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Pneumomediastinum in a Patient with Systemic Lupus Erythematosus with Pulmonary Renal Syndrome: Pneumomediastinum in a Patient with Systemic Lupus Erythematosus with Anca-P+ Pulmonary Syndrome

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INTRODUCTION

Lung-kidney syndrome is a clinical syndrome involving the presence of diffuse alveolar hemorrhage and rapidly progressive glomerulonephritis, which progresses to acute respiratory failure and end-stage renal disease. It is usually associated with rheumatologic diseases, which can be divided into ANCA-associated vasculitis (AAV) and autoimmune complex-mediated vasculitis.

The diseases frequently implicated are those associated with anti-neutrophil cytoplasmic antibodies (ANCAs) and anti-glomerular basement membrane disease (anti-GBM). The incidence of lung-kidney syndrome is estimated to be between 2.4 and 2.7 cases per million, frequently occurring in females between the ages of 40 and 60 years. Positive ANCA antibodies are observed in 30% to 50%, renal diseases are frequently associated with proteinase 3 antibodies (Anti-PR3) which are prevalent in America and Europe. Pulmonary hemorrhage occurs in the same percentage and can be fatal. Systemic Lupus Erythematosus (SLE) is responsible for 80%, is caused by antibodies directed against double-stranded DNA (anti-dsDNA), which are deposited in immunocomplexes in the glomerulus, and activate the proinflammatory response. Pleural and pulmonary disease occurs in 70% of cases, however, lung-kidney syndrome occurs in 4% of cases with mortality rates of 50%. [43]

In the kidney there is the formation and deposition of immunocomplexes in the glomerulus which results in the activation of a proinflammatory cascade[8.] In the lung, there is vasculitis of small vessels and necrosis, at alveolar and interstitial level, there is necrotizing pulmonary

capillaritis, with neutrophilic infiltration and edema, with fibrin thrombi. Weakening the alveolar integrity, allowing the entrance of red blood cells that cross pulmonary capillary basement membranes, as a consequence the entrance to the interstitial space and flooding the alveoli. Pneumomediastinum is a condition in which air is present in the mediastinum secondary to physical trauma to the chest wall. When it is spontaneous it is usually self-limited. In the lung-kidney syndrome it may be secondary to rupture of alveoli adjacent to the mediastinum. In pneumomediastinum, the most associated causes are related to diseases such as asthma, smoking, drug use and triggering factors such as Valsalva maneuvers. Within the rheumatologic field, it is an entity described in connective tissue diseases, predominantly in polymyositis and dermatomyositis. However, the evidence related to systemic lupus erythematosus is insufficient. There are reported cases associated with vasculitis, however, its presentation is not defined as a primary agent or secondary cause.

CASE REPORT

We present the case of a 27-year-old female patient, restaurant employee. Hemotype B positive. Mother died secondary to a hemorrhagic cerebrovascular event at 41 years of age, with a history of systemic lupus erythematosus. Grandmother died at age 52, with a history of systemic lupus erythematosus. Gynecobstetric history, she had a cesarean section 4 years ago secondary to preeclampsia, she denies history of abortions. She denies toxic manic habits. She denies chronic degenerative diseases prior to current illness and history of blood transfusions. She starts current condition with diffuse alopecia and madarosis, as well as pallor of the integuments for the previous 3 months. She denies malar erythema, as well as annular or papulosquamous lesions. No fever, no weight loss, no arthralgias, no gingivorrhage, no ecchymosis.

She started on 04/06/24 with small effort dyspnea which gradually progressed to medium effort, adding tachypnea and tachycardia. Initial medical attention was sought on 12/04/24. Initial laboratory studies were performed, of which the following stand out: Anemia of chronic disease pattern with iron deficiency, proteinuria with active sediment, renal failure with a glomerular filtration rate (GFR) of 16 ml/kg/1.73m3 , moderate hyperkalemia and metabolic acidosis with elevated gap anion. Subsequently, antibody studies were performed, of which ANCA positive (AC-ANTI P-ANCA (1:10) AC Anti C-ANCA negative. Anti nuclear positive AC-5 (1:5120), negative cytoplasmic pattern, presence of C3 (35), C4 (3.8) complement. Cardiolipin IgM antibodies 2.54, Anti-Cardiolipin (IgG) 25.20. Given the clinical conditions of acute respiratory failure, she was referred to the Hospital General de Zona #2, she arrived at the emergency area on 04/13/24 where 2 erythrocyte concentrates were transfused as part of her initial management due to hemoglobin of 4.1 g/dL. Conventional oxygen support was started with a high flow device with reservoir mask at 10 liters per minute, with a FiO2 calculated at 51%. Physical examination revealed sinus tachycardia with a heart rate of 113 beats per minute, pulmonary fields with thick rales in both bases with tachypnea in the range of 23-28 breaths per minute, globose abdomen at the expense of adipose panniculus, and asignologic. Diuresis on admission without quantification. Initial imaging study with chest X-ray with presence of diffuse alveolar infiltrate, predominantly in bases and peri-hilar level. A diagnosis of systemic lupus erythematosus SLEDAI 24 points and lung-kidney syndrome was integrated. That same day he was admitted to the intensive care unit. Subsequently, acute renal injury was documented with a glomerular filtration rate of 6ml/min/1.73m2 (Grade V) and the presence of severe refractory hyperkalemia, for which anticalemic measures were initiated.

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Methylprednisolone pulses were started at 1g per day for 5 days, as well as cyclophosphamide doses calculated at 25mg/kg. Replacement therapy was started with hemodialysis preserving diuresis of 0.9ml/kg/hour. On 04/14/24, a tomographic study was performed showing the presence of interstitial alveolar pulmonary infiltrates randomly distributed with diffuse bilateral pneumonic foci with centrolobulillar septal thickening giving a cobblestone appearance compatible with diffuse alveolar hemorrhage and the presence of pneumomediastinum at the right paratracheal level, with lower mediastinal predominance. His clinical evolution showed cardiorespiratory improvement. On 04/26/24 during her stay she was managed with empirical antibiotic therapy with vancomycin, meropenem and fluconazole adjusted to the glomerular filtration rate, secondary to the onset of fever. However, she presented new respiratory deterioration which again required supplemental oxygen support with simple mask at 6 liters per minute. New laboratory studies showed leukocytosis of 12,000 at the expense of 96% neutrophils, persistence of normochromic hypochromic anemia, hemoglobin of 4.6. Urea of 224mg/dl, creatinine of 5.2 mg/dl. After the hemo transfusion, he presented progression of acute respiratory failure of refractory character, requiring advanced management of the via area, that same day he presented massive hemoptysis that caused cardiac arrest, cardiopulmonary resuscitation maneuvers were initiated without obtaining return of the pulse and death was declared on 04/27/24.

PARACLINICS

EXAMINATIONS 12/04/24	**RESULTS**	**REFERENCE VALUES**
Leukocytes	14.8	(4.0-10.5)
Neutrophils	11.0	(1.5-7.0)
Hemoglobin	4.1	(11-18)
Hematocrit (%)	13.3	(34-50)
Mean corpuscular volume	90.50	(80-110)
Platelets	318.0000	(150-400)
Lymphocytes	17.8%	(2.0-10)
Monocytes	2.3%	(0.0-5)
Eosinophils	0.7%	50-80
Prothrombin time	15.1	(11.7-15.3)
TP Witness	13.3	(10-15)
INR	1.14%	(0.8-7.2)
RENAL FUNCTION AND ELECTROLYTES		
Glucose	106	(136-145)
Urea	132	(70-105)
Bun	61	(20-43)
Uric Acid	9.7	(2.4-7.0)
Sodium	131	(135-145)
Potassium	6.44	(3.60- 5.00)
Chlorine	104	98-107
Phosphorus	6.8	(3.60-4.5)
Magnesium	2.07	(1.60-2-45)
Calcium	7.7	(8.6-10.0)

GENERAL URINE TEST	**RESULTS**	**NORMAL VALUES**
DENSITY	1.020	(1.010-1.030)

PH	5	(5.50-7.00)
NITRITES	NEGATIVES	NEGATIVES
PROTEINS	500	NEGATIVES
HEMOGLOBIN	250	NEGATIVE
URINARY SEDIMENT		
LEUCOCYTES	1-3 PER FIELD	NEGATIVE
ERITROCYTES	UNCOUNTED PER FIELD	NEGATIVES
EPITHELIAL CELLS	SCARCE PER FIELD	NEGATIVES.
BACTERIA	REGULARS PER FIELD	NEGATIVE
HYALINE CYLINDERS	0-2	NEGATIVE.

EXAMINATIONS	**RESULTS**	**REFERENCE VALUES**
Leukocytes	7.5	(4.0-10.5)
Neutrophils	7.25	(1.5-7.0)
Hemoglobin	6.0	(11-18)
Hematocrit (%)	17.9	(34-50)
Mean corpuscular volume	91.80	(80-110)
Platelets	135.000	(150-400)
Lymphocytes		(2.0-10)
Monocytes	1.9	(0.0-5)
Eosinophils	0.5	50-80
C-REACTIVE PROTEIN	5.10	<0.5
RENAL FUNCTION AND ELECTROLYTES		
Glucose	102	(136-145)
Urea	218	(70-105)
Bun	102	(20-43)
Uric Acid	9	(2.4-7.0)
Sodium	131	(135-145)
Potassium	4.57	(3.60- 5.00)
Chlorine	96	98-107
Phosphorus	7-1	(3.60-4.5)
Magnesium	1.99	(1.60-2-45)
Calcium	8	(8.6-10.0)

DISCUSSION

Patients with lung-kidney syndrome may have a fulminant course. If a patient deteriorates after steroid and antineoplastic therapy, the possibility of sepsis should be considered given the level of immunosuppression. In SLE, disease recurrence after treatment is common.

In the management of the patient with acute renal injury, due to rapidly progressive glomerulonephritis, administration of corticosteroids such as methylprednisolone in pulses of 500-1000mg daily, for induction of remission. Continuing with oral corticosteroid with prednisolone at doses of 0.6-0.8mg/kg/day. However, according to the guidelines by the KDIGO organization, the use of immunosuppressants in conditions such as ANCA-positive glomerulonephritis, in which the deterioration of renal function is very rapid and is associated with severe complications including alveolar hemorrhage. It is managed with cyclophosphamide with renal adjustment at doses of 250 - 750mg/m2. Showing reduced relapse rate. However, the patient has a torpid evolution, probably combined with the poor

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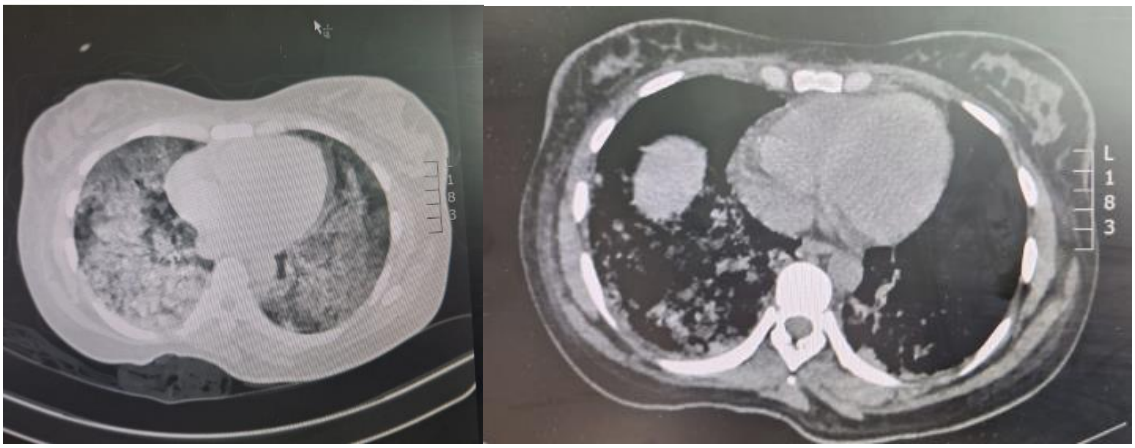
prognosis of the same, according to the clinical classification system to predict the prognosis in patients with RPGN of 5 points-Grade II. In the treatment of pneumomediastinum there is no established consensus, the great majority of the reported cases the treatment is conservative since it is a self-limited condition as a whole. In our presented case the definitive management was conducted to the resolution of the patient's condition.

TOMOGRAPHY



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