

Joint disease and Henoch–Schönlein Purpura: a case review

Abstract

Schönlein-Henoch Purpura is the most common type of vasculitis in childhood. Its most frequent clinical manifestations are: palpable purpura, arthritis, abdominal pain, intestinal bleeding and nephritis although any organ may be affected. A large number of studies conclude that IgA plays a large role in its pathogenesis. It is more prevalent in males and the mean age of onset is at 6-7years. Joint symptoms are the second most frequent manifestation, after palpable purpura. Joint involvement occurs in 40-82% of patients, being more frequent in children under 7years. This may be the presenting symptom in 15-43%. Joint manifestations include arthritis and arthralgia. The affection is usually oligoarticular. Pain is migratory and recurrent. The most involved joints are knees, ankles, feet and hips. The diagnosis is clinical and the therapeutic management is conservative, with non steroidal anti-inflammatory drugs at an early stage, requiring only corticosteroids if pain is intractable. Arthralgia-arthritis is transient and resolves within a few days without consequences. The recurrence rate of joint involvement is very low.

Keywords: purpura, henoch-schönlein, arthritis, vasculitis

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Case report

Four-year-old girl was referred to the Emergency Department with a palpable purpuric exanthema in the lower limbs and the buttocks, pain and swelling in the left ankle (Figure 1) and on the second finger of the right hand. As antecedent, the previous week, she had acute pharyngitis.



Figure 1 Hematoma and photogenic signs in the left ankle.

Supplementary tests

- i. Microbiology:** negative rapid detection of streptococcus β -hemolytic group A, sterile urine culture, negative blood culture.
- ii. Hemogram:** 8320/mm³ leukocytes, hemoglobin 13g/dl, and platelets 276 000/mm³, Erythrocyte Sedimentation Rate (ESR) in the first hour: 18mm/h.
- iii. Coagulation tests:** Prothrombin time: 13.9seconds (range 10-14), Quick index: 69% (range 70-150), International Normalized Ratio (INR): 1.25 (range 0.85-1.15), Activated Partial Thromboplastin Time: 31.4 seconds (range 26-36), Fibrinogen: 429ng/ml (range 200-400).
- iv. Biochemistry:** Glucose 82mg/dl, Urea 20mg/dl, Creatinine 0.40mg/dl, Total/direct bilirubin 0.4/0.2mg/dl, Proteins 7.5g/dl, Albumin 3.5 g/dl, Total Calcium 8.6mg/dl, GOT/GPT 23/15

IU/L, GGT 15 IU/L, Lactate dehydrogenase (LDH) 254 IU/L, FA 127 IU/L.

v. Ions: Na 130mEq/l, K 3.5mEq/l, Cl 103mEq/l.

vi. C-reactive protein 1.05mg/dl.

vii. Urine sediment: no proteinuria is seen, no significant hematuria.

Due to the clinical characteristics, the case suggests as first possibility to Henoch-Schönlein Purpura with joint involvement. The case evolved satisfactorily with oral non-steroidal anti-inflammatory and rest.

Discussion

The Henoch-Schönlein Purpura (HSP) is the most common type of vasculitis in childhood. 200years ago, William Heberden described the first case in a five-year-old boy with purpuric exanthema, macroscopic hematuria, abdominal pain, bloody stools and vomiting. In 1837, Johann Schönlein added the joint component and called this entity “rheumatic peliosis” or “Purple rubra” years later, Eduard Heinrich Henoch, student of Schönlein, completed his description.¹ The most common clinical manifestations of this entity are: palpable purpura, arthritis, abdominal pain, intestinal bleeding and nephritis, although any organ can be affected. A great number of studies, published between 1960 and 1970, established that IgA had a great role in the pathogenesis of HSP, deposits of IgA were found in the renal mesangium and dermal vessels, also elevated serum concentrations of IgA and circulating immune complexes were found.¹

From an epidemiological point of view, the prevalence is greater in the male sex, and the average age of appearance is between six and seven years old. The debut is more common in winter (33%), autumn (27.4%) and spring (28.3%) and less frequent in summer (11.3%). Skin involvement in the form of palpable purpura is almost universal (98.1%), being the joint symptoms the second more common manifestation.²

Few articles focus on addressing these joint manifestations. We will proceed to comment some of them. Joint involvement occurs

in 40-82% of the patients.¹⁻⁵ Articular symptoms were observed more often in children under seven years, being the first symptom of the disease in 15-43% of patients.^{1,2} 6.65% of the patients with joint involvement developed this one to three days after the appearance of the purpura, 27% do it 4-7 days later, and 6% do it at least one week later.⁶ It is not known exactly what mechanism underlies joint involvement, beyond vasculitis. It seems that synovitis occurs concomitantly, but this fact is not proven.⁶ Joint manifestations include arthritis and arthralgia. The affectation is usually oligoarticular. Characteristically the inflammation is periarticular, painful, without erythematous or heat, but with limitation to ambulation. The pain is migratory and recurrent. Although the upper limb can be affected, the most involved joints are the knees, ankles, feet and hips.^{1,6,7} The diagnosis is clinical, and the therapeutic management is conservative, with non-steroidal anti-inflammatory in an initial step.¹ Only those cases that present with tractable pain require treatment with corticoids,¹ without, to date, being protocolized in a uniform manner the administration dose of these nor the time of corticoterapia. The evolution is favorable in most cases. The usual thing is that arthralgia-arthritis is transitory, and resolves in a few days without leaving sequels. It is rarely invalidating.^{1,6} 33% of the Patients with PSH can have recurrences, especially skin and digestive. The recurrence rate of joint involvement is very low.^{7,8}

Conclusion

Joint involvement is one of the most frequent manifestations of PSH. Its evolution is benign, self-limited in time and usually yield satisfactorily with conservative measures. Only in those refractory cases is it necessary to refer to a specific unit of Pediatric Rheumatology.

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Conflict of interest

Author declares that there is no conflict of interest.

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