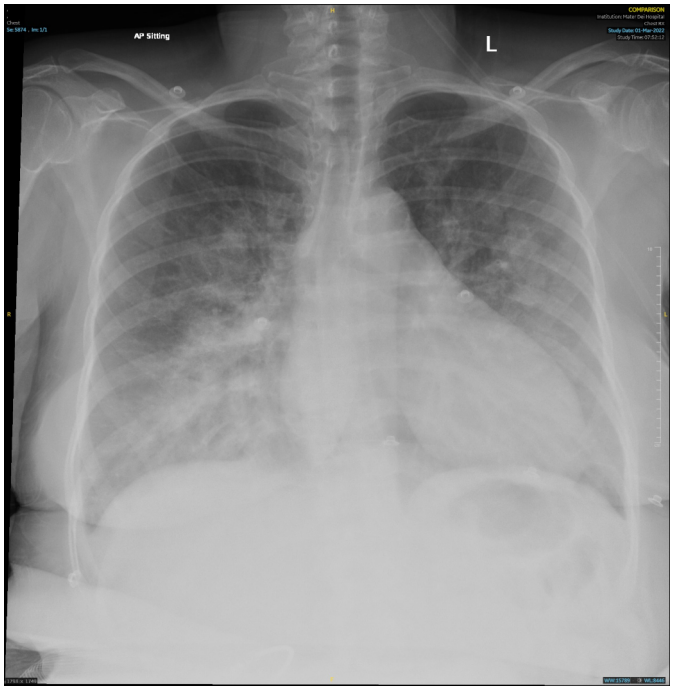

Figure 1 *An anteroposterior chest X-ray taken on admission showing bilateral airspace shadowing more prominent on the right*

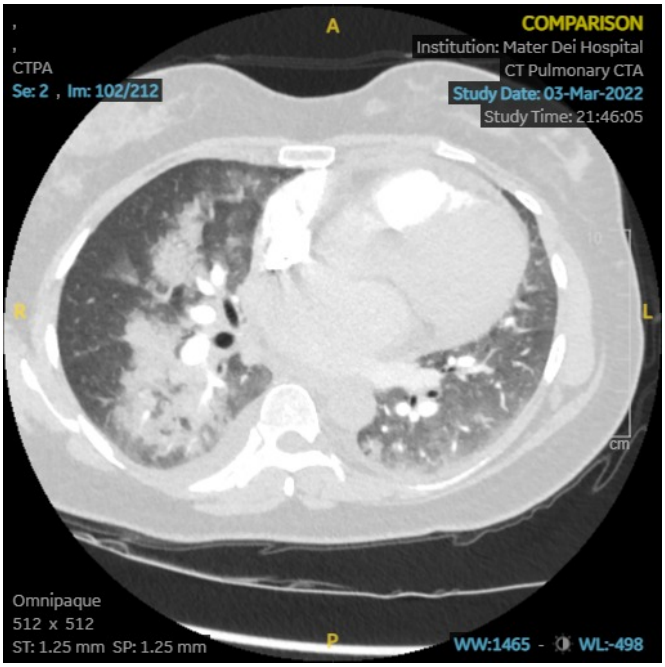

Figure 2 *An axial view of a CTPA showing well-defined area of ground glass opacification in the right lower lung, interlobular septal thickening and bronchovascular bundle thickening. All signs are consistent with pulmonary oedema secondary to congestive heart failure.*

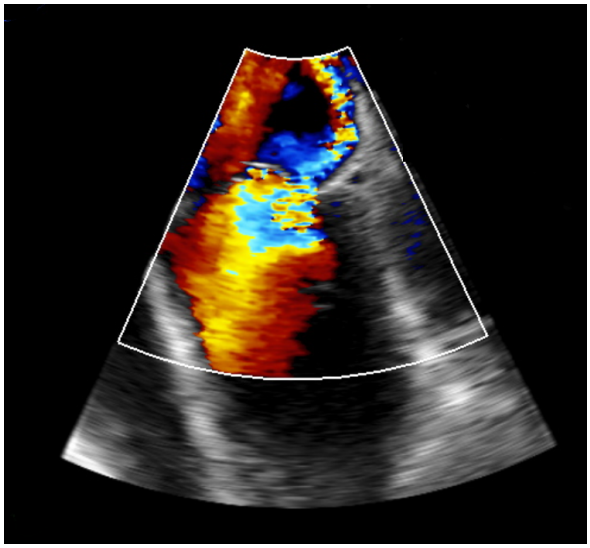

Figure 3 *A still image from the TOE showing regurgitant blood flowing into the left atrium secondary to a posteriorly-directed MR jet*

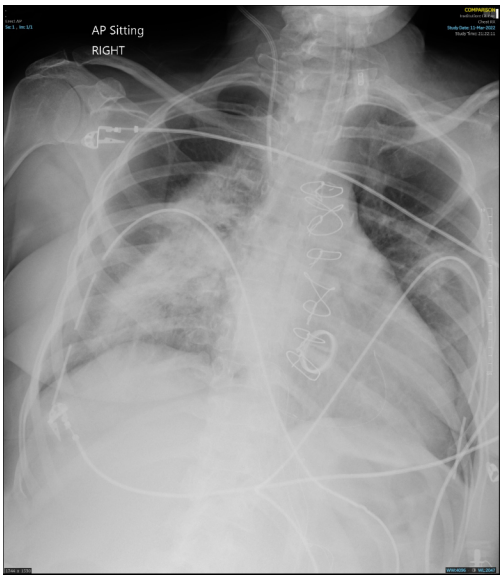

Figure 4 *An anteroposterior chest X-ray post mechanical MV replacement showing midline sternotomy wires and clear demarcation of a right sided UPE*

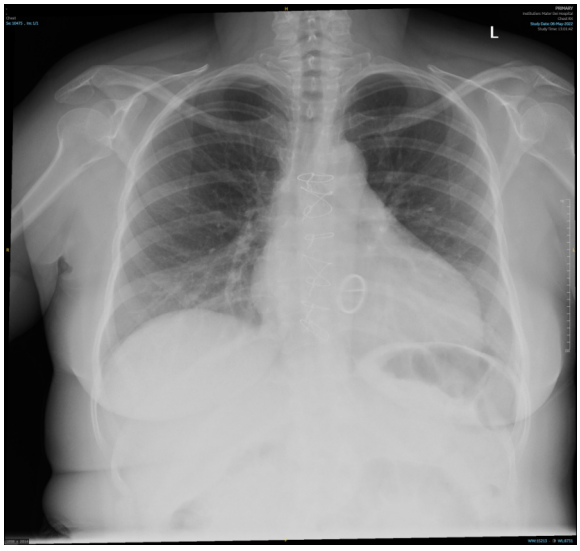

Figure 5 *A six-week follow-up anteroposterior chest X-ray showing complete resolution of right sided pulmonary oedema, mechanical MV replacement and midline sternotomy wires*

was done. This showed a mildly dilated left ventricle with good systolic function and a normal sized right ventricle with good function. It also confirmed the presence of a flail and prolapsing A2 leaflet of the mitral valve with a posteriorly-directed MR jet (Figure 3). This resulted in systolic flow reversal in the pulmonary veins which is consistent with severe MR (grade of ≥3/4). There were no vegetations visible on the cardiac valves.

In light of these findings, the patient underwent an urgent mitral valve (MV) replacement. Adjustments to the patient's medical treatment were done and repeat chest X-rays post-operatively showed improving right-sided lower lobe pulmonary oedema (Figure 4, Figure 5).

DISCUSSION

Literature to date has shown that UPE is strongly associated with severe organic or functional mitral valve regurgitation. Due to its rarity and atypical clinical presentation, UPE is often misdiagnosed, resulting in an independent increased risk of mortality due to the delay in treatment.

Our patient presented with typical clinical features of heart failure and pulmonary oedema. Initial findings on chest X-ray showed a radiological pattern in keeping with bilateral opacification, which was more prominent in the right middle lung zone. Pneumonia is one of the differentials for these X-ray findings. However, as demonstrated in other articles, the unilateral radiological findings could have been secondary to the regurgitant MV jet or the anatomical drainage and position of the pulmonary veins.[1]

Other reports frequently documented leucocytosis as being a common finding in patients with UPE secondary to severe MR.[1,2] This is echoed in our case as our patient had a leucocytosis in the absence of fever and with negative blood cultures. In fact, there was no significant clinical improvement with antibiotic therapy.

An observational study by Attias et al, highlights the importance of taking into account the clinical course along with the patient's physical condition when making the correct diagnosis.[1] A comprehensive assessment including adequate history taking, clinical examination, appropriate use of laboratory investigations and conventional radiological imaging is key to diagnosing UPE. Storto et al suggest that CT imaging is not necessary to diagnose UPE but is useful to recognise the radiological pattern of increased hydrostatic pulmonary congestion.[3]

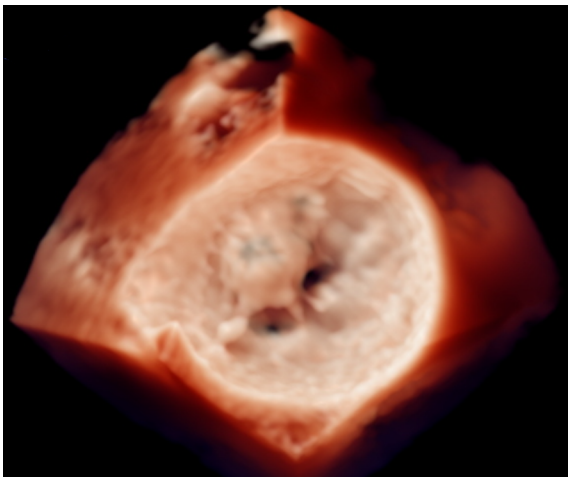

Figure 6 *A 3-dimensional reconstruction of the TOE images showing papillary chordal rupture as seen from the left atrium*

Given the difficulty in detecting MR on clinical examination, especially in patients presenting with acute SOB, bedside transthoracic echocardiographic examination remains an important tool in the diagnosis of UPE. In this case, an echocardiogram showed thickening of the anterior leaflet at the site of chordal rupture. TOE showed a flail and prolapsing A2 anterior leaflet of the mitral valve with a posteriorly-directed MR jet. The direction of the regurgitating jet is positioned in such a way so as to cause an asymmetric systolic flow reversal, whereby the mean pulmonary capillary pressures are higher in the right middle lung lobe leading to UPE.

As a consequence, the patient was investigated for an organic cause of MR, possibly resulting from rheumatic fever or infective endocarditis. With regards to the latter, no vegetations were visible on the cardiac valves and blood cultures were negative. Ischemic MR was excluded since troponin levels, ECG findings and relatively normal left ventricular dimensions on echocardiography were not consistent with dilated cardiomyopathy or with coronary artery disease. Moreover, a 3-dimensional reconstruction of the TOE images from the left atrium showed a papillary chordal rupture (Figure 6). Having previously been diagnosed with undifferentiated chronic mixed connective tissue disease and Raynaud's disease, the cause in our case points towards organic rather than functional MR.

CONCLUSION

UPE is an uncommon condition which typically manifests as an opacity in the right lung caused by severe mitral valve regurgitation. The direction of the regurgitating jet is positioned in such a way so as to cause an asymmetric systolic flow reversal, with a predilection to the right pulmonary veins. An abnormal increase in hydrostatic pressure in the pulmonary circulation causes fluid to accumulate in the extravascular compartment of the lung parenchyma resulting in pulmonary congestion and oedema.

An important take home message from this case is that UPE should be considered to be one of the differentials when radiological imaging is consistent with unilateral opacification. Maintaining a high index of suspicion is crucial in making the correct diagnosis since early detection and treatment of UPE significantly decreases the risk of mortality with good patient outcome.

ABBREVIATIONS

UPE	Unilateral pulmonary oedema
LRTI	Lower respiratory tract infection
SOB	Shortness of breath
PND	Paroxysmal nocturnal dyspnoea
WCC	White cell count
CRP	C-Reactive protein
CTPA	computerized tomography pulmonary angiogram
MR	Mitral regurgitation
TOE	Transoesophageal echocardiogram
MV	Mitral valve

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