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Goodpasture's Syndrome in Children: About a Case Report

Azzahiri I[1*], Benfaddoul O[1], Zouita B[1], Basraoui D[1], Jalal H.[1]

[1]Department of Radiology, Mother and child hospital, Mohamed VI university hospital, Cadi Ayyad University, Marrakech, Morocco

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***Corresponding author:** Azzahiri I
Department of Radiology, Mother and child hospital, Mohamed VI university hospital, Cadi Ayyad University, Marrakech, Morocco

Abstract **Case Report**

Pulmonary-renal syndrome is a medical condition that encompasses a range of clinical symptoms that can appear in various diseases. It is identified by the occurrence of both pulmonary hemorrhage and glomerulonephritis. While this syndrome is rare in children, it can pose a medical emergency if it does occur. Goodpasture's syndrome (GPS) is one of the infrequent causes of pulmonary-renal syndrome in childhood. This article presents a case study detailing the diagnosis of Goodpasture's Syndrome in a 5-year-old pediatric patient with a medical history of nephrotic syndrome, glomerulonephritis at end-stage renal failure, and the presence of anti-glomerular basement membrane (anti-GBM) antibodies. The patient was admitted to the emergency department due to hemoptysis, a hallmark symptom of this syndrome.

Keywords: Goodpasture's Syndrome, hemoptysis, infants, nephrotic syndrome.

INTRODUCTION

The medical condition referred to as "pulmonary-renal syndrome" is a set of clinical symptoms that are commonly observed in various diseases. This syndrome is characterized by the presence of both pulmonary hemorrhage and glomerulonephritis, and while it is a rare occurrence in children, it can be a medical emergency when it does occur. One of the infrequent causes of pulmonary-renal syndrome in childhood is known as Goodpasture's syndrome (GPS). *(Bayat et al.,* 2012)

Ernest Goodpasture was the first to describe a case of pulmonary hemorrhage and renal failure in an 18-year-old male patient during the influenza pandemic in 1919. (Goodpasture, 2009) In 1958, Stanton and Tange reported several patients who died of pulmonary hemorrhage and glomerulonephritis and coined the term Goodpasture's syndrome (GS). (Stanton & Tange, 1958) Over the following decade, the understanding of GS as a distinct disease entity developed, and it was hypothesized that the pathological process is driven by an autoimmune mechanism.

In this article, we present a case of Goodpasture's Syndrome in a 5-year-old boy with a medical history of nephrotic syndrome, glomerulonephritis at end-stage renal failure, and anti-glomerular basement membrane (anti-GBM) antibodies. The patient presented to the emergency room with hemoptysis, which is a common symptom of this syndrome.

CASE REPORT

In this report, we describe a case of Goodpasture's Syndrome in a 5-year-old boy who had a medical history of nephrotic syndrome, glomerulonephritis leading to end-stage renal failure, and anti-glomerular basement membrane (anti-GBM) antibodies. The patient arrived at the emergency room with dyspnea and hemoptysis, but no other symptoms. During the physical examination, the patient displayed nasal congestion and bilateral lung crackles on pulmonary auscultation. Initial laboratory tests revealed normal hemoglobin level, hematocrit value, and low creatinine clearance, while other parameters, such as white blood cell count, platelet count, coagulation profile, C-reactive protein level, and electrolytes were within normal limits. Chest computed tomography (CT) was performed, which revealed multiple diffuse and bilateral nodular ground glass and consolidated opacities suggestive of intra-alveolar hemorrhage. These imaging findings, along with the patient's medical history, led to the diagnosis of Goodpasture syndrome. See Figure 1 and 2 for a visual representation of the CT scan.

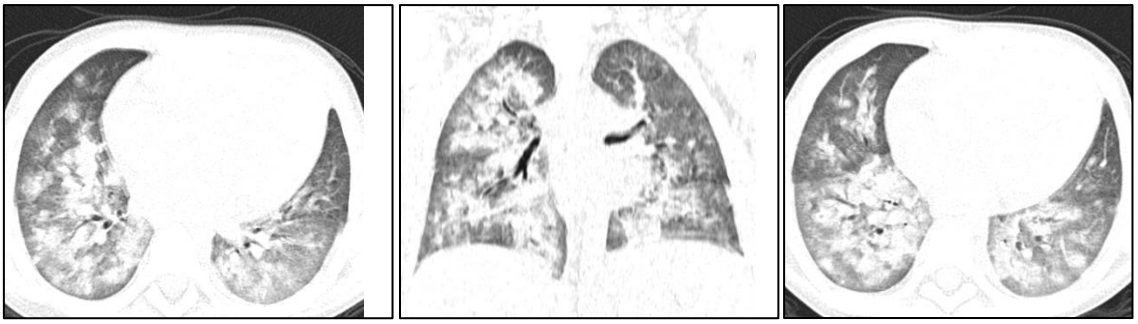

Figure 1: Chest CT scan without injection parenchymal window showing foci of diffuse alveolar hemorrhage with respect to the sub pleural regions.

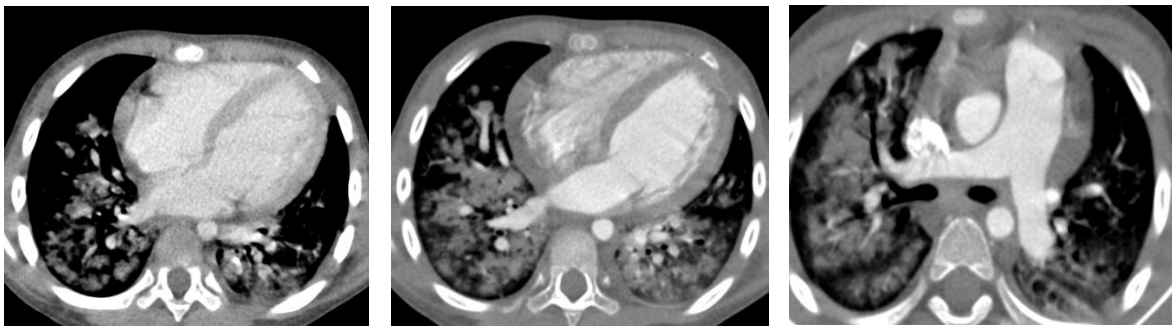

Figure 2: Chest CT with mediastinal window injection showing foci of parenchymal condensation corresponding to foci of alveolar hemorrhage. The trunk and pulmonary arteries are permeable.

DISCUSSION

Goodpasture's syndrome is a medical condition that is characterized by the triad of pulmonary hemorrhage, glomerulonephritis (GN), and the presence of anti-glomerular basement membrane (anti-GBM) antibodies. Goodpasture's disease, on the other hand, refers to the presence of GN and GBM antibodies in the absence of pulmonary hemorrhage. The term antiglomerular basement membrane antibody disease (aGD) is used to describe patients with serum antibodies against the basement membrane and encompasses both Goodpasture's syndrome and Goodpasture's disease.

Pulmonary renal syndromes are a group of disorders that affect both the lungs and kidneys and are characterized by pulmonary and glomerular manifestations. aGD is a classic example of a pulmonary-renal syndrome, while the term specific pulmonary-renal syndrome refers to disorders that associate pulmonary and glomerular manifestations, including Wegener granulomatosis, microscopic polyangiitis, Churg-Strauss syndrome, and systemic lupus erythematosus (SLE). The term nonspecific pulmonary-renal syndrome, on the other hand, refers to either pulmonary thromboembolism, pulmonary edema, or pulmonary infection that complicates the course of glomerular disease, or to glomerular diseases following pulmonary disease, mostly an infection.(Godfrey, 2004)

Although pulmonary-renal syndrome can occur in both childhood and adulthood, it is rare in childhood, and aGD as the cause of pulmonary-renal syndrome is extremely rare. It is important to promptly evaluate and appropriately manage children with hemoptysis to reduce the risk of complications and improve outcomes.

The incidence of anti-glomerular basement membrane disease (aGD) in adults is estimated to be less than 0.5-1 per million per year and is even rarer in children. Among all types of glomerulonephritis, aGD is responsible for 1 to 5% of cases in adults and is the cause in 10 to 20% of patients with crescentic glomerulonephritis. Although aGD occurs across all racial groups, it is most common in European Caucasians. The age distribution follows a bimodal pattern, with a peak in young males and a second peak in females aged 60-80 years. The target of the anti-GBM antibodies in aGD is the NC1 domain of the alpha-3 chain of type IV collagen, which is one of six genetically distinct gene products found in basement membrane collagen. Since this domain is primarily located in the kidneys and lungs, these organs are most affected by aGD. It has been suggested that young infants lack the anti-GBM antigen normally present in older children and adults, and that this transition occurs between the ages of 3 months to 3 years. There is a strong association between aGD and certain HLA alleles, with over 80% of patients carrying HLA-DR15 or DR4, while HLA-DR1 and DR7 are underrepresented, suggesting a possible protective effect against the disease.(Anand *et al.*, 1978) (Thorner *et al.*, 1989)

Anti-glomerular basement membrane disease (aGD), also known as Goodpasture syndrome, is a rare autoimmune disorder that affects the kidneys and lungs. The condition is caused by the production of autoantibodies against the glomerular basement membrane (GBM) and alveolar basement membrane (ABM), which leads to inflammation and damage in these tissues.(Bombassei & Kaplan, 1992)

The principal clinical features of aGD relate to the development of renal failure due to rapidly progressive glomerulonephritis (GN) or pulmonary hemorrhage. GN and pulmonary hemorrhage occur more frequently in young men, whereas GN alone is more common in older women. General symptoms such as malaise, weight loss, fever, or joint pain may be the first signs of aGD, but these symptoms are less frequent and prominent compared to primary vasculitic diseases like Wegener's granulomatosis and polyarteritis nodosa.(Bayat *et al.*, 2012) (Harrity *et al.*, 1991)

In addition to these general symptoms, other specific symptoms of aGD may include nausea and vomiting, weight loss, myalgia and/or arthralgia. The most common physical findings on examination are pallor and anemia, followed by fever, edema, tachycardia, and tachypnea.(Greco *et al.*, 2015)

Radiological features of aGD are non-specific, appearing as bilateral, coalescent airspace opacities on plain radiographs that resolve in several days to give reticular opacities in the same distribution. On thoracic CT scans, ground glass and airspace opacities can be seen, which progress to a reticular "crazy paving" pattern over a few weeks. Hilar lymphadenopathy may be present, but no interlobular septal thickening is seen in the acute phase, which is a distinguishing feature of diffuse alveolar hemorrhage.

The diagnosis of aGBM disease is dependent on the identification of anti-GBM antibodies in circulation or kidney tissue. Renal biopsy is necessary unless contraindicated, as the accuracy of serological tests can vary. Additionally, renal biopsy provides vital information on the activity and chronicity of renal involvement, which can help guide therapy.(Turner & Rees, 1996)

Early detection and management are crucial in the management of anti-glomerular basement membrane (aGBM) disease. Treatment typically involves immunosuppressive therapy to suppress the autoimmune response and reduce inflammation in the affected tissues. Plasmapheresis may also be employed to remove the autoantibodies from the bloodstream. Failure to promptly and effectively treat aGBM can result in irreversible kidney and lung damage, as well as potentially fatal complications such as pulmonary fibrosis and end-stage renal disease.Untreated aGBM disease can rapidly progress to end-stage renal failure. Therefore, early diagnosis is a crucial determinant of therapy response and long-term prognosis. Recommendations for the treatment of aGBM disease are mainly based on studies in adults. The preferred treatment for aGBM antibody disease is plasmapheresis, in combination with corticosteroids and cyclophosphamide.(Lockwood *et al.*, 1976) (Apaydin, 2018)

Prior to the advent of immunosuppressive therapies such as corticosteroids and plasmapheresis, the mortality rate for patients with anti-glomerular basement membrane disease (aGD) was high. The majority of deaths were caused by pulmonary hemorrhage (54%) or renal failure (46%). However, advancements in expert management have dramatically reduced mortality among adults to less than 20%. Conversely, the mortality rate for children with aGD is estimated to be greater than 30%. Prognosis for aGD is affected by several factors, including oliguria at presentation, a serum creatinine level greater than 600 µmol/L (6.8 mg/dL), or more than 50% crescent formation in the glomeruli. While some studies have suggested that serum anti-GBM antibody titers do not necessarily correlate with disease severity, others have found that higher titers are associated with a greater likelihood of pulmonary hemorrhage and a poorer prognosis. Despite the severity of pulmonary hemorrhage, most patients who recover do not experience residual pulmonary deficits or fibrosis. Recurrence of the disease with antibody production is rare, but has been reported in some cases. Overall, aGD is a serious and potentially life-threatening condition that requires prompt diagnosis and expert management to optimize patient outcomes.(Shin *et al.*, 2022).

CONCLUSION

Goodpasture's syndrome (GPS) is a rare condition in children that requires a prompt diagnosis and immediate treatment to prevent the development of renal failure. In recent decades, therapeutic management of GPS has evolved significantly due to the introduction of intensive immunosuppression. This has improved the vital and renal prognosis of patients who receive early treatment. Immunoadsorption, a treatment that is currently underutilized, has the potential to provide more targeted management with fewer side effects.

In addition, it is important to consider the possibility of antiglomerular basement membrane antibody disease (aGD) in patients who present with pulmonary or renal disease, as aGD is often misdiagnosed. Furthermore, aGD can present with pulmonary symptoms alone or with minimal renal involvement, making it important to conduct appropriate testing for aGD in these cases.

DISCLOSURE OF INTEREST

The authors declare that they have no conflicts of interest concerning this article.

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