Case Report

Unusual Pattern of Pulmonary Edema in a Case with Partial Anomalous Pulmonary Venous Drainage

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A B S T R A C T

Introdcution: Cardiogenic pulmonary edema manifests radiologically as bilateral symmetric air space opacification with central perihilar distribution. Asymmetric pulmonary edema is a relatively uncommon finding at imaging and is often misdiagnosed. There are a number of causes of unilateral or asymmetric pulmonary edema including postural and perfusion related factors.

Case report: We report an unusual pattern of pulmonary edema in a 47-year-old male who presented with shortness of breath to our emergency department. A chest radiograph and subsequently high-resolution computed tomography of chest was performed for evaluation. Both lungs showed diffuse ground glass opacities with patchy areas of consolidations along broncho-vascular bundles. Strangely the upper lobe of left lung was spared with normal appearing lung parenchyma. On further viewing an interesting observation was made. The left superior pulmonary vein was noted draining through a vertical vein into the left brachiocephalic vein thereby suggesting partial anomalous pulmonary venous drainage.

Conclusion: Awareness of the various causes of unilateral / asymmetrical pulmonary edema with unusual imaging patterns aids the diagnosis and avoids misinterpretation of underlying pathology.

Key-words: Pulmonary Edema, Partial Anomalous Pulmonary Venous Drainage, High Resolution Computed Tomography, Ground Glass Opacities, Left Upper Lobe, Asymmetric.

Key Messages: Partial anomalous pulmonary venous drainage should be considered as a cause of asymmetrical pulmonary edema while interpreting the imaging findings

INTRODUCTION

Pulmonary edema is defined as abnormal accumulation of fluid in the extravascular compartments of the lung. The relative amounts of intravascular and extravascular fluid in the lung are mostly controlled by the permeability of the capillary membrane as well as the oncotic pressure. Pulmonary edema can be divided into four main categories on the basis of pathophysiology: (a) increased hydrostatic pressure edema, (b) permeability edema with diffuse alveolar damage (DAD), (c) permeability edema without DAD, and (d) mixed edema due to simultaneous increased hydrostatic pressure and permeability changes.[1] Radiologically the patterns of pulmonary edema presentation differ in each patient of cardiac, renal, and capillary permeability edema related conditions. Unilateral or asymmetric pulmonary edema (UPE) is uncommon, accounting for 2% of cardiogenic pulmonary edema cases and usually involves the upper lobe of the right lung.[2] Various causes of unilateral / asymmetrical pulmonary edema are encountered in our day today practice and these are usually position or perfusion related. We present an unusual case of asymmetrical cardiogenic pulmonary edema with sparing of left upper lobe due to partial anomalous pulmonary venous drainage.

CASE REPORT

A 47-years-male presented to the emergency department with shortness of breath. He was a known diabetic and hypertensive with coronary artery disease and history of inferior wall myocardial infarction in the past.

On general physical examination, his pulse rate was 96/minute and blood pressure were 100/70mmHg. Cardiovascular system examination was unremarkable. On auscultation of lungs bilateral basal crepitations were present. Blood investigations including hemogram, renal function tests and serum electrolytes were unremarkable. Serum lactate dehydrogenase (LDH) (408 U/L) and serum creatine phosphokinase (222 U/L) levels were elevated. Electrocardiogram (ECG) showed features of left ventricular hypertrophy and 2D Echocardiography revealed severe

mitral regurgitation with mild diastolic dysfunction. A chest radiograph was performed that showed haziness in bilateral mid and lower lung zones. For further evaluation a non-contrast CT chest was performed. Scanogram of the chest showed asymmetric air space opacities in the right lung in perihilar and central distribution with normal appearing left lung (Figure 1). HRCT thorax showed diffuse ground glass opacities with patchy areas of consolidations in peri broncho-vascular distribution in both lungs with strikingly spared left upper lobe parenchyma (Figure 3). On careful observation note was made of anomalous drainage of the left upper lobe pulmonary vein through the vertical vein into the left brachiocephalic vein. This represents a partial anomalous left upper lobe pulmonary venous connection (Figure 2). Rest of the pulmonary veins and cardiovascular structures were normal in appearance, connection. Patient was managed for congestive cardiac failure. A follow up radiograph showed clearing of the pulmonary edema.

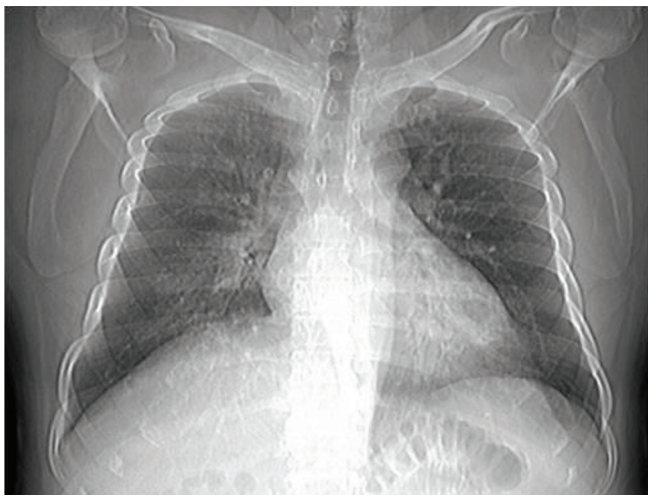

Figure-1: CT Scanogram of chest showing asymmetric air space opacities in the right lung in perihilar and central distribution with normal appearing left lung.

DISCUSSION

Pulmonary edema is defined as an abnormal accumulation of fluid in the extravascular compartments of the lung. Increased hydrostatic pressure leads to two pathophysiological and radiological phases in the development of pressure edema, a) interstitial edema and b) alveolar edema. These phases are virtually identical for left-sided heart failure and fluid overload, the two most frequently observed causes of pressure edema in intensive care and emergency patients. Interstitial edema occurs with an increase of 15–25 mmHg in mean pulmonary capillary wedge pressure and results in the early loss of definition of subsegmental and segmental vessels, mild enlargement of the peribronchovascular spaces, the appearance of Kerley lines, and subpleural effusions. If the quantity of extravascular fluid continues to increase, the edema will migrate centrally with progressive blurring of vessels, first at the lobar level and later at the level of the hilum. At this point, lung radiolucency decreases markedly, making identification of small peripheral vessels difficult. Peribronchial cuffing becomes apparent, particularly in the perihilar areas. With increases in mean pulmonary capillary wedge pressure greater than 25 mm Hg, fluid drainage from the extravascular compartment is at maximum capacity and the second phase (alveolar flooding) commences, leading to a sudden extension of edema into the alveolar spaces, thus resulting in acinar opacities that coalesce into frank consolidations.[1,2]

Bat wing pattern of edema refers to bilateral, central,

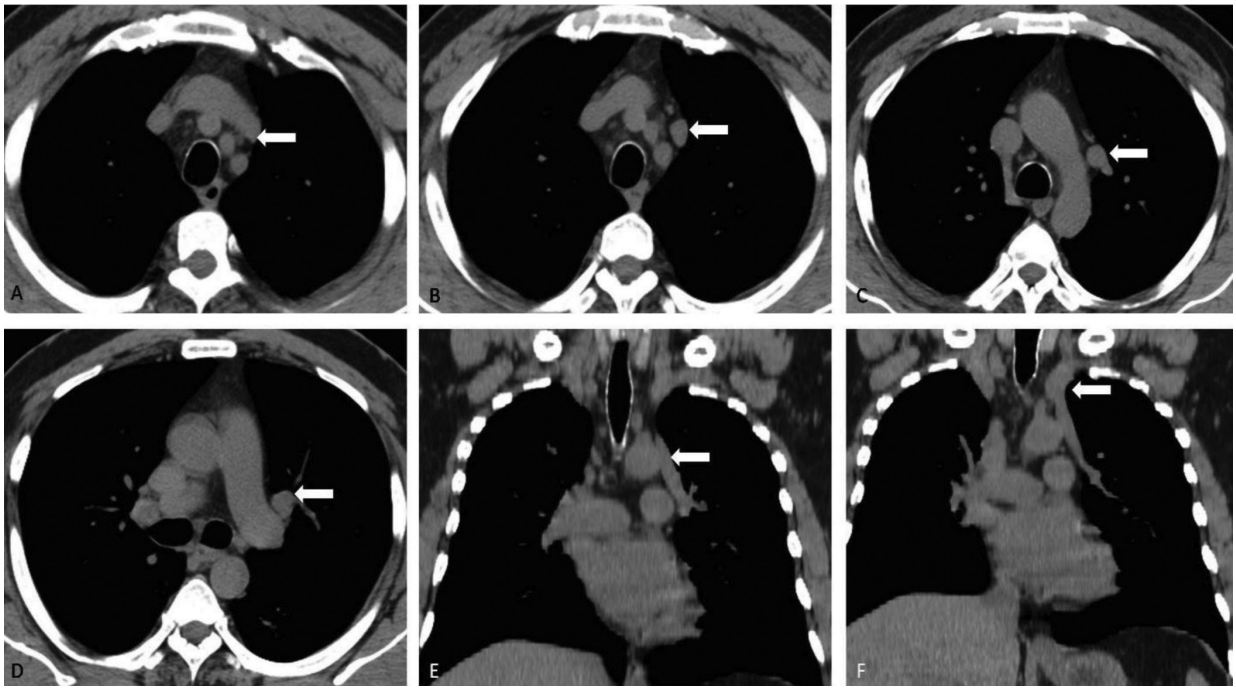

Figure-2: Serial axial and coronal reformatted images of CT chest in mediastinal window revealed drainage of left upper lobe pulmonary vein into the left brachiocephalic vein. This represents a partial anomalous left upper lobe pulmonary venous connection (arrows).

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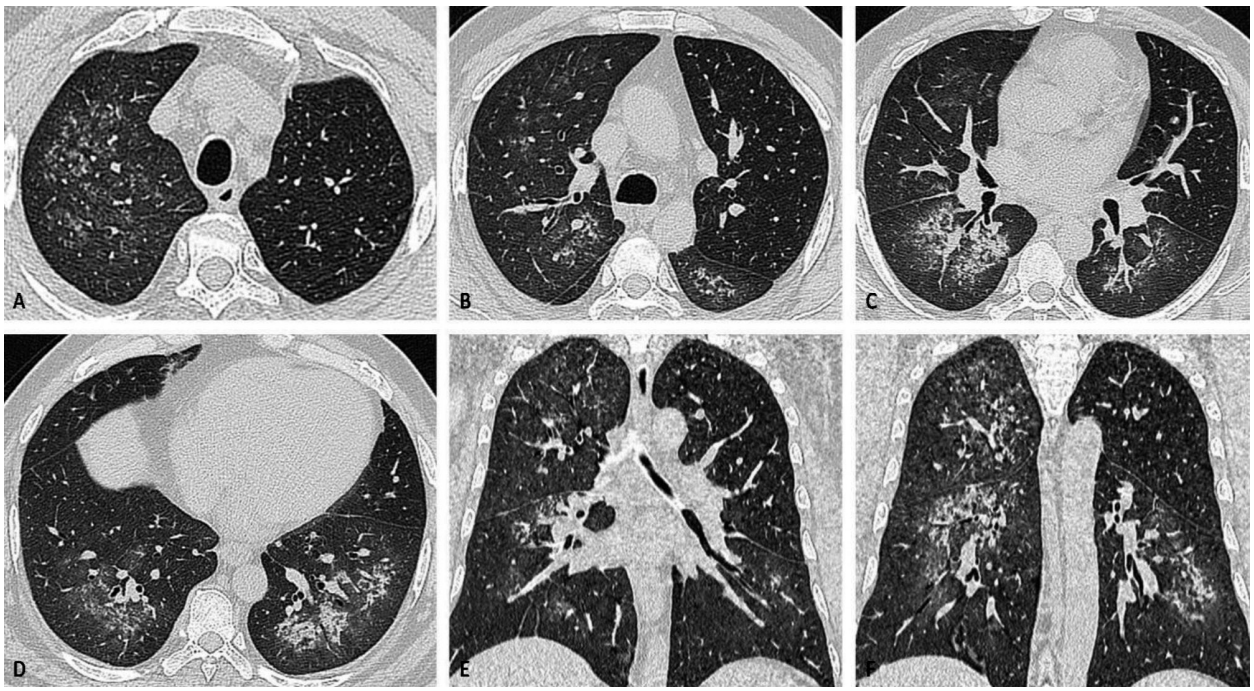

Figure-3: Serial axial and coronal reformatted images of HRCT chest in lung window showed diffuse bilateral ground glass opacities with patchy areas of consolidations along broncho-vascular bundles. Note the relative sparing of left upper lobe parenchyma.

nongravitational distribution of alveolar edema. It generally occurs in the setting of rapidly developing severe cardiac or renal failure.[1,2] The most frequent cause of asymmetric distribution of pressure edema is morphologic changes in the lung parenchyma in chronic obstructive pulmonary disease. In cardiac failure, extensive lung emphysema of the apices (seen in heavy smokers) or marked destruction and fibrosis of the upper and middle portions of the lungs (seen in end-stage tuberculosis, sarcoidosis, or asbestosis) will result in pulmonary edema that predominates in the regions that are less affected by these disease processes. Hemodynamic factors can also cause asymmetric distribution of pulmonary edema. Edema associated with mitral regurgitation has been shown to predominate in the right upper lobe as a result of flow impairment caused by the reflux stream that is directed toward the right upper pulmonary vein. Such asymmetric distribution occurs in 9% of adults and 22% of children with grade III or IV mitral regurgitation. Finally, the position of the patient also influences intra- and extravascular fluid distribution. In supine patients, axial CT usually demonstrates an anteroposterior gradient, whereas more asymmetric distribution of edema secondary to prolonged surgery or immobilization is frequently observed in the lung fields in recumbent patients. This distribution is typically seen in congestive heart failure but is also observed in overhydration.[1,2]

Various pathologies present with different patterns of pulmonary edema, few of them are discussed here. 1) Post obstructive pulmonary edema typically manifests radiologically as septal lines, peribronchial cuffing, and, in more severe cases, central alveolar edema. 2) Pulmonary edema with chronic pulmonary embolism manifests as sharply demarcated areas of increased ground-glass attenuation. 3) Pulmonary edema with veno-occlusive disease manifests as large pulmonary arteries, diffuse interstitial edema with numerous Kerley lines, peribronchial cuffing, and a dilated right ventricle. Stage 1 near drowning pulmonary edema manifests as Kerley lines, peribronchial cuffing, and patchy, perihilar alveolar areas of airspace consolidation; stage 2 and 3 lesions are radiologically nonspecific. 4) Pulmonary edema following administration of cytokines demonstrates bilateral, symmetric interstitial edema with thickened septal lines. 5) High-altitude pulmonary edema usually manifests as central interstitial edema associated with peribronchial cuffing, ill-defined vessels, and patchy airspace consolidation. 6) Neurogenic pulmonary edema manifests as bilateral, rather homogeneous airspace consolidations that predominate at the apices in about 50% of cases. 7) Reperfusion pulmonary edema usually demonstrates heterogeneous airspace consolidations that predominate in the areas distal to the recanalized vessels. 8) Post-reduction pulmonary edema manifests as mild airspace consolidation involving the ipsilateral lung, and 9) pulmonary edema due to air embolism initially demonstrates interstitial edema followed by bilateral, peripheral alveolar areas of increased opacity that predominate at the lung bases.[1,2,5]

In our case, cardiogenic pulmonary edema appeared asymmetric with sparing of the left upper lobe due to partial anomalous pulmonary venous drainage of the lung parenchyma into systemic central vein. In cases with partial anomalous pulmonary venous drainage (PAPVD), the pressure changes in the left atrium during left heart failure do not result in venous congestion of the lung parenchyma as the pulmonary vein drains into the systemic veins and not into the left atrium.

Congenital abnormalities of the major mediastinal venous

structures occur in less than 2% of population and are often detected at imaging. Partial anomalous pulmonary venous drainage is a rare abnormality with failure of connection between the initial draining system of the lungs and the common pulmonary vein. In this anomaly, blood from the anomalously drained portion of the lung is recirculated to the right side of the heart, creating a left-to right shunt. Right sided anomalous return is the most common form of anomalous connection, with left sided anomalous return uncommon.[3,4] Normal pulmonary venous anatomy consists of four separate pulmonary veins: right and left superior and inferior veins, which drain individually into the left atrium. The right superior pulmonary vein (RSPV) drains the right upper and middle lobes of the lung with the right lower lobe drained by the right inferior pulmonary vein (RIPV). Then left superior pulmonary vein (LSPV) drains the lingual and left upper lobe, whilst the left lower lobe is drained by the left inferior pulmonary vein (LIPV). This anatomical arrangement is found in 60–70% of the population.

Stephen Lyen et al. (2017) reported that left upper lobe PAPVD will spare vascular congestion of the left upper lobe in left heart failure, as the left upper lobe pulmonary vein does not drain into the left atrium which is similar to present our study. Similarly, in right heart failure the same anomaly causes pulmonary oedema isolated to the left upper lobe.[3,4]

PAPVD is an infrequent cause of asymmetric pulmonary edema and cross-sectional imaging is helpful in evaluation of such patients.

CONCLUSION

In day-to-day practice, radiologists encounter various usual and unusual patterns of pulmonary edema. Awareness of partial anomalous pulmonary venous drainage as a cause of asymmetrical pulmonary edema reduces the chances of misdiagnosis and aids in appropriate case management

ACKNOWLEDGMENTS

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ABBREVATIONS

PAPVD - Partial anomalous pulmonary venous drainage
HRCT - High resolution computed tomography
DAD - Diffuse alveolar damage
ECG - Electrocardiogram
LDH - Lactate dehydrogenase
HIV ELISA - Human immunodeficiency virus Enzyme linked immunosorbent assay

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