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Case report

Adult onset immunoglobulin A vasculitis (Henoch-Schonlein purpura) with alveolar hemorrhage

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A B S T R A C T

S. pyogenes is the cause of many important human diseases, ranging from mild superficial skin infections to life-threatening systemic diseases. The post streptococcal syndromes are immune mediated phenomena including Immunoglobulin A Vasculitis (Henoch-Schönlein purpura).

HSP is more common in children and usually self limited but it can cause skin, joint, renal, gastrointestinal and rarely respiratory involvement. We present a case with *Streptococcus pyogenes* pneumonia that presented with respiratory failure, pulmonary hemorrhage, extensive rash and renal failure.

Introduction

The Group A beta hemolytic *Streptococcus* (GAS), also known as *Streptococcus pyogenes*, causes a broad range of infections and complications such as glomerulonephritis and vasculitis. Immunoglobulin A vasculitis (IgAV), formerly called Henoch-Schönlein purpura (HSP) [1], is the most common form of systemic vasculitis in children. It is less common in adults but adults are reported to have worse renal outcomes [2,3]. About one-half of the cases of IgAV are preceded by an upper respiratory tract infection, especially those caused by GAS. Other infectious agents, vaccinations, and insect bites also have been implicated as possible triggers for HSP IgAV [4,5].

HSP usually involves kidneys, joints, gastrointestinal system and skin but severe lung involvement such as pulmonary hemorrhage is very rare [6]. We present a rare case of GAS associated IgAV causing systemic vasculitis and alveolar hemorrhage.

Case presentation

76 year old female originally from China with past medical history of diabetes mellitus, hypertension, gout, atrial fibrillation and permanent pace maker insertion presented with 2 weeks of rash. About a week prior she was seen by her primary care doctor and was started on prednisone, diphenhydramine and furosemide. As the rash did not subside, she went to an emergency room but was discharged with similar regimen. Her rash progressively worsened and she developed cough, shortness of breath, dysphagia, arthralgia and hemoptysis.

On exam she was normotensive and afebrile but hypoxic to 89%. She was ill appearing with bilateral rales on lung examination, petechiae on conjunctiva and oral mucosa, and a diffuse purpuric and petechial rash on her palms, soles, chest and abdomen with bullae [Figs. 1–3]. Initial laboratory work up showed, white blood cell count of 9.1 K/uL with 87% neutrophils, hemoglobin: 9.2 g/dL, Platelet: 185 K/uL, Sodium: 117 mmol/L, Potassium: 5.6 mmol/L, Blood Urea Nitrogen: 75 mg/dL, Creatinine: 2.24 mg/dL, with normal liver function test. The urinalysis had 5–10 Red Blood Cells and CXR showed bilateral airspace opacities suggestive of multifocal pneumonia. She was intubated for respiratory distress and started on hydrocortisone, hypertonic saline, vancomycin, piperacillin/tazobactam empirically and also doxycycline added in view of her rash. Chest CT also showed extensive patchy bilateral pulmonary opacities. Further work up demonstrated negative blood cultures, respiratory viral panel, *Mycoplasma, Legionella, Anaplasma, Ehrlichia,* Rocky Mountain Spotted Fever, *Leptospira* and *Treponema* serologies. Patient underwent bronchoscopy which was consistent with alveolar hemorrhage. The BAL was negative for AFB smear. Skin biopsy showed small vessel acute vasculitis with differential diagnosis of microscopic polyangiitis, hypersensitivity angiitis, leukocytoclastic vasculitis, IgAV, and less likely an unusual presentation of Wegener's granulomatosis. As the patient's renal function worsened and the etiology of respiratory failure with alveolar hemorrhage was not clear, patient was started on emergent plasmapheresis.

Further work up showed an antinuclear antibody (ANA) of 1:320 with speckled pattern, high rheumatic factor: 25.6 but negative anti CCP and ANCA. Cultures from the BAL revealed GAS and ASO subsequently rose from 852 to 1220 IU/mL. Antibiotics were switched to ceftriaxone for 7 days. Renal biopsy revealed proliferative glomerulonephritis with immune deposits suggestive of IgA nephropathy. The skin biopsy was repeated and showed weak dermal vessel stain for IgA

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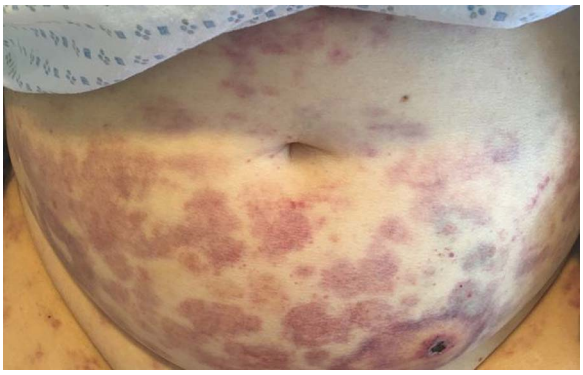

Fig. 1. xxx.

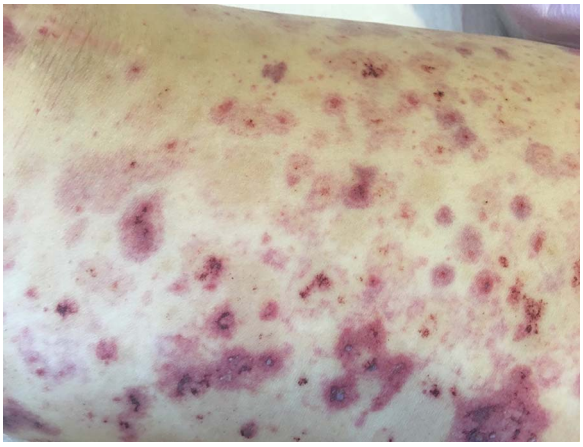

Fig 2. xxx.

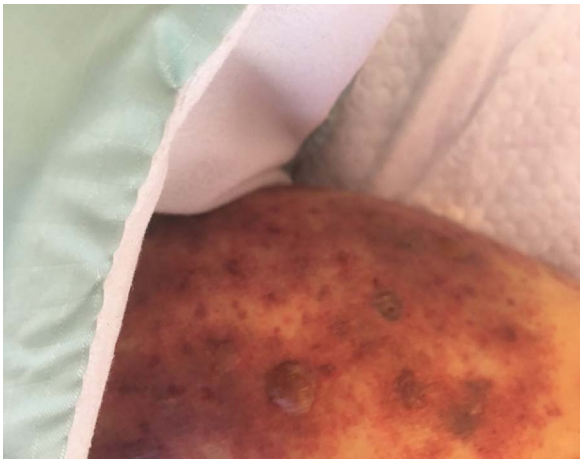

Fig. 3. xxx.

and C3. Ultimately the diagnosis was determined to be post streptococcal vasculitis with IgAV and the patient was tapered on prednisone after the pulse steroid therapy. The course of ceftriaxone was completed and hemoptysis, rash and renal failure resolved. Patient was discharged with no other immune suppressants.

Discussion

Streptococcus pyogenes causes an array of infections involving the respiratory tract and soft tissues ranging in severity from mild to severe. The streptococcal virulence factors and antigen mimicry are responsible for various complications including rheumatic fever, post streptococcal glomerulonephritis, post streptococcal vasculitis and IgAV [5].

IgAV is a common vasculitic syndrome of childhood which is mostly self limited but can sometimes occur in adults and rarely cause pulmonary involvement which manifests as diffuse alveolar hemorrhage (DAH) and occasionally as usual interstitial pneumonia or interstitial fibrosis [6].

Conclusion

Streptococcus pyogenes can cause different immune mediated syndromes including vasculitis and IgAV. IgAV can be life threatening in adults due to pulmonary hemorrhage and multi organ failure including renal failure with poor outcome.

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