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Diagnostic Difficulties in A Case of Henoch-Schönlein Purpura at The Pediatric University Hospital Complex of Bangui.

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Abstract

Henoch-Schonlein purpura is an idiopathic IgA-associated systemic vasculitis characterized by the clinical tetrad of palpable purpura, arthralgias, abdominal pain, and renal dysfunction. It is a rare disease mainly seen in the pediatric setting. Its diagnosis is often problematic for clinicians in our environment.

We describe a clinical case of Henoch-Schonlein purpura suspected clinically in a 10-year-old adolescent who underwent laparotomy for intestinal intussusception. The objective was to analyze the diagnostic difficulties that may lead to the misrecognition of this pathology.

Keywords: Diagnostic difficulties, Henoch-Schönlein purpura, adolescent, Bangui.

Introduction

Henoch-Schönlein purpura (HSP) is a disease that results in the inflammation of small blood vessels (capillaries). It was described in 1837 by Doctors Enoch and Schönlein, hence the name Enoch-Schönlein purpura **[1]**. It affects 10 to 20 children per 100,000 per year, 90% under 10 years old **[2]**. This inflammation usually affects the small blood vessels in the skin, intestines, and kidneys **[3]**. The diagnosis of HSP is clinically characterized by purpura that can be associated with abdominal pain and arthritis.

The causes of HSP remain unknown. Viruses and bacteria are thought to cause the disease, which often starts after an upper respiratory infection. However, cases of HSP have also been observed following the intake of medication, insect bites, exposure to colds, chemical toxins, or the ingestion of specific food allergens. HSP can be a secondary reaction to an infection or a very aggressive response of the child's immune system **[4]**. The short-term prognosis is good and depends essentially on the digestive attack.

This work aims to clarify that atypical presentations of HSP are frequent, complicating the diagnostic approach.

Clinical case

Patient Y, 10 years old and weighing 29 kg admitted to surgery for abdominal pain. We note in his history a prior consultation for osteoarticular pain of the upper and lower limbs associated with skin lesions evolving in a febrile context. The patient's clinical examination concluded with an acute abdomen requiring an exploratory laparotomy, the postoperative report of which mentioned acute intestinal intussusception. Two weeks after his surgery, there was a resurgence of osteoarticular pain in the limbs and the persistence of skin lesions. The clinical examination on D7 postoperative noted a preserved general condition; the blood pressure is 94/73 mm Hg. Clinically, we observe purpuric papules close to petechiae, which are palpable and symmetrical on the legs and arms with no joint swelling **(Figures 1 and 2)**.

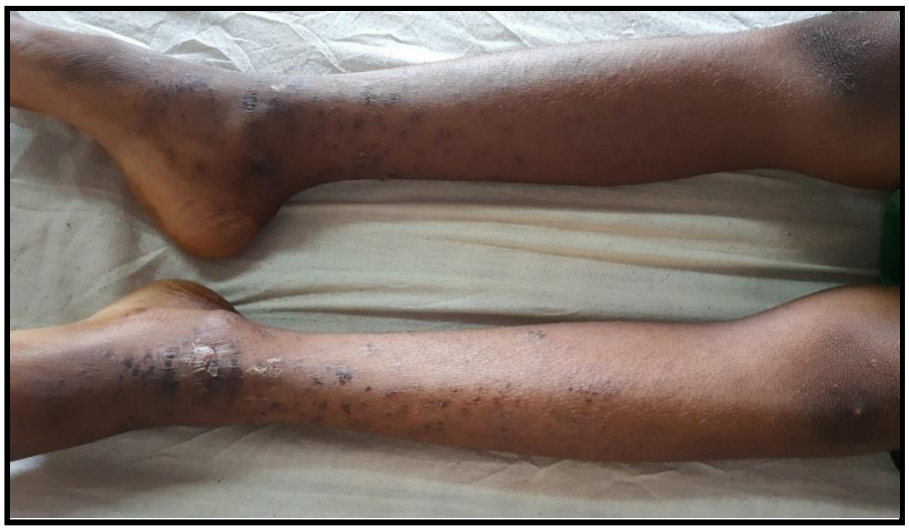

Figure 1 : Clinical picture of palpable involvement of purpura of the bilateral lower limbs (legs) by Petechiae and Purpuras.

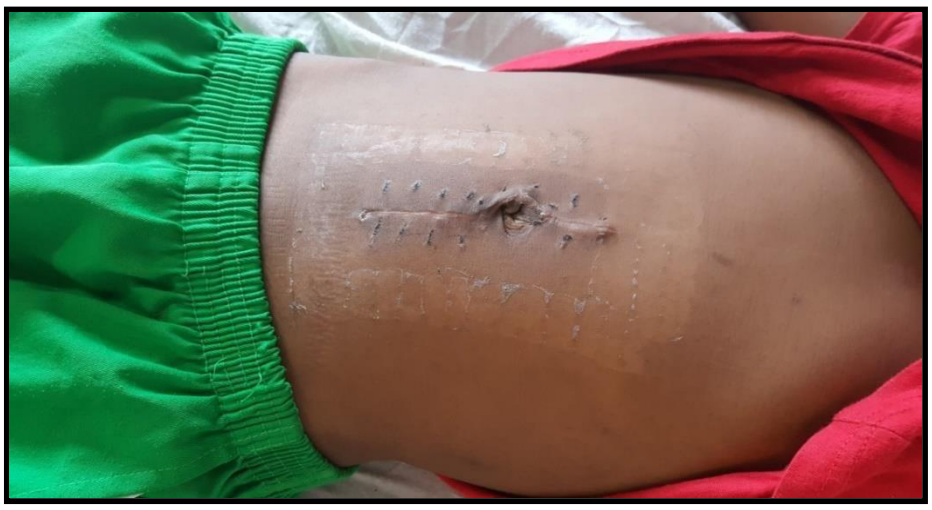

Figure 2: White line exploratory laparotomy scar.

On the paraclinical level, we note a moderate normochromic, normocytic anemia (Hb: 10.1g/dl), C-Reactive Protein (CRP) at 9 mg/l, serum creatinine at 4.1mg/l (creatinine clearance is at 108ml /min/1.73m2). Urinalysis showed no hematuria or proteinuria. A stool examination for occult blood was positive. Skin lesion biopsy and IgA assay were not performed.

The diagnosis of Henoch-Schonlein purpura was made using the criteria of the European Society of Pediatrics **(Table 1)**.

The patient was hospitalized and treated with Tramadol, then oral Ibuprofen on discharge. Regular monitoring of renal functions was done for six months, and the evolution to date is favorable, with no renal function disorder.

Table 1: Clinical manifestation of Henoch-Schönlein purpura adapted from the European Society of Pediatrics **[17].**

Organes	**Incidences**	**Présentations**
Peau	100 %	Purpura palpable, symétrique, sur la face d'extension des jambes, genoux et bras
Articulations	80 %	Oligoarthrite surtout des chevilles et des genoux
Tractus gastro-intestinal	65 %	Coliques abdominales post prandiales
		Hématémèse, méléna
		Nécrose, perforation
		Invagination, pancréatite
Rénal	35 % (20 à 60 %)	Micromacrohématurie
		Protéinurie
		Hypertension artérielle
		Syndrome néphrotique ou néphritique
		Insuffisance Rénale Aigue
Urogénital		Orchite, urétrite
Neurologique	2 %	Convulsion
		Hémorragie intracérébrale
Pulmonaire	< 1 %	Pneumonie interstitielle
		Hèmorragie alvéolaire

Discussion

Abdominal pain is a common reason for seeking emergency care. Atypical presentations of common diseases are frequent, further complicating the diagnostic process. Abdominal pain represents 5 to 10 % of the reasons for referral to Emergency Services **[5]**. They continue to pose diagnostic problems for emergency physicians. Despite the means of investigation available, the diagnosis remains imprecise for about 25 % of patients leaving the emergency department and 35 to 41 % of patients admitted to the hospital **[6]**. The associated symptoms often lack specificity, and atypical presentations of common diseases are frequent, further complicating the diagnostic process. In many cases, the range of differential diagnoses is wide. If abdominal pain is often the result of a benign pathology, this symptom can also reveal a pathology potentially life-threatening and requiring urgent therapeutic care.

Rheumatoid purpura is also called immunoglobulin A (IgA) vasculitis. This new nomenclature was retained at the Chapel Hill Revised Consensus Conference in 2012 **[7]**. IgA vasculitis has been reported in several locations over the past 25 years. The patient is 10 years old and male. Research conducted has revealed consistent annual incidence estimates of this condition ranging geographically from 6.2 to 70.3 per 100,000 in children under 17 with a slight male predominance (M: F = 1.2: 1.0). The peak age incidence is 4 to 6 years and 90 % of cases of Henoch-Schönlein purpura occur before the age of 10 years **[8]**. It is not easy to understand the pathophysiological mechanism of Rheumatoid Purpura. It would seem that an abnormal response of an immature immune system to an external antigenic attack is at the origin of this pathology. However, based on American and European criteria and epidemiological and clinical aspects, we can only confirm our case hypothesis as Henoch-Schönlein purpura. It is a systemic vasculitis of small vessels related to deposits of immune complexes containing immunoglobulins A. These complexes of antigens and antibodies, mainly IgA, are formed following bacterial and viral infections, vaccinations, drugs, and autoimmune mechanisms **[7]**. These antigen-antibody complexes deposit in small vessel walls and activate the alternative complement pathway, which leads to an accumulation of neutrophils resulting in inflammation and vasculitis without a granulomatous reaction. It can involve multiple systems, including the skin, gastrointestinal tract, kidneys, and joints, but it can affect any organ system. It is characterized by the association of cutaneous, joint, and gastrointestinal signs that can occur in successive outbreaks. Renal involvement may be associated with these manifestations and conditions in the long-term prognosis. This vasculitis is more common in children than in adults, where it is characterized by clinical polymorphism and particular severity **[9]**.

The diagnostic difficulty in this patient is based on the acute abdominal syndrome, which requires rapid management without exploration of the associated signs. The patient presented oligo-arthritis type osteoarticular pain in the knees and ankles with palpable purpura, symmetrical on the extensor face of the legs, arms, and knees. Henoch-Schönlein purpura is a clinical diagnosis. Cutaneous involvement is always present in the typical forms. These are noticeable purpuric lesions, which can sometimes coalesce into ecchymotic patches **[10]**. They predominate in the lower limbs but can also affect the elbows, forearms, and, more rarely, the trunk and the face. Digestive involvement mainly involves pain and vomiting; upper or lower gastrointestinal bleeding should lead to the search for a complication that determines the short-term prognosis. Digestive difficulties are primarily represented by digestive bleeding, acute intestinal intussusception, and perforation **[11]**. This patient's complication is characterized by intussusception that required immediate treatment without further questioning. Vascular deposition of immune complexes containing IgA1 plays a pathogenic role. Complement activation, cell damage, and IgA deposition suggest that HSP is a dysregulated IgA-mediated immune response to an antigen. However, even if their presence is an argument in favor of an HSP, it is not pathognomonic since IgA is found in other disorders such as Systemic Lupus Erythematosus (SLE), endocarditis, IgA nephropathy, and drug hypersensitivity reactions, among others. In addition, in HSP, it should be noted that the search for IgA in skin biopsies can be harmful in nearly 25 % of cases **[12]**. It would have been better to do this immunological analysis to at least rule out this possibility, but the technical platform did us harm.

Digestive ultrasound reveals an anteroposterior digestive mass in a "roundel" shapes 40 mm in diameter, reinforcing the hypothesis of acute intestinal intussusception, a non-exceptional complication.

The biological signs are non-specific and make it possible to eliminate other diagnoses: the inflammatory syndrome is absent or moderate.

We administered a symptomatic treatment, Tramadol, for a short time with Ibuprofen orally as a relay. On the digestive level, small doses of steroids have been shown to help reduce the intensity and duration of pain **[13]**. For this patient, we did not use corticosteroid therapy.

The hospitalization was of short duration, one week, and the evolution was favorable. In the literature, healing is spontaneous and occurs within a few days **[14]**. The follow-up was done for 6 months, and the patient did not present renal complications. According to the literature, the long-term prognosis depends on renal involvement **[15]**. Renal involvement manifests between the third day of diagnosis and the first year. Thus, 91 % of patients who develop kidney damage do so within the first six weeks, 97 % within the first six months, and 99 % in one year **[16]**.

Conclusion

Henoch-Schonlein purpura is a classic pediatric pathology whose presentation and diagnostic criteria are well-defined in the literature. Despite significant advances in research in recent years, the pathogenesis remains poorly understood. The precise evaluation of a child presenting this pathology will make it possible to direct care.

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