**Scholars Journal of Medical Case Reports**
Abbreviated Key Title: Sch J Med Case Rep
ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online)
Journal homepage: https://saspublishers.com

OPEN ACCESS

**Radiology**

# Good-Pasture Syndrome: A Rare Entity, about two Observations

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**DOI:** 10.36347/sjmcr.2023.v11i05.032 | **Received:** 11.03.2023 | **Accepted:** 19.04.2023 | **Published:** 11.05.2023

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**Abstract** **Case Report**

Goodpasture syndrome is an uncommon autoimmune disorder that can harm the lungs and kidneys. This condition occurs when the body's immune system mistakenly targets and damages the tissues in these organs. The antibodies produced by the body attack the collagen in the basement membrane of the lungs and kidneys, which results in inflammation and injury to these organs. Symptoms of Goodpasture syndrome include tiredness, coughing up blood, breathing difficulties, and kidney failure. Diagnosis typically involves blood and urine tests to look for the specific antibodies associated with the syndrome, as well as imaging tests to assess the extent of lung and kidney damage**.** We report two cases of Good-Pasture syndrome admitted to the emergency room with alveolar hemorrhage, whose anatomopathological examinations later confirmed the diagnosis.

**Keywords:** Goodpasture syndrome, body's immune system, kidney failure, Diagnosis.



## INTRODUCTION

Good-Pasture's disease, also known as Anti-Glomerular Basement Membrane (anti-GBM) disease, is an extremely rare autoimmune disease characterized by the presence of antibodies that selectively target glomerular and alveolar basement membrane antigens (Shiferaw *et al.*, 2016a).

Good-Pasture's disease was first described by Ernest Good-Pasture, an American pathologist, in 1919(Lettieri & Pina, 2001). It can cause the pneumo-renal syndrome, which is characterized by rapidly progressive glomerulonephritis, pulmonary hemorrhage, and circulating anti-GBM antibodies. This condition can be life-threatening in some cases.

We present two cases of patients with Goodpasture's disease who were admitted to the emergency department due to diffuse alveolar hemorrhage.

## CASE 1

An 18-year-old male with a history of pediatric surgery for tetralogy of Fallot and type 1 diabetes on insulin therapy was admitted to the nephrology department for severe acute renal failure and impure nephrotic syndrome. During her hospitalization, the patient developed dyspnea with two episodes of acute pulmonary edema that did not improve after daily hemodialysis sessions. The dyspnea worsened with the development of moderate hemoptysis, prompting an emergency chest CT scan.

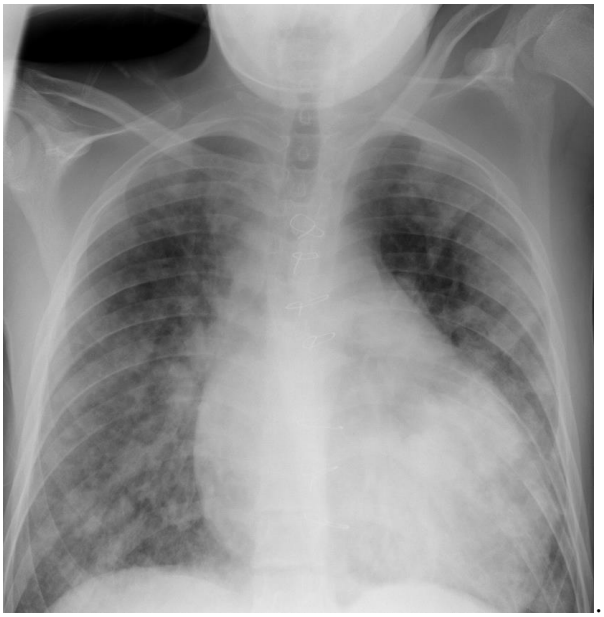

**Figure 1: Chest x-ray showing diffuse nodular alveolar opacities**

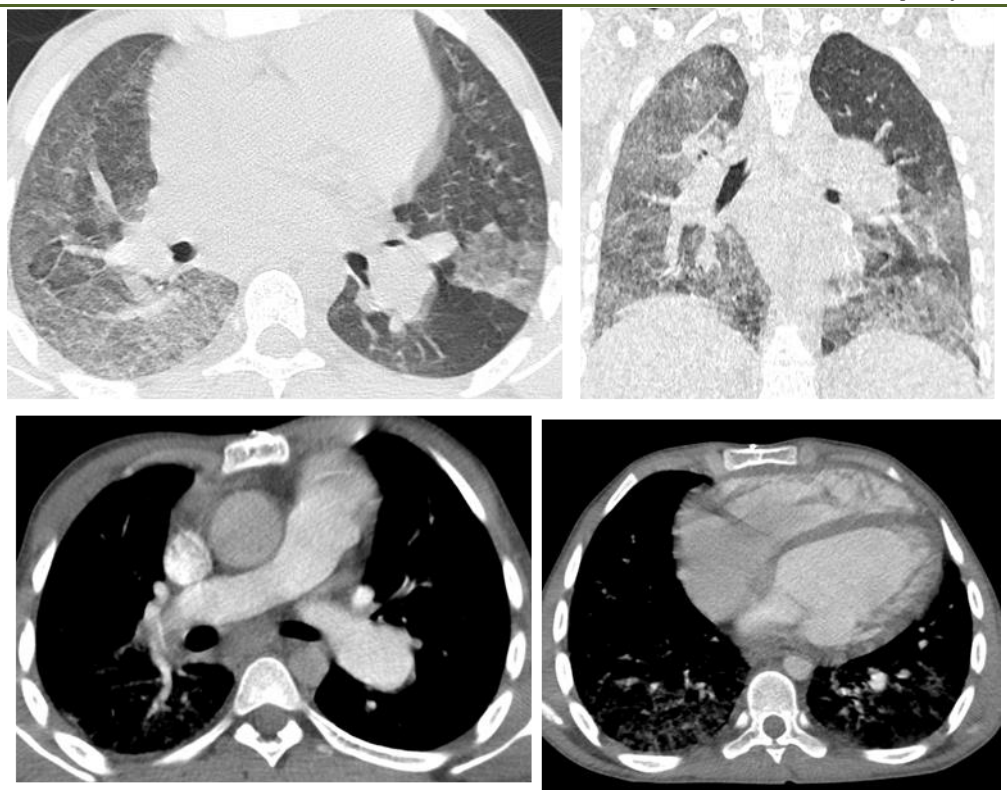

**Figure 2: Chest CT in parenchymal and mediastinal windows showing diffuse ground-glass areas in both lung hemifields, respecting the subpleural regions, in connection with alveolar hemorrhage. No image of proximal pulmonary embolism or cardiomegaly after injection of the contrast medium**

The blood tests showed anemia with a hemoglobin level of 8g/dl and leukocytosis with a count of 16,000. The ionogram revealed an elevated creatinine level of 41 mg/l, hyponatremia at 130, and bicarbonates at 14. The CRP was normal at 0.44.

A thoracic CT scan was performed according to the following protocol: a non-contrast scan, a second pulmonary arterial phase to rule out pulmonary embolism, and a third portal venous phase for better study of vascular structures. The thoracic CT scan showed diffuse ground-glass opacities, more pronounced in the dorsal segments, sparing the apical segment of the culmen and the upper lingula, with reticulations forming a crazy paving pattern, with alternating hypo- and hyperdense areas, and vessels of the same caliber and uniform distribution, creating a mosaic pattern of ground-glass opacities. All these lesions led us to consider, as a first hypothesis, an intra-alveolar hemorrhage that could be part of Goodpasture syndrome.

A renal biopsy was performed, which showed an extracapillary glomerulonephritis with anti-MBG antibodies on direct immunofluorescence. Therefore, a diagnosis of Goodpasture syndrome was established.

## CASE 2

A 22-year-old woman with end-stage renal disease on hemodialysis due to recently diagnosed anti-glomerular basement membrane disease causing isolated glomerular involvement presented with moderate hemoptysis and chest pain for five days. Clinical examination revealed stage III dyspnea with crackling rales on auscultation and 94% O2 saturation.

The laboratory tests showed a normal level of hemoglobin and leukocytes. Creatinine was elevated at 45.2 mg/L, with normal levels of sodium and potassium. CRP was elevated at 78 mg/L, and the estimated prothrombin level was low at 56%.

A CT scan was requested to determine the underlying cause, which showed multiple scattered ground-glass opacities respecting the subpleural regions associated with bilateral basithoracic intra-parenchymal consolidation and a small left pleural effusion without contrast injection.

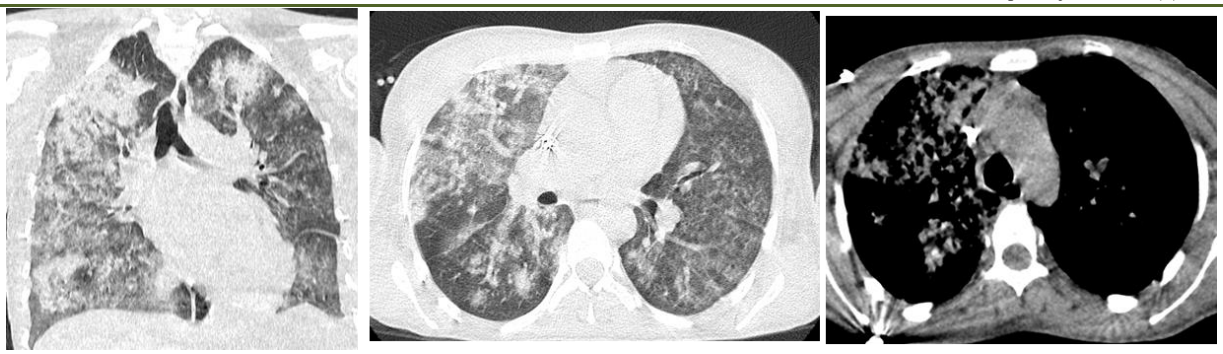

**Figure 3: Chest CT scan in parenchymal and mediastinal windows showing diffuse ground glass areas in both lung hemifields, respecting the sub pleural regions, related to an alveolar hemorrhage**

The diagnosis of Goodpasture's disease was made based on the combination of alveolar hemorrhage, glomerular involvement, and renal biopsy showing an extracapillary glomerulonephritis with anti-MBG antibodies on direct immunofluorescence. The patient was treated with corticosteroids and antibiotics and had a good clinical outcome.

Five months later, the patient underwent a thoracic CT scan due to persistent dyspnea, which showed features suggestive of pulmonary arterial hypertension with mosaic perfusion of the lung parenchyma.

## DISCUSSION

Good-Pasture's syndrome is a rare organ-specific autoimmune disease, caused by specific antibodies called anti-glomerular basement membrane (anti-GBM), with an estimated incidence of 1 case per 1 million per year (Tang *et al.*, 2013).

The terms "Good-Pasture disease" and "Good-Pasture syndrome" were used to refer to the pneumo renal syndrome before the discovery of auto-antibodies. Although these terms persisted for some time, the term "anti-GBM antibody disease" is now more commonly used to describe this condition, whether or not it affects the lungs(Marques *et al.*, 2020).

It usually manifests as rapidly progressive glomerulonephritis (RPGN) which can lead to acute renal failure and life-threatening pulmonary hemorrhages.

Renal involvement alone or with pulmonary involvement is commonly observed in Good-Pasture disease. However, isolated pulmonary hemorrhage as the main manifestation of this disease is rare (Lettieri & Pina, 2001). Some case series have reported an associated gastrointestinal bleeding in Good-Pasture syndrome, although it remains rare, with an estimated incidence of only 3-5% among cases of gastrointestinal bleeding (Gunjan *et al.*, 2014) (Kawabata *et al.*, 2020).

The typical presentation of Goodpasture syndrome consists of a combination of renal and pulmonary failure. Sixty to 80% of patients have apparent clinical manifestations of both renal and pulmonary disease, 20 to 40% have only renal involvement, and less than 10% have a disease limited to the lungs(Lazor *et al.*, 2007).

Renal signs of Good-Pasture syndrome may include hematuria, edema, hypertension, and altered renal function tests. Pulmonary symptoms may consist of cough, dyspnea, hemoptysis, and chest pain with decreased oxygen saturation, although some patients may be asymptomatic(Shiferaw *et al.*, 2016b). It is also possible to observe that pulmonary symptoms may occur before the onset of diagnosed renal signs, and they may also occur after patients have started dialysis treatment.

Both patients in our case presented with a combination of renal and pulmonary symptoms, preceded by rapidly progressive renal insufficiency, followed by pulmonary symptoms dominated by dyspnea and hemoptysis as major serious signs. Exploration of the origin of hemoptysis often involves imaging exams, mainly high-resolution CT scan. Generally, imaging reveals diffuse alveolar hemorrhage, which appears on chest radiography as alveolar opacities in more than 80% of cases, predominantly in the lower lung fields.

A CT scan of the chest without contrast shows diffuse bilateral ground-glass opacities or consolidation, often sparing the subpleural regions and apices in the acute phase (Collard & Schwarz, 2004). The parenchymal opacities often appear suddenly, with major changes in location, extent, and density observed in short-term follow-up studies (Cortese *et al.*, 2008). During the subacute phase, pseudo-nodular opacities without a preferential distribution may appear, associated with areas of ground-glass opacity. In the chronic phase, a Crazy Paving appearance is observed, characterized by areas of ground-glass opacity with thin linear reticulations (Specks, 2001).

Linear reticulations are not due to a secondary thickening of interlobular septa, but to the presence of hemorrhagic material deposited inside the alveoli at the periphery of the lobule (Collard & Schwarz, 2004).

CT, especially high-resolution CT, is considered the radiological examination of choice and the most sensitive for identifying diffuse alveolar hemorrhage, but it is not specific.

In a study of the radiological features of twenty patients with alveolar hemorrhage, Goodpasture syndrome was diagnosed in two patients whose CT scans showed diffuse involvement of both lung fields with sparing of the apices and cardiophrenic regions. Pulmonary consolidations were demonstrated in one patient, while ground-glass opacities were a constant finding in both patients (Cortese *et al.*, 2008). This was the same observation for the cases presented above.

Diffuse alveolar hemorrhage in the context of Goodpasture's syndrome must be differentiated from other causes such as Wegener's granulomatosis and microscopic polyangiitis (Cortese *et al.*, 2008) as well as other etiologies of opacities due to alveolar filling such as cardiogenic pulmonary edema which is often associated with bilateral pleural effusion and cardiomyopathy, and inflammatory exudate from infectious pneumonia (Lee & Specks, 2004).

The CT scan appearance is often suggestive of alveolar hemorrhage and in combination with clinical data such as rapidly progressive renal failure, one may suspect Goodpasture's syndrome. However, the definitive diagnosis is established by the histological detection of Anti-GBM antibodies.

## CONCLUSION

The Good-Pasture disease is a rare cause of renal failure and pulmonary hemorrhage with a poor prognosis in the absence of treatment. Therefore, it is crucial to make a prompt diagnosis and treatment to improve clinical outcomes.

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