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Dilated Cardiomyopathy Revealing a Henoch-Schönlein Purpura in a Young Patient: About a Case and Review of the Literature…

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Abstract **Case Report**

Schönlein-Henoch purpura is a non-thrombocytopenic systemic vasculitis of small-caliber vessels due to immune complexes. We report the case of an adult 30 year old man admitted for a generalized edematous syndrome with purpuric lesions in the 2 lower limbs revealing a dilated cardiomyopathy whose etiological assessment was in favor of a Schönlein-Henoch purpura with cardiac, renal, dermatological involvement. The diagnosis was difficult to establish given the clinical polymorphism. Also, management was complicated given the multifocal involvement. The treatment was based on corticosteroid therapy and rituximab but the evolution was, unfortunately, not satisfactory.

Keywords: Schönlein-Henoch purpura, dilated cardiomyopathy, small-vessel vasculitis, immune complexes, adult purpura, immunoglobulin A vasculitis.

INTRODUCTION

Schönlein-Henoch purpura is a non-thrombocytopenic systemic vasculitis of small-caliber vessels due to immune complexes, especially immunoglobulin A (IgA). HSP in adults remains rare but more serious and its severity is potentially linked to cardiac and renal involvement. Its physiopathology remains unknown today but IgA would play a central role.

The etiology of HSP is also unknown. It can be triggered by any immunological attack: drug, infectious and even post-vaccination.

CASE PRESENTATION

Herein, We report the case of a young 30-year-old Moroccan patient, cannabis smoker and occasional alcoholic, admitted to the emergency department for a 3 weeks history of generalized edematous syndrome, arthralgia, epigastric pain, jaundice, worsening dyspnea in a context of declining general condition.

Our clinical examination found a conscious patient, hemodynamically stable with signs of global heart failure with abundant ascites, signs of arthritis next to the 2 ankles. The urine dipstick test showed hematuria (++)proteinuria(+++) without pyuria or bacteriuria, and autoantibodies were negative. This symptomatology was followed by the appearance of extensive infiltrated purpuric spots on both lower limbs complicated by necrosis and hemorrhagic signs (Figure 1). The TTE shows hypokinetic heart disease at the dilated stage with LVEF: 25% (SBP), bi- auricular dilatation, severe TR by ***dilatation of*** the ***tricuspid*** annulus (Figure 2). Laboratory test finds anemia at 8.1g/dl hypochromic microcytic with hyperferritinemia, the aspartate transaminase/alanine transaminase ratio was elevated to 98/67 IU/L, without renal insufficiency, CBEU found hematuria with dysmorphic red cells and the 24h proteinuria was persistently positive at 1g/24h.

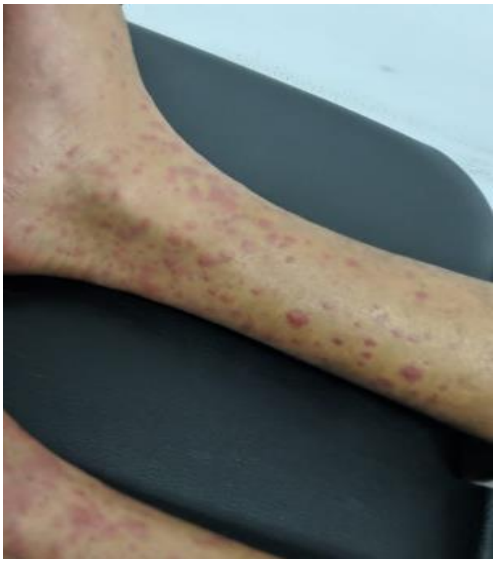

Figure 1: Confluent purpuric skin lesions

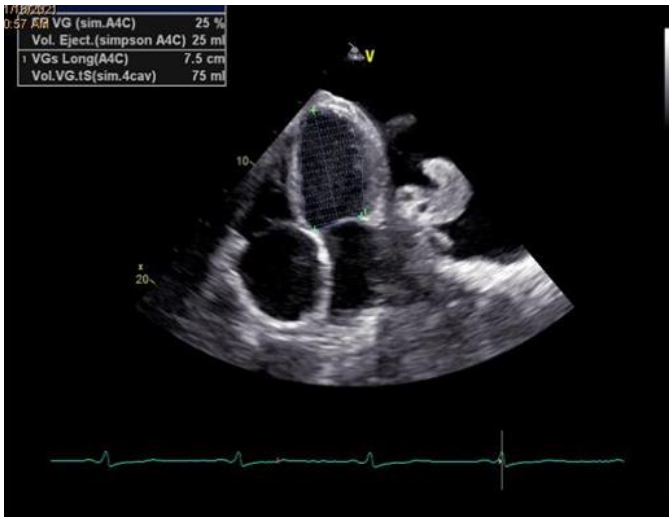

Figure 2: TTE showing severe LV dysfunction

Skin biopsy shows leukocytoclastic vasculitis with perivascular neutrophilic infiltrate and Immunofluorescence was positive for multifocal IgA deposits along the walls of dermal vessels (Figure 3). Renal biopsy showed mesangial proliferative glomerulonephritis with IgA deposits at IF.

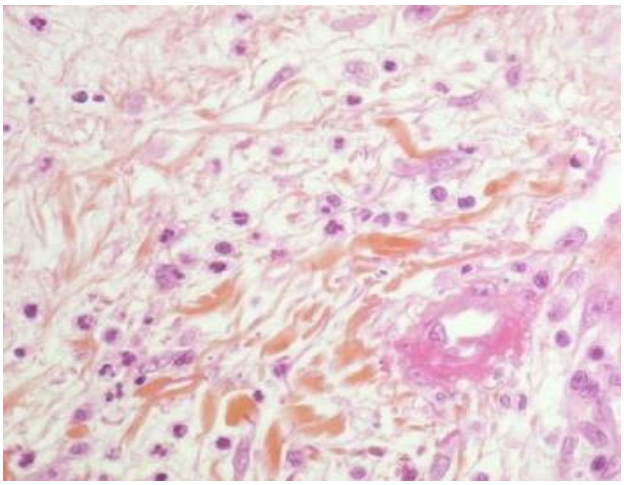

Figure 3: Anatomopathological study of the skin biopsy

The thoraco-abdominal CT angiography shows only an hepatosplenomegaly and an abundant ascites.

The diagnosis of HSP with cutaneous, articular, renal and cardiac involvement was retained.

The patient was put on treatment for heart failure and treated with high doses of methylprednisolone then with prednisone at 1 mg/kg/d with a good initial clinical evolution then a relapse mainly on the renal and cardiac level requiring the introduction of cyclophosphamide 1g/m²/ especially since biotherapy (such as rituximab) was not recommended in our patient given the seriousness of the cardiac damage that rituximab can also increase.

The subsequent course was marked by the worsening renal failure deterioration of the renal function and the installation of a cardio-renal syndrome, respiratory failure, without improvement of the LVEF. The patient died despite maximal supportive care.

DISCUSSION

HSP is a rare systemic small-vessel vasculitis characterized by the deposition of IgA complexes in the tissues. It generally has a self-limiting course and its manifestations are mainly cutaneous, gastrointestinal, renal, less frequently cardiac which makes the severity of the pathology [1].

The skin rash is the most frequent presentation [2, 3], it sits on the lower limbs and buttocks and can extend to the trunk and upper limbs. Purpura is often palpable, erythematous, and variable in size. Skin biopsy and histopathological analysis establish the diagnosis by objectifying leucocytoclastic vasculitis, and direct immunofluorescence shows an IgA deposit [4, 5].

The etiology of adult HSP is variable and poorly understood. Previous immunological reaction, such as bacterial or viral infection was suspected [2, 5]. A study by Albaramki published in 2016 showed that the majority of patients had a previous infection, especially a respiratory infection [2]. Incidents of purpura following infection with streptococcus, Mycoplasma, Epstein-Barr virus (EBV), parvovirus B19 have also been documented. Additionally, certain drugs and vaccines, have also been observed to trigger vascular reactions [6, 7].

The positive diagnosis is based on studies conducted by the European League Against Rheumatism (EULAR), the European Society of Pediatric Rheumatology (PRES) and the International Pediatric Rheumatology Trial Organization (PRINTO) in 2010. The category dictates the presence of purpura or petechiae, and the patient must have at least one of four other criteria: abdominal pain, histopathology, arthralgia and renal involvement. In addition to the EULAR/PRES/PRINTO clinical criterion, a definitive diagnosis can also be established by histopathological analysis demonstrating leukocytoclastic vasculitis and IgA deposition in the walls of blood vessels [8].

Cardiac involvement is extremely rare, and may be in the form of : acute or chronic myocarditis, pericarditis, atrioventricular block and even a real myocardial infarction [9].

Treatment of HSP is based on glucocorticoids, immunosuppressive agents and angiotensin receptor blockers/angiotensin converting enzyme inhibitors [10]. The effectiveness of treatment depends on the time between the onset of symptoms and the initiation of treatment, and the presence of comorbidities [10, 11].

Our young patient was treated with glucocorticoids and cyclophosphmide but the evolution was not satisfactory given the delay in seeking care.

CONCLUSION

This observation presents a rare case of rheumatoid purpura appearing in adulthood with multi-systemic involvement. This case underlines the great importance of an early diagnosis of this pathology in order to identify the disease and initiate the adequate treatment in a timely manner.

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