

The Ground Glass of the Pulmonary-Kidney Syndrome Case Report

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Abstract

The pulmonary-Kidney syndrome encompasses genetic and autoimmune diseases that involve tissue structures of the same origin. Radiological findings are usually found with alveolar opacities and ground glass. We present a related clinical case and the diagnostic approach of the most frequent differential diagnoses of these findings.

Keywords: Microscopic Polyangiitis; Ground Glass; Pulmonary Kidney Syndrome

Abbreviations: SLE: Systemic Lupus Erythematosus; GBM: Glomerular Basement Membrane; ANCA: Antineutrophil Cytoplasmic Antibodies; NSIP: Non-specific Interstitial Pneumonia.

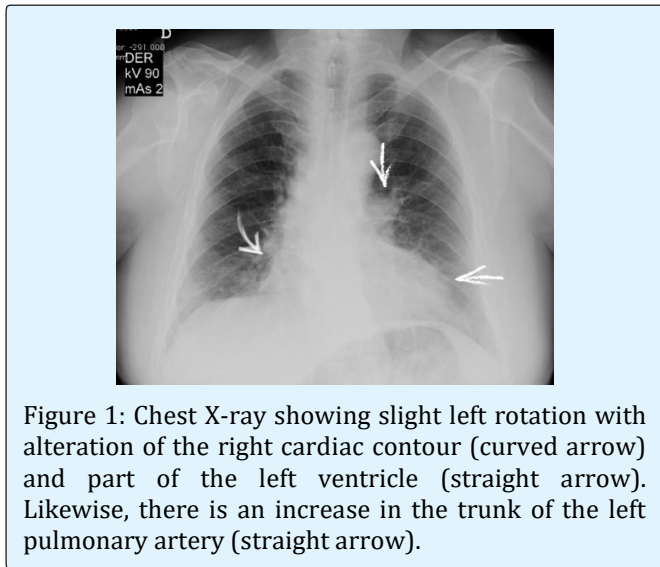
Introduction

The diseases that affect the lung and the kidney at the same time can be of genetic or autoimmune origin. The pulmonary-kidney syndrome was initially associated with the pathologist Ernest Goodpasture in 1919, due to his findings in autopsies in the epidemic influenza [1]. Taking into account that the etiological diagnosis is vasculitis, an annual incidence of 20 / million inhabitants is estimated [2]. Within the autoimmune spectrum, it is important to know epidemiologically which are the most frequent and which imaging spectrum they develop, in order to have an etiological approach from the diagnostic images of the thorax [3,4]. We presented a case of a patient with pulmonary-kidney syndrome due to vasculitis and the imaging approach of this diagnostic challenge.

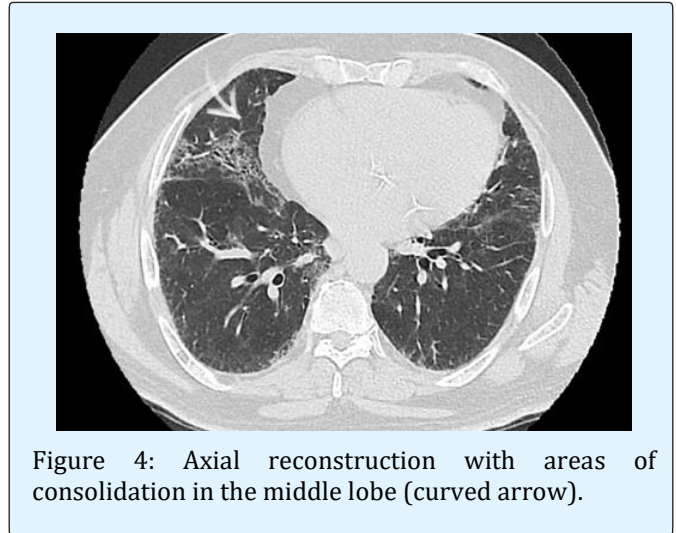
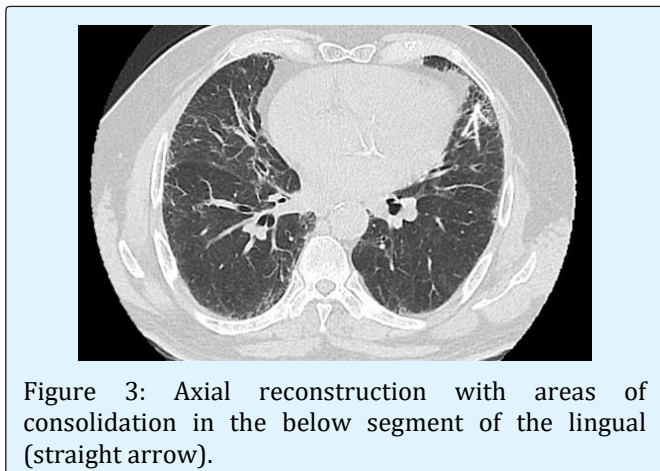
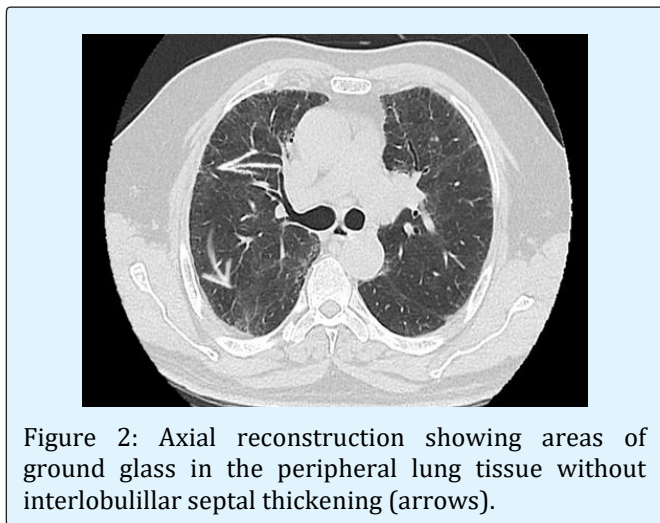
Clinical Case

A 66-year-old patient diagnosed with controlled hypertension without renal disease who presented a 5-month course consisting of impairment of his functional class from I / V to III / V with dyspnea as the main symptom. Dyspnea is exacerbated during the last 15 days with dry cough without expectoration, so consult the emergency department. At admission, the patient had a laboratory test of interest: hemoglobin without alterations with leukocytosis without neutrophilia, the uroanalysis presented incipient proteinuria. Creatinine values of 1.58 without alterations in nitrogen. The patient was taken to the radiology department where a chest x-ray and a high-resolution tomography (Figures 1-4). The patient presented clinical deterioration with sudden renal alteration despite medical management. Within a few days, it was determined that the patient had positive anti myeloperoxidase by indirect immunofluorescence compatible with microscopic polyangiitis. He is given medical treatment with clinical improvement and early hospital discharge.

Figure 1: Chest X ray AP projection



Figures 2-4: High-resolution tomography in axial reconstruction.



Discussion

The kidneys and lungs share special diseases due to their high perfusion and similarity in the structure of the basement membrane and microtubules [3]. The distribution of kidney or lung disease depends on each etiology. It is very important to know the clinical progression, the distribution of the age group in terms of the social and demographic variables, the comorbidities that worsen the clinical picture and the chronological imaging findings (Table 1). The radiological approach in radiography and tomography is very difficult because most of the findings focus on alveolar and ground glass opacities as in the case of our patient. Therefore, it is always essential to know as a radiologist the clinical history and the reduced spectrum of autoimmune diseases that can develop this lung-kidney syndrome. In the case of the patient, the nonspecific findings were tomographically reported and on the other hand his renal injury progressed without apparent cause. Epidemiologically, the most frequent etiology in these cases of kidney lung syndrome is microscopic polyangiitis, as it was in our patient [4]. However, the radiologist must be more analytical and evaluate the associated findings, as in the case of Goodpasture syndrome, which is the main differential diagnosis [5]. However, it occurs more in young people and smokers [3,4].

Granulomatosis with polyangiitis, presents with additional deterioration of the upper airway (90%), subglottic stenosis and nodules or lung masses of peripheral distribution. Eosinophilic Granulomatosis with Polyangiitis presents with peripheral neuropathy, in young patients, and with asthma up to 90% [6]. Systemic lupus erythematosus (SLE) is more common in women and presents pleuritis and pulmonary embolism up to 50%

[7], unlike IgG4-Related Sclerosing Disease, where pancreatic abnormality is characteristic. Systemic sclerosis presents with dermatological alterations [8], the Erdheim-Chester Disease with metaphyseal cortical alterations of the legs, while the sarcoidosis debuts with nephrocalcinosis and a characteristic pattern in the tomography without ground glass generally (Table 1).

The clinical relevance of ground glass in this type of patient speaks of the progression of the disease with dual

pulmonary-kidney involvement. It is important to always look for the clinical context of the patient and always ask if there is renal involvement at the time of assessing the finding of ground glass. Frequently the clinician looks for the radiologist to ask about the relevance of this finding in the tomography. It is important that the radiologist evaluate the post-test probability of the findings of the ground-glass opacities, which increases in the context of a patient with acute renal impairment directing a diagnostic approach to pulmonary-kidney syndrome.

Syndrome	Clinical progression	Social and demographic distribution / Comorbidities	Imaging findings	Physiopathology
Goodpasture	Rapidly progressive glomerulonephritis 40-60%. Alveolar hemorrhage	Young / Smoker	Rx: Solitary, multifocal or diffuse alveolar opacities. CT: ground glass, thickening of interlobular septa, consolidation	Glomerular basement membrane antibodies (GBM)
Granulomatosis with polyangiitis (before Wegener)	Upper airway deterioration 90%. Glomerulonephritis 85%.	40-65 years old / Treatment with corticosteroids, proteinuria > 1g / day. Previous kidney injury.	Nodules or lung masses, generally peripheral, ground glass, nodules cavitated in 50%, subglottic stenosis, and alteration of the posterior membrane of the trachea.	Antineutrophil cytoplasmic antibodies (c-ANCA)
Eosinophilic Granulomatosis with Polyangiitis (before Churg Strauss)	Peripheral neuropathy 75%, glomerulonephritis 25%.	3rd decade of life 90% / Asthma 90%. Eosinophilia	Ground glass, consolidative airspace opacities, pulmonary nodules, interlobular septal thickening.	Antineutrophil cytoplasmic antibodies (c-ANCA)
Systemic lupus erythematosus (SLE)	Arthritis, rash, glomerulonephritis (50%), pleuritis and pericarditis.	Women / Immunosuppression	Pleuritis 50%. Pulmonary embolism, opportunistic pneumonia.	ANA or anti-dsDNA
Systemic sclerosis	50% have renal impairment but renal failure is uncommon	Scleroderma, Thickening of the skin Interstitial lung disease 75%. Pulmonary hypertension 50%.	Non-specific interstitial pneumonia (NINE). Peripheral and peribronchovascular fibrosis. Ground glass, traction bronchiectasias.	Deposits of collagen in soft tissues and vasculature [9].
IgG4-Related Sclerosing Disease	Autoimmune pancreatitis (>patients), renal injury 35%, biliary disorder 77%, lymph nodes 33%, retroperitoneum and mediastinum.	67 years old (25-93). Inflammatory bowel disease.	Pancreatic thickening and hypoenhancement, pulmonary alterations 13%: peribronchial inflammation, discrete nodules, reticulation or septal thickening, or consolidation. Renal involvement is common and involves the cortex. The most common appearance is discrete nodules or wedge-shaped lesions that are T2 hypointense and	Infiltration of plasma cells and positive T lymphocytes for IgG4. IgG4 levels elevated 86%.

			hypoenhancing with restricted diffusion	
Erdheim-Chester Disease	metadiaphyseal cortical thickening in the lower extremity long bones 96%. Renal involvement: perirenal soft tissue rind altered.	non-Langerhans cell histiocytosis.	septal thickening and ground-glass opacities 50%. Pleural and pericardial effusions >50%.	BRAF proto-oncogene
Sarcoidosis	Nephrocalcinosis > 50%.	Alteration of the skin, central nervous system, eyes (uveitis), as well as liver and spleen.	Hilar lymph nodes. Eggshell calcifications. Upper-lung predominant perilymphatic nodules. peri-bronchovascular distribution, with fibrotic bands and traction bronchiectasis en enfermedad avanzada	autoimmune disorder of granulomatous inflammation
Microscopic polyangiitis [4]	Necrotizing glomerulonephritis 80%. Diffuse pulmonary hemorrhage 50%, with dyspnea and hemoptysis.	Alveolar hemorrhage	Diffuse bilateral air opacities. Ground glass.	Systemic necrotizing vasculitis not granulomatous

Table 1: Distribution of autoimmune diseases that present lung-kidney disorders [3].

Conclusion

Vasculitis of small vessels is the main cause of the alteration of the kidney and the lung in this type of patients, where at least twenty different varieties are described. The most important manifestation is the renal alteration that may be incipient, associated with pulmonary manifestations, where up to 50% of the cases do not present alterations in the chest X-ray. When radiographic alterations occur, images of alveolar occupation without alteration of diaphragmatic cost angles are evident [2]. In spite of this, in the high-resolution tomography, the pattern in ground glass without interlobular thickening, which can guide the diagnosis with diffuse alveolar hemorrhage, is acutely evidenced that associated with the renal alteration is highly suggestive of a pulmonary-kidney syndrome caused by Vasculitis. Microscopic polyangiitis presents with segmental focal glomerulonephritis in addition to the imaging findings already described [10]. As a final recommendation it can be described that whenever we are confronted with this non-specific finding in tomography with important clinical deterioration of the patient, we must ask ourselves and follow up the renal function to be able to evaluate if it is a pulmonary kidney syndrome in the context of these vasculitis.

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