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CASE REPORT

Classic IgA-Vasculitis With Nephrotic Range Proteinuria - Rare Presentation in an Adult

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Abstract

IgA vasculitis formerly known as Henoch-Schonlein Purpura is characterized by leukocytoclastic vasculitis and IgA immune complex in small vessels of the affected organ. IgA vasculitis can involve any organ system depending upon the deposition of the IgA immune complex. IgA vasculitis is a clinical diagnosis which manifest with abdominal pain, arthralgia/arthritis, palpable purpura, and kidney involvement. Occasionally, serum IgA levels or skin or kidney biopsy can help in confirming the diagnosis. Treatment is usually supportive, but studies have proved that prednisone or immunosuppressive agents can help in the prevention or progression of the disease. Hereby we present a case of 54-year-old Caucasian male who developed classic tetrad findings of IgA vasculitis most likely after receiving monkeypox vaccine which patient received three weeks prior to presentation to the hospital. Kidney involvement was present but surprisingly proteinuria was above nephrotic range making it as a rare presentation of IgA vasculitis.

Keywords: Classic, Vasculitis, Nephrotic, Proteinuria, Rare, Presentation, Adult

1. Introduction

IgA vasculitis is leukocytoclastic vasculitis characterized by the formation of IgA dominant immune deposits in small sized blood vessels.[1,2] It is the most common form of vasculitis among children but can happen less frequently in adults.[3] It manifests with palpable purpura, arthralgia/arthritis, abdominal pain, and renal problems. Usually, earliest symptoms include skin and joints, but occasionally gastrointestinal symptoms may precede cutaneous manifestations.[4] IgA vasculitis is a clinical diagnosis, but skin or renal biopsy might help to confirm the diagnosis. Treatment is supportive, but studies have proved that patient responds well to prednisone or other immunosuppressive drugs.[4] Here, we report a case of 54-year-old Caucasian gentleman with newly developed IgA vasculitis secondary to possibly monkeypox vaccine which patient received three weeks prior to presentation to the hospital, with typical presentation but having nephrotic range proteinuria which is very uncommon in adult population.

2. Case presentation

54-year-old Caucasian gentleman with significant past medical history of recent COVID-19 infection last month presented to the emergency department with complaint of upper abdominal pain for one week, associated with nausea and vomiting. On further inquiry, patient reported that his initial symptoms including intermittent blood in the stool, arthralgias, petechial rash and swelling in bilateral lower extremity started three weeks prior to the emergency department visit, just after receiving a Monkey Pox vaccine. He was prescribed short course of prednisone 40 mg daily for a total of two weeks by primary care physician leading to transient resolution of the symptoms which recurred after the course of prednisone was completed.

Physical examination was significant for diffuse tenderness in the abdomen, hyperactive bowel sounds, bilateral lower extremity swelling with palpable purpura [Fig. 1]. Rest of the physical examination was unremarkable. Laboratory investigation confirmed white blood cell count of 17.9 k/uL, Creatinine of 1.58 mg/dl with baseline creatinine of

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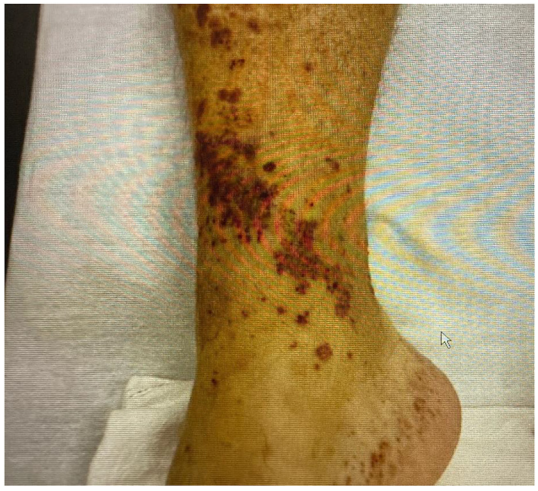

Fig. 1. Palpable purpura in lower extremity.

0.9 mg/dl, C-Reactive Protein of 9.4 mg/dl. Urine analysis showed large blood, 300 mg/dl of protein, >100 RBCs, 10–20 WBCs with no signs of urinary tract infections, raising the concerns for glomerulonephritis. Complement levels were unremarkable. Rest of the laboratory investigations were unremarkable. CT of the abdomen showed signs of small bowel obstruction with colonic diverticulosis. In the light of small bowel obstruction, general surgery got consulted and recommended conservative management including making the patient Nothing Per Oral (NPO), Intravenous fluids, pain management, which led to resolution of symptoms in couple of days.

Patient was started on IV Methylprednisolone 125 mg due to concerns of glomerulonephritis most likely secondary to vasculitis and nephrology was consulted. Nephrology planned for 24-h urine protein which showed 5 g of protein consistent with Nephrotic range proteinuria. Considering Nephrotic range proteinuria, suspicion for IgA nephropathy was less likely despite of classic presentation of the patient, hence patient was planned for skin as well as renal biopsy. Skin biopsy came back positive for leukocytoclastic vasculitis. Rheumatology was consulted for the evaluation and management of vasculitis. Rheumatology planned for ANA, Rheumatoid factor, SPEP, UPEP, ANCA antibodies and results came back unremarkable. Patient was transitioned from IV Methylprednisolone to oral prednisone 40 mg daily. Kidney biopsy came back positive for IgA vasculitis with nephritis. Patient got diagnosed with IgA vasculitis with nephritis and having proteinuria of nephrotic origin which is very rare and is typically associated with worst prognosis. Patient responded well to steroids; his symptoms of small bowel obstruction was resolved. Patient became stable during the hospital stay. Patient was scheduled to be discharged on steroids with follow up with primary care physician, nephrology, and rheumatology with a rationale to considering adding on azathioprine for disease control, awaiting results of Thio-Purine Methyltransferase (TPMT levels).

3. Discussion

IgA vasculitis, formerly known as Henoch-Schonlein Purpura, is characterized by leukocytoclastic vasculitis and IgA immune complex deposits in the affected organs. It has a male predominance with predilection among children. Among children, it manifests as classic tetrad of palpable purpura, arthralgia or arthritis, abdominal pain, and kidney disease in contrast to the adults, among which presentation is usually atypical. Palpable purpura is the most common feature of IgA vasculitis followed by arthralgia or arthritis. Arthralgia or arthritis usually manifest in a migratory pattern, which is often accompanied by periarticular swelling and tenderness but less frequently displays with joint effusion, erythema, or warmth.[5,6] Abdominal pain or bleeding associated with IgA vasculitis is usually caused by submucosal hemorrhage or edema. It can also be accompanied by submucosal edema, ulceration, and ileal and jejunum spasms.[7] Intussusception is the most common gastrointestinal complication of IgA vasculitis among children but considered as a rare presentation among adults.[8] However, kidney disease in contrast is more prevalent in adults with IgA vasculitis when compared to children. IgA vasculitis patients having kidneys involvement usually manifest with hematuria with or without red blood cell casts and mild proteinuria, Nephrotic range proteinuria is very uncommon in patients with IgA vasculitis (which was seen in our patient) and is usually correlated with increased risk of progressive kidney disease, ultimately leading to end stage renal disease.[8,9]

IgA vasculitis is typically a clinical diagnosis, but laboratory investigations can be used to support the diagnosis of IgA vasculitis.[10] Serum IgA levels have been found to be elevated in 50–70% of patients and are usually associated with renal involvement.[11] Other laboratory markers can potentially point to the trigger that pushes the patient into IgA vasculitis. Patients who had a preceding bacterial infection would demonstrate leukocytosis and an elevated ESR while patients who had a preceding viral infection would not have any acute phase reactant elevation.[12] Among adults, skin or renal biopsy are necessary to confirm the diagnosis due to

the rarity of the disease among adult population. Skin biopsy of the purpuric lesions will demonstrate leukocytoclastic vasculitis and IgA deposition,[13] while renal biopsy will show IgA deposition in the renal mesangial as was observed in our patient.[13] Treatment for IgA vasculitis is dependent on the symptoms. We begin with non-steroidal anti-inflammatory agents for abdominal or joint pain and treat with prednisone if the pain is severe.[14]

Our patient developed IgA vasculitis after administration of monkeypox vaccine. It is not certain what triggers the onset of IgA vasculitis in our patient but most common triggers in literature includes infection or chemical triggers.[15] There are several case reports that show IgA vasculitis occurring after vaccine administration. One case report found a significant increased risk of IgA vasculitis after administration of the MMR vaccine in children.[16] Our patient had classic clinical presentation, which is very uncommon among adults, Also, despite having IgA vasculitis, patient had nephrotic range proteinuria, which is usually seen in less then 1% of the patient getting diagnosed with IgA vasculitis. IgA vasculitis cases includes children in 90% of the cases. The occurrence in adults is about 3.4–14.3 cases per million. Although classic presentation among adults is very uncommon, high clinical suspicion and recognition of the symptoms of IgA vasculitis is important to start the treatment, to prevent the morbidity associated with the disease which could include end stage renal disease or acute abdomen secondary to small bowel obstruction or intussusception leading to bowel perforation.

In conclusion, IgA vasculitis is a leukocytoclastic vasculitis which has predilection for children but occurrence among adults is uncommon, its typically a clinical diagnosis and manifest with classic tetrad including abdominal pain, palpable purpura, kidney disease and arthralgias except among adults. Kidney involvement commonly causes glomerulonephritis but nephrotic range proteinuria if present is an indication of poor prognosis ultimately causing end stage renal disease. Skin or renal biopsy can augment to confirm the diagnosis. And if there is suspicion depending upon clinical presentation, treatment should be started with steroids to prevent progression of the disease even before biopsy results came back.

Conflict of interest

There is no conflict of interest while conducting this study.

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