

Unusual face involvement in Henoch-Schönlein purpura

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ABSTRACT

Henoch-Schönlein Purpura is an idiopathic vasculitis characterized by deposits of immunoglobulin, mainly IgA, on the walls of small vessels, typically involving the skin, gut, joints, and renal glomeruli. Cutaneous involvement affects specially the lower limbs and buttocks, and it is seldom found on the face and upper limbs. We report the case of a 6-year old girl with purpuric lesions over the auricular, periorbital, malar, and mentonian regions and the upper limbs, arthralgia, and intestinal torsion. After surgical treatment and pulse therapy with glucocorticoids, her symptoms subsided without further complications.

Keywords: Henoch-Schönlein Purpura, skin, facial lesion, cutaneous involvement.

INTRODUCTION

Henoch-Schönlein Purpura (HSP) is the most common type of vasculitis in children 3 to 10 years old, and it is characterized by the development of purpuric lesions, mainly on the lower limbs and gluteal region, and it might be associated with abdominal pain, arthralgia, and nephritis.¹

Epidemiologic studies demonstrated greater predominance of boys.² In a Brazilian study, Silva *et al.* observed a higher incidence of this disorder in 6-year old girls, which contrasts with the literature.^{3,4}

This syndrome was initially described in 1801 by Heberden and, in 1837, Schönlein defined the association between cutaneous involvement, with purpuric lesions, and arthralgia. In 1874, Henoch described a syndrome of purpura, abdominal pain, and melena.⁵

We report a case of HSP with involvement of the lower and upper limbs and face, as well as intestinal torsion and arthralgia of the ankles, wrists, and proximal interphalangeal joints.

CASE REPORT

This is a 6-year old white female from Minas Gerais referred to the Hospital Monte Sinai of Juiz de Fora (MG, Brazil) on May 27, 2008. Three days before admission, the patient developed subcutaneous edema in the lumbar region and pain in the lower limbs, with worsening of the edema 24 hours later, along with the development of purpura on the lower and upper limbs and gluteal region, arthralgia of the left wrist, proximal interphalangeal joints, and right ankle. On admission, the patient was awake, her mucous membranes were pink, she was anicteric, acyanotic, afebrile, hydrated, and without abdominal and cardiovascular changes. She had purpuric lesions on the lower limbs, hands, face, and auricular region, as shown in Figure 1. Table 1 shows the laboratorial tests requested on admission and before discharge from the hospital.

Serum IgA levels were normal, 268 mg/dL (29 to 270 mg/dL), and urinalysis (UA) did not show abnormalities. Initially, the patient was treated with 20 mg of prednisone every 12

Received on 11/09/2008. Approved on 05/29/2009. We declare no conflict of interest.

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Figure 1. Purpuric lesions on face, predominantly on malar, maxillary, and mandibular regions.

hours, acetaminophen, anti-histamines, and omeprazole. Subcutaneous edema improved significantly; however, purpuric lesions spread, affecting also the face, and the patient developed arthralgia in the upper limbs, and abdominal pain.

Abdominal ultrasound demonstrated large amount of fluid in the abdominal cavity and reduced intestinal mobility; abdominal X-ray showed distension of intestinal loops. That same day, the abdominal pain increased and the patient developed nausea and vomiting. Intravenous hydration and antiemetics were instituted, and the diagnosis of Henoch-Schönlein purpura was made. The patient underwent surgery due to the suspicion of intestinal necrosis. Laparotomy did not show evidence of intestinal necrosis, only occlusive acute abdomen with intestinal torsion of the ileus, which was successfully reversed.

The patient was treated with pulse therapy with intravenous methylprednisolone (30 mg/kg/dose) for three days and, after surgery, she received parenteral nutrition and the intravenous association of metronidazole and ampicillin.

Biopsies of the appendix and skin of the arm and right leg showed, respectively, serous congestion and lymphoid hyperplasia of mucous membrane, edema of the dermis with extravasation of erythrocytes, ectasia of superficial and deep capillaries with perivascular deposit of fibrinous material and neutrophils permeating the vascular walls, interstitial infiltration of neutrophils, and collagen degeneration. This confirmed the presence of leukocytoclastic vasculitis, with more pronounced changes in the right arm skin sample.

After improvement of her symptoms, the patient was discharged on decreasing doses of prednisone, starting with 20 mg with slow decrements until discontinuation after 30 days.

DISCUSSION

The etiology of Henoch-Schönlein purpura (HSP) is unknown, although triggering factors, such as upper airways infections, and infections with group A streptococci, adenovirus, parvovirus B19, and mycoplasma, have been described.¹

Skin histology continues to be the most reliable method of diagnosing HSP.^{6,7} Deposits of IgA and C3 on direct immunofluorescence (DIF) are non-specific, and they seem to be observed more often in younger children. Although this test can lead to the diagnosis of HSP when associated with clinical symptoms, it is not always positive.⁸ Besides, the sensitivity of DIF is higher for deposits of IgA in the blood vessels if done within 48 hours of the onset of the disease because immunocomplexes are removed from the walls of the vessels

Table 1. Laboratorial tests on the day of admission and hospital discharge

Laboratorial tests	Admission	Hospital discharge	Reference values
Hemoglobin	14.5 g/dL	13 g/dL	11.5 a 16 g/dL
Hematocrit	42.2%	38.2%	36 a 47%
RBC	4.97 million/mm ³	4.21 million/mm ³	4 a 5.6 million/mm ³
ESR	62 mm/h	35.6 mm/h	Up to 10 mm/h
Platelets	365,000 mm ³	361,000 mm ³	140,000 a 450,000 mm ³
WBC	24,700 mm ³	16,200 mm ³	3,500 a 10,000 mm ³
CRP	37.95 mg/L	0.57 mg/dL	Up to 6 mg/dL
Creatinine	0.5 mg/dL	0.3 mg/dL	0.4 a 1.3 mg/dL

or destroyed after this period.⁹ In the case presented here, DIF was not performed because she was admitted to the hospital three days after the onset of symptoms, which decreases the efficacy of the test.

Symptoms of HSP are basically divided in four groups: skin lesions, affecting 100% of the cases; abdominal pain, nausea, and vomiting, affecting 65% of the cases; arthritis and arthralgia, in 70% of the cases; and kidney involvement, in 25% of the cases.¹⁰

As for skin manifestations, the classical lesion affects, predominantly, the gluteal region, knees, extremities of the lower limbs, and abdomen, and rarely affects upper limbs, trunk, face, and mucous membranes.¹¹

Face involvement is rare in HSP. Nussinovitch *et al.*¹², analyzing 155 children with this diagnosis, observed that only 4.5% had purpura or edema in the upper limbs and face. In the present report, besides classical lesions, the patient also had involvement of the upper limbs, especially extremities, and face, with purpuric lesions on auricular, periorbital, malar, and mentonian regions.

The reason for the low incidence of facial involvement is not known; some authors suggested that the location of the lesions depends on gravity, which would cause greater intravascular pressure in the lower limbs and gluteal region, therefore stimulating extravasation of plasma in those regions.¹²

Intestinal torsion and necrosis represent surgical complications affecting 0.7-13.6% of the cases and require fast and effective intervention to avoid the loss of intestinal segments.⁶

Burke Marks¹³ observed that patients with recurring episodes of vomiting develop facial purpura after a few days despite normal levels of hemoglobin, platelets, leukocytes, and prothrombin time. Those manifestations started to subside approximately 24 hours after the end of the episodes of vomiting. Vomiting or coughing can cause facial purpura due to an increase in intrathoracic pressure and consequent reduction in venous return.¹³ This would generate a sudden increase in local capillary pressure and plasma extravasation, forming purpuric lesions in the face in detriment of lower areas.¹⁴

Corticosteroids are recommended in more severe cases of HSP, especially with gastrointestinal manifestations, and it is more effective when instituted early.⁴

We conclude that, although rare, facial involvement should not be forgotten in cases of HSP.

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