Granulomatosis with polyangiitis as a differential diagnosis in intensive care unit: a case report

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ARTÍCULO ORIGINAL

Abstract: *Granulomatosis with polyangiitis, a small vessels vasculitis, involves mostly upper respiratory tract, lungs and kidneys. It's a multisystemic pathology, with no sex predominance, affecting adults over 40 years old. Case report: K.S.D, 64 years, female, came to emergency room with dyspnea, cough and coryza, 15 days after a recurrent acute otitis media surgical treatment. A CT showed bilateral pleural effusion, consolidation and a lung mass, being admitted in Intensive Care Unit. Subsequently, developed mastoiditis and conjunctival hyperemia and used several antibiotic therapys. Patient was discharged 40 days later, with clinical improvement. One month later, was admitted again with pulmonary septic shock, in addition to sinusitis, polyarthralgia, purpura and acute renal injury, requiring hemodyalisis. Biomarkers were requested, such as c-ANCA and rheumatoid factor, with positive result and antinuclear factor, with negative result. A renal biopsy also has been solicitated, showing glomerulonephritis. The diagnosis was granulomatosis with polyangiitis, treated with methylprednisone pulse therapy and cyclophosphamide, progressing to clinical stability and receiving discharge with outpatient follow-up. Conclusion: Granulomatosis with polyangiitis is a rare condition, with that should be considered when patient presents the triad: upper and lower airways and glomerulonephritis. Early diagnosis and immunosuppressive therapy are essential to reduce its morbimortality.*

Keywords: *biomarkers, granulomatosis, polyangiitis*

INTRODUCTION

Granulomatosis with polyangiitis (GPA), is immunologically mediated, which is characterized by an inflammatory reaction pattern, such as necrosis, granulomatous inflammation and vasculitis1. It involves mostly upper and lower respiratory tract, eyes, skin and kidneys, with no sex predominance, affecting adults over 40 years old2.

GPA is included in antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis (AAV)1, which is a group of systemic inflammatory diseases with necrosis of small blood vessels that can attack multiple organ systems, and includes: GPA, microscopic polyangiitis (MPA) and eosinophilic granulomatosis with polyangiitis (EGPA, formerly Churg-Strauss Syndrome)3. Recently, it has been suggested contribute to better understanding this diseases physiopathology and diagnosis. The subtypes are myeloperoxidase (MPO-ANCA) and proteinase 3 (PR3-ANCA), and the PR3-ANCA appears to be most commonly associated with GPA3.

Upper airways involvement occurs in approximately 90% of patients, who develops ear, nose and throat manifestations such as otitis media, earache, mastoiditis, otorrhea, persistent rhinorrhea, blood nasal discharge, sinusitis, oral and nasal ulcers and retro-orbital masses4. In several cases, it could occur more severe sequels, such as permanent hearing impairment, bone and cartilage destruction and cranial nerve entrapment5. Lower airways involvement may present dyspnea, stridor, tracheal stenosis, pulmonary consolidation and pleural effusion4.

The chest radiograph findings are variable and includes nodules, fleeting pulmonary infiltrates, hilar adenopathy and even tumor-like masses[4,5].

The most common renal manifestation is glomerulonephritis, which may range from asymptomatic hematuria to rapidly progressive glomerulophritis[1,3]. Other manifestations includes cutaneous (purpura, nodules or focal necrosis and ulceration) and ophthalmic (conjunctivitis, episcleritis, uveitis, retinal vasculitis) involvement[6,7].

Diagnosis is based on clinical manifestations, a positive c-ANCA serology and histological evidence of necrotizing vasculitis, glomerulonephritis or granulomatous inflammation from an organ biopsy, such as skin, lung or kidney. Positive ANCA serology is not essential for the diagnosis of GPA if clinical and histological findings exclude vasculitis mimics or other types of systemic vasculitis, but the presence of this biomarker in GPA is a great aid to diagnosis[1].

Treatment of GPA involves immunosuppression, which can be managed with methotrexate or mycophenolate associated with glucocorticoid, when there are no organ threatening disease. In case of organ or life threatening disease, patient should be treated with cyclophosphamide or rituximab associated with glucocorticoid and, in severe manifestations, such as rapidly progressive glomerulonephritis or pulmonary haemorrhage, plasma exchange should be considered[8].

Difficulty in diagnosis often delays the initiation of treatment, and the disease occasionally progresses to the irreversible phase1. This case report discusses a patient who was managed in an intensive care unit and, due to early correct treatment, presents a good prognosis. Therefore, the aim of this study is draw attention to GPA as a differential diagnosis in an intensive care unit.

CASE REPORT

K.S.D, 64 years, female, with chronic obstructive pulmonary disease and hypothyroidism, former smoker, came to emergency room with dyspnea, asthenia, cough and coryza, 15 days after a recurrent acute otitis media surgical treatment. Computed tomography (CT) chest scans showed bilateral pleural effusion, consolidation and a lung mass to be investigated (Figure 1). Admitted in intensive care unit, treated for pulmonary sepsis. Subsequently, developed mastoiditis and conjunctival hyperemia and used several antibiotics and glucocorticoids. A bronchoscopy with biopsy was realized, which showed an acute and chronic inflammatory infiltrate. She was discharged 40 days later, with clinical improvement.

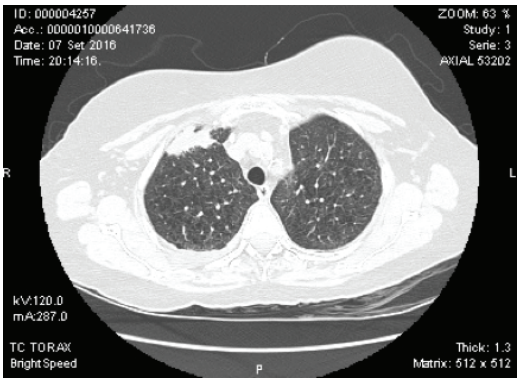

Figure 1. CT chest scan showing a lung mass, in first hospitalization.

One month after discharge, was admitted again with pulmonary septic shock, in addition to sinusitis, polyarthralgia, purpura, skin nodules (Figures 2 and 3) and acute renal injury, requiring hemodialysis during hospitalization.

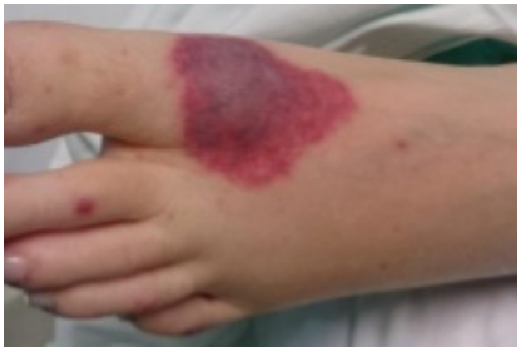

Figure 2: Cutaneous manifestations: purpura and subcutaneous nodules.

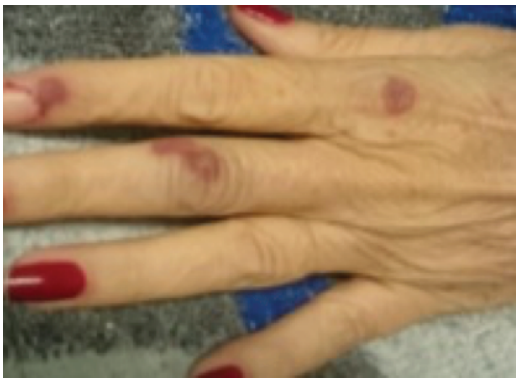

Figure 3: Cutaneous manifestations: purpura and subcutaneous nodules.

In a new CT scan, the pulmonary mass was no longer present and suggests a transient infiltrate and a bilateral pleural effusion (Figure 4).

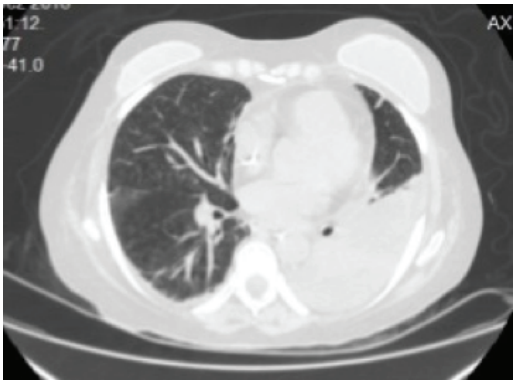

Figure 4: CT chest scan showing a transient infiltrate, with no longer a lung mass, in second hospitalization.

Biomarkers were requested, such as c-ANCA, p-ANCA and rheumatoid factor, with positive result, and antinuclear factor, with negative result. A renal biopsy also has been solicitated, presenting glomerulonephritis. The diagnosis was granulomatosis with polyangiitis, treated with methyl prednisone pulse therapy and cyclophosphamide, progressing to clinical stability and receiving discharge with outpatient follow-up.

DISCUSSION

In this case report, the patient initially presented upper respiratory tract involvement, dyspnea, asthenia and a history of recurrent otitis media. CT chest scans showed an image that, despite being a common presentation, differs from the classical pattern, which consists in multiple, bilateral and cavitate infiltrates2. Thus, despite the suggestive clinical signs, the low prevalence of this pathology turns it into a difficult diagnosis1. The patient develops renal injury, associated with polyarthralgia, sinusitis, purpura and skin nodules, presenting the GPA's triad: upper and lower airways involvement and glomerulonephritis[4].

Considering a rare vasculitis as a differential diagnosis was essential to lead the correct investigation, with a positive c-ANCA serology and the result of glomerulonephritis in renal biopsy. Thus, an early treatment could be managed, even in an uncommon environment for this kind of rare conditions, such as an intensive care unit. In this context, it was possible to avoid a worst prognosis, such as a terminal renal injury, that could lead to permanent hemodialysis or renal transplant, a pulmonary hemorrhage, permanent hearing impairment or deformations due to bone and cartilage destruction[2,8].

CONCLUSION

Granulomatosis with polyangiitis is a rare condition, with a difficult diagnosis, that should be considered when patient presents upper respiratory tract, lungs and kidneys symptoms. Early diagnosis and immunosuppressive therapy are essential to reduce its morbimortality.

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