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Case Report

Eosinophilic Granulomatosis polyangiitis (EGPA) in Pediatrics Population, Rare Disease with a Challenging Diagnosis: Case Report

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ABSTRACT

Eosinophilic Granulomatosis polyangiitis (EGPA) is a rare ANCA-associated multisystem vasculitides that affects mainly adult population. Occurrence in pediatrics population is extremely rare with only few cases been reported in adolescent patients. Eosinophilic granulomatosis with polyangiitis (EGPA)—or, as it was traditionally termed, Churg-Strauss syndrome—is a rare systemic necrotizing vasculitis that affects small-to-medium-sized vessels and is associated with severe asthma and blood and tissue eosinophilia. Like granulomatosis with polyangiitis (Wegener granulomatosis), and the microscopic form of periarteritis, EGPA is an antineutrophil cytoplasmic antibody (ANCA)–associated vasculitide.

Keywords: Eosinophilic; Rare Disease; Granulomatosis; Immunosuppressive

INTRODUCTION

Clinical manifestations include respiratory, renal, gastrointestinal, skin, joints and others. Perinuclear Anti Neutrophil Cytoplasmic Antibodies (C-ANCA) especially directed toward myeloperoxidase antibodies very helpful in diagnosis in addition to biopsy. Systemic corticosteroids and immunosuppressive modulators are the cornerstone in management.

CASE STUDY

A 10 years old Middle Eastern female with a known history of recurrent urticaria and microscopic hematuria presented to the emergency department with 2 days of non bilius non bloody emesis associated with right upper quadrant abdominal pain. Patient denied any fever, diarrhea or sick contact. Family denied any recent travel but admitted to have pigeons as pets at home. Regarding her medical history, patient has been followed by Allergy/ Immunology service with possible autoimmune urticaria treated with oral anti-histamine. She is also been followed by pediatrics nephrology for microscopic hematuria and proteinuria. No kidney biopsy was done. She always maintained normal blood pressure and renal function. Significantly, her younger brother has a history of gross hematuria and was found to have mesangiopathy/ avssculopathy on renal biopsy. He also has uveitis. In the emergency department patient was noted to be pale with tachycardia. Initial diagnostic blood work was done. Complete blood count showed white blood cell counts of 8.4 with neutrophilic predominance and eosinophils 6%, hemoglobin of 6.8 MCV 54. Elevated reticulocytes of 1.8%. Comprehensive metabolic profile showed normal electrolytes, liver enzymes, pancreatic enzymes and renal function test. Urinalysis showed moderate blood without protein. CRP 160 (Normal value <9) and ESR 41. Complement C3 was slightly decreased with normal C4. Chest Xray showed pneumonitis with interstitial infiltrates suggesting atypical pneumonia or pulmonary hemorrhage. Complete abdominal ultra-sonogram was done and showed normal liver, kidney and bowel. Patient was admitted to the pediatrics service for further investigation with differential diagnosis of infectious versus rheumatologic etiology. Pediatrics nephrology, pulmonology, infectious disease and rheumatology were consulted. Shortly after arriving to the pediatrics floor patient started to exhibit signs of respiratory distress with increase work of breathing and decrease oxygenation. Patient was started on oxygen via Venturi mask and Pediatrics ICU team was consulted. Patient also received blood transfusion. Chest CT with contrast was recommended by Pulmonology that showed signs of multifocal pneumonia and possible alveolar hemorrhage. Patient was intubated in the ICU and stabilized. Alveolar hemorrhage was confirmed later by bronchoscope (Figure 1).

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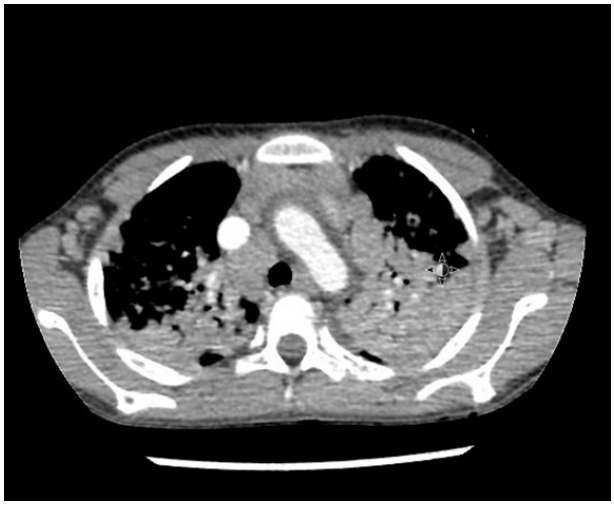

Figure 1: Alveolar hemorrhage was confirmed later by bronchoscope.

Patient was started on empiric antibiotics including Vancomcyin, Ceftriaxone and Azithromycin to cover for possible pneumonia. Respiratory panel PCR was positive for Rhinovirus. Extensive infectious work up was also done including Legionella, Histoblasmosis, Coccidoimidosis, blastomycosis, Cryptococcus, sputum culture, blood culture, HIV, Tuberculosis, Mycoplasma titers and Chlamydia psittaci with negative results. Rhinovirus is a very common viral cause of upper respiratory infection, however it was considering as a coincidental finding and less likely contributor to patient's presentation. Pediatrics Rheumatology evaluated the patient and had a high suspension of autoimmune process. Autoimmune pneumonitis, EGPA, Wagner's disease, Sarcoidosis and idiopathic vasculitis were on the differential diagnosis. Bronchoscopy with bronchoalveolar lavage was done and showed numerous hemosiderin- laden macrophages. Diagnosis of EGPA was confirmed with highly positivity for P-ANCA with presence of myeloperoxidas antibodies. Patient was started on high dose of methyl prednisone and Rituximab. Patient was successfully extubated and her respiratory status started to improve. Inflammatory markers started to trend down and they were back to normal upon discharge. Patient was discharged home on course of Systemic Steroids and weekly Rituximab. As pigeon feathers antigens may be responsible of triggering esinophilic reaction, family was asked to give up the pigeons. Patient will be following up with Pediatrics Rheumatology team, Pediatrics Pulmonology, pediatrics Immunology and Nephrology.

RESULTS AND DISCUSSION

ANCA-Associated Vasculitides (AAVs) are multisystemic disease include Microscopic Poly Angiitis (MPA), granulomatosis with polyangiitis (GPA) and Esinophilic Polyangiitis (EGPA) [1]. AAV's are extremely rare in pediatrics population and has been described in case reports. The annual incidence of AAV reported in a recent French study including 66 children, the largest in Europe is 0.5 per million children [2]. Higher incidence rate up were reported in a Canadian study [3]. AAV's are more common in female and their onset peaks in pediatrics papulation is at age of 11-14 years [1,2]. Another large databases reveals that childhood onset EGPA accounts of less than 2% of all cases of pediatrics vasculitis [4]. As pathogenesis of EPGA is not entirely clear, environmental triggers, viral infections and genetic predisposition are believed to be contributors to the developmental and progression of the immune cascade. These triggers are responsible of dysregulation of the immune system including pathogenic production of ANCA. ANCA (Antineutrophil cytoplasmic antibodies) are immunoglobulin G antibodies directed toward components of the neutrophils, they are produced by B cell lymphocytes PR3-ANCA account of the majority of ANCA with cytoplasmic immunofluorescence C-ANCA and are commonly associated with EGPA [6,7]. EPGA are systemic diseases with multiorgans involvement. Lungs, kidneys, skin, joints and other organs are commonly affected. It's generally characterized by ear-nose- throat involvement followed by constitutional symptoms [8,9] renal, lower respiratory tract, muscoskeletal and cutaneous involvement [8]. Respiratory tract involvement includes rhinitis, sinusitis, hearing loss, asthma and rarely pulmonary hemorrhage. Renal disease is very common 50-100% of pediatrics patients that range from isolated renal abnormality to rapidly progressive renal glomerulonephritis. Skin involvement present with palpable purpura, nodules to urticarial rash. Diagnosis depends on laboratory findings after clinical suspicion. ANCA positivity is common with PR3-ANCA positive in about 70% of pediatrics cases. Hematological, renal and inflammation markers abnormalities are quite common. Renal involvement is common and usually have better prognosis with EPGA than other AAVs. Proteinuria, hematuria, low complements and even hypertension have been described. Renal biopsy helps making the diagnosis. In our case, renal biopsy was delayed by nephrology team as patient is responding well to treatment and will be considered if renal symptoms persist or renal function started to decline [10,11]. Treatment options of AAVs have evolved and showed major advances in the last decade. Systemic glucocorticosteroids are still the cornerstone of the treatment with high dose steroids. Immunomodilators have proven good results in controlling disease flares up including renal and respiratory symptoms. Our patient showed a promising response to Steroids and Rituximab with normalization of her inflammation markers [12,13].

CONCLUSION

EPGA is a very rare ANCA-mediated vasculitis in pediatrics population. The clinical presentation of asthma symptoms with hematuria and history of chronic urticarial rash were helpful in making the diagnosis. PR3-ANCA is usually positive in majority of cases. Lung biopsy and renal biopsy are helpful in making the diagnosis of EPGA. Systemic steroids and immunosuppressive mediations are essential for treatment and controlling of symptoms. In spite of their rare incident, Clinicians should consider ANCA vasculitis on their work up for pediatric patients presenting with wheezing and hematuria.

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