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Case Report

IgA Vasculitis Associated *Salmonella enteritidis* Infection: A Case Report

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Abstract

Immunoglobulin A (IgA) vasculitis, or Henoch-Schönlein purpura (HSP), is a vasculitis with pathophysiology not well elucidate and infections are one of proposed triggers. We reported the case of a 55-years old man who had fever, diarrhea improved by ciprofloxacin then a vascular purpura associated knee's arthritis. He presented an acute left abdominal contracture with circulatory collapse. CT-scan revealed renal and hepatic aneurysms complicated by a rupture and a voluminous retroperitoneal left hematoma. He had an urgent embolization with clinical and radiological good results. Skin biopsy showed leukocytoclastic vasculitis with IgA deposition. **C**ytobacteriologic urine examination revealed the presence of *Salmonella enteritidis.* An articular ultrasound for the right hip joint did not show an effusion but MRI described a right coxofemoral osteoarthritis with myositis and a collection in the homolateral iliopsoas muscle. Treatment was ciprofloxacin for continued 3 months and steroid with well evolution. The review of the literature did not found aneurism as vascular manifestation of HSP whereas abdominal aneurism was associated to *Salmonella* infection.

Keywords: IgA Vasculitis; *Salmonella*; Aneurism; Embolization; Coxofemoral Osteoarthritis

Introduction

Immunoglobulin A (IgA) vasculitis, formerly called Henoch-Schönlein purpura (HSP), is a vasculitis that affects small vessels with Ig A-dominant immune deposits. It is the most common childhood vasculitis but can also be observed in the adult [1,2]. There is no specific laboratory tests and diagnosis is generally based on the clinical presentation. There is no specific treatment for HSP. In general, the treatment is symptomatic. However, corticosteroids, immunosuppressive and modulating drugs can be used in severe forms especially with renal involvement [3].

Most cases of HSP occur in autumn and winter. Proposed triggers include upper respiratory tract infections, medications, vaccinations, and malignancies [2,4]. The pathophysiology behind HSP is not yet completely understood [3].

Cases of HSP associated with *Salmonella* infection has been reported just in a limited number of cases [4,5]. We proposed report the case of an adult patient, presenting HSP complicated with a rupture of an abdominal aneurysm and a concomitant *Salmonella enteritidis* infection.

Case Presentation

A 55-years old man was admitted for vascular purpura associated knee's arthritis. He was chef in a hotel. He has personal medical history of hypertension, since five years,

without particular medical family history. Ten days before his admission, he presented fever and diarrhea improved by 7 days of ciprofloxacin. In our department, examination showed fever at 38.2°C, correct hemodynamic state without meningeal syndrome nor heart murmur. Right knee's arthritis and purpura over the lower limbs were noted and the rest of physical examination was normal. Hematuria was detected on urinary sediment with traces of proteinuria. Infectious endocarditis was eliminated by transesophageal echocardiogram.

Blood count showed normal platelets rate and elevated rate of white blood cells (15800e/µL). Inflammatory anemia was noted (Hb=11,8 g/dL) associated to elevated CRP (204 > 6 mg/L) and procalcitonine (3,66 > 0.5 ng/mL). No hepatic problem nor renal failure was initially noted and with negative bacteriologic urine examination. Viral serology for HIV, B and C hepatitis, Cytomegalovirus, Parvovirus B19 and Syphilis was negative. Immunological investigation was negative for rheumatoid factor, Anti-neutrophil cytoplasmic antibodies (ANCA) and anti-nuclear antibody, associated to normal complement levels (C3=1,7 g/L, C4 =0,48 g/L).

On the fifth day investigation, the patient presented an acute left abdominal contracture with circulatory collapse. Hemoglobin passed from 11 to 9 g/dl. An urgent abdominal CT-scan revealed renal and hepatic aneurysms. The most voluminous was in the upper polar segmental renal artery complicated by a rupture and a voluminous retroperitoneal left hematoma. The patient had an urgent embolization with clinical and radiological good results (Figure 1).

Figure 1: (a) Preembolization image showing an extraction of the contrast medium. (b) Post embolization image illustrating regression of contrast medium extraction.

Polyarthritis Nodosa had been the most suspected diagnosis for associating aneurysms and purpura (medium and small sized arteries involvement). No other vascular aneurism was noted. ANCA vasculitis protocol of treatment was started using steroid on bolus doses and cyclophosphamide repeated after two weeks.

However, skin biopsy showed leukocytoclastic vasculitis with IgA deposition in direct immunofluorescence. The diagnosis turned into IgA vasculitis. The man presented an acute urinary infection associated to right hip joint pain. Cytobacteriologic urine examination revealed the presence of *Salmonella enteritidis.* An articular ultrasound for the right hip joint did not show an effusion but MRI described a right coxofemoral osteoarthritis with myositis and a 20 mm collection in the homolateral liliopsoas muscle (Figure 2). No endocarditis nor infected retroperitoneal hematoma nor nephritis was detected on renal and cardiac ultrasounds.

Figure 2: MRI showing right coxofemoral osteoarthritis with myositis.

In conclusion, urinary and articular *Salmonella* was treated by ciprofloxacin at the dose of 750 mg twice a day for continued 3 months. The treatment of Henoch-Schönlein purpura was continued by only steroid with a rapid regression doses. Hematoma regressed on abdominal ultrasounds. Treatment of hypertension was switched from conversion inhibitor enzyme to a calcic inhibitor. The patient had well evoluted with the total regression of hematoma and psoas abscess. After 2 years evolution, MRI showed the absence of all aneurism. Renal scintigraphy revealed normal kidneys function whereas hypertension still no controlled on three therapy.

Discussion

Ig A vasculitis,or Henoch–Schönlein purpura, is an immune complex vasculitis caused by the deposition of IgA immune complex

in the walls of vessels [6]. It is predominantly affecting small vessels, marked by palpable purpura, arthritis, glomerulonephritis and gastrointestinal manifestations [3].

It occurs mostly in infants. It is less frequent in adults in whom it may be associated with a worse outcome and it is more frequent in males, with a male/female ratio of 1.5 [7].

Although the cause of the disease remains unknown, it is clear that IgA system plays a central role in the pathophysiology. Increased IgA synthesis could be related to antigen exposure processed by the mucosa associated immune system. Bacteria, virus or parasitic agents were suspected to trigger the disease in genetically prone individuals, but causative agents and factors remain to be identified [6]. In this reported case, *Salmonella enteritidis* infection was suspected inducing IgA vasculitis.

Clinical spectrum of the disease mainly includes cutaneous purpura, arthralgias and/or arthritis, acute enteritis and glomerulonephritis, with gastrointestinal and renal involvements representing the main causes of morbidity and mortality in adults. Myocarditis, orchitis, alveolar hemorrhage or episcleritis represent very rare manifestations of IgA vasculitis. Central nervous or peripheral nervous system involvement may also occur [1].

Looking in the literature, aneurism was not described as vascular manifestation of HSP.

IgA vasculitis diagnosis is based on clinical criteria. The revised criteria developed by EULAR/PRINTO/PRES were published in 2010, and are the gold standard for the diagnosis of HSP. The sensitivity and specificity are respectively 100% and 87% when applied to children and 99.2% and 86% in adult [8]. There are currently no specific biomarkers useful for diagnosis of HSP. Skin biopsies are the gold standard for diagnosing any cutaneous vasculitis. IgA-predominant vascular deposits are characteristic for HSP, but not sufficient for its diagnosis. However, since it is an immune-mediated disease, corticosteroids and immunosuppressive therapy can be used in severe forms with especially renal manifestation [3]. Treatment should concider causative agent when it is diagnosed as in this case of incriminated *Salmonella enteritis* infection in IgA vasculitis occurence.

Non-typhoidal *Salmonella* (NTS) is an entero invasive bacterium and causes infections that may have different clinical presentations. It is known that *Salmonella* infection occurs mostly in immunocompromised patients including who have systemic diseases particularly SLE [9-11]. Gastroenteritis is the most common presentation. Bacteremia and focal infections are less frequent but more severe presentations. Focal infections may affect various sites in the body, creating different disorders, which usually occur during or after *Salmonella* bacteremia [12,13].

IgA vasculitis associated to *Salmonella* infection has been rarely reported in the literature. Three cases were published, which occurred in children, and two other cases happened in adult one of them was due to *Salmonella hirschfeldii* in a 19 years old male [4,14]. The other case was association with *Salmonella typhi* [5].

In this reported case, *Salmonella enteritidis* seems to be the trigger cause of HSP. Clinical manifestations were abdominal pain, diarrhea and arthritis. Secondary localizations were urinary, articular, psoas abces and abdominal aneurism. Fatal complication was aneurism rupture.

Abdominal aneurism in this case could be correlated to *Salmonella* infection. Bacterial aneurysm was first described by Osler in 1885. Syphilis, Tuberculosis, and untreated endocarditis were the most common causes of mycotic aneurysms. Recently, the most common cause reported worldwide has been *Staphylococcus aureus*, followed by *Salmonella* [15]. *Salmonella* has a strong affinity for large blood vessels, and can easily adhere to the damaged vascular wall. Recent literature review of Mycotic aneurysm due to *Salmonella* species showed that it was more frequent in male over sixteen years old. Hypertension, diabetes, atherosclerosis, hyperlipidemia, smoking, acquired immunodeficiency syndrome, chronic kidney disease, drug abuse, and autoimmune diseases treated by biological agents were considered the most common comorbidities present in patients presenting *Salmonella* mycotic aneurysm [16-18].

Symptoms were fever and diarrhea. Species that are more isolated were *S. enteritidis* and *S. choleraesuis*. Aneurism localisation was Abdominal aorta but in few cases in Thoracic aorta, aorta arch, Iliac and beyond artery. Complications were aneurysm rupture, psoas abscess, surrounding abscess and spondylodiscitis. Treatment of patients, with mycotic aneurysm caused by *Salmonella*, should include antibiotic therapy and surgery. The surgical options for mycotic aneurysms included in situ reconstruction, extra-anatomic

bypass, and endovascular aneurism repair (EVAR) [16]. EVAR was routine treatment for patients who were not suitable for open surgeries. However, the risk of infection recurrence is relatively increased. Huang et al. reported that the 30-day mortality of EVAR-treated mycotic aneurysms was 0%, and the 1-year survival was 81.8% [19]. Furthermore, the potential factors of persistent infection after EVAR include fever during operation and aneurysm rupture [16].

Conclusion

This case elaborates on the fact that we should always seek an infectious trigger to HSP especially severe forms since we are to use immunomodulating drugs that can lead to a more important infectious risque. *Salmonella* infection should be considered when searching for the trigger and should be adequately treated to avoid complications and focal localizations since this can be particularly severe in immunocompromised patients. More studies on HSP cases in adults are needed for a better comprehension of this disease and more adequate treatment.

Ethical Approval

The case report is anonymous and the patient approved the publication.

Competing Interests

No competing interests were present.

Authors' Contributions

Ben yahia, Saafi, Ben Cheikh, and Bouker are the physicians directly involved in the study. Ben yahia, Guiga, and Saafi contributed to the study conception and design. Ben yahia and Saafi contributed to the redaction and interpretation of results. Ghannouchi N performed study supervision. All authors reviewed the results and approved the final version of the manuscript.

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Availability of Data and Materials

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