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Henoch Schonlein Purpura with Facial Palsy

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Authors' contributions

This work was carried out in collaboration between all authors. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Henoch–Schönlein purpura is an acute vasculitis syndrome. It is a self limited condition with characteristic skin, joint, gastrointestinal, renal, and neurological manifestations. It is more common in children and can be associated with fatal complications such as gastrointestinal bleeding and nephritis. Nervous system involvement in HSP is usually underestimated. Headaches, seizures and mental status changes are the most frequent neurologic symptoms. We experienced a nine years old boy diagnosed as HSP with palpable purpura and developed lower motor neuron left facial palsy four weeks from his diagnosis.

Keywords: Henoch–schönlein purpura; facial palsy; rash; vasculitis.

ABBREVIATIONS

C3: Complement 3; CNS: Central Nervous System; FMF: Familial Mediterranean Fever; HSP: Henoch–Schönlein Purpura; Ig: Immunoglobulin; LMNL: Lower Motor Neuron Lesion; MRI: Magnetic Resonant Image.

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1. INTRODUCTION

Henoch–Schönlein syndrome is the most common vasculitis disorder in children. The characteristic clinical features include: palpable purpura concentrated in dependent areas, arthralgia or arthritis, abdominal pain and glomerulonephritis. Headache is also common [1,2]. More rarely, Henoch–Schönlein patients present with signs of either CNS or peripheral nervous system dysfunction [3,4]. In Henoch–Schönlein syndrome peripheral facial palsy may result from vasculitic neuropathy. There is no specific diagnostic laboratory test to assess the markers of HSP. The peak age prevalence of HSP in children is 3-10 years, but the condition is also seen in adults [5].

2. CASE REPORT

A 9 years old boy presented with skin rash over the lower extremities and on his buttocks lasted for 2 weeks. The rash was associated with joint pain around the ankles and difficulty in walking. The boy was diagnosed as having Henoch–Schönlein purpura based on purpuric rash and joint pain. His laboratory results were within normal. He didn't require hospital admission. One week later, he was presented with difficulty in closing his left eye, disappearance of the left nasolabial fold and deviation of the mouth to the same side giving the picture of left lower motor neuron facial palsy. His purpuric rash and joint pain improved at that time. He didn't complain from headache, fever or urinary symptoms. On clinical examination he was vitally stable with blood pressure 124/84 mmHg (95% centile), fainting purpuric rash over lower extremities and left lower motor facial palsy. The rest of his neurological examination as regard the tone, power of upper and lower extremities and higher function were normal. There was no gastrointestinal or renal problems. His urine routine, IgA level and other laboratory tests were normal. Full neurological assessment of the patient was done by pediatric neurologist who decided to start him on prednisolone 0.5 mg per kg for two weeks then gradual tapering. Physiotherapy also was done. The patient was reevaluated after stopping the steroids showing a dramatic improvement.

3. DISCUSSION

HSP is a systemic vasculitis syndrome which affects small vessels (arterioles and venules) and presents a variety of clinical manifestations. The etiology is unknown, but it was clear that IgA plays a role in the immunopathogenesis of HSP. The diagnosis is confirmed by skin biopsy showing a leukocytoclastic vasculitis with neutrophil infiltration. Granular deposits of IgA with little amounts of C3 and fibrin were shown by immunofluorescent studies.

HSP is common in children with male dominance (1.5: 1) and greater in spring and autumn. Gastrointestinal bleeding does not respond to conservative management and transfusions with surgical interference may be required. Pulmonary and cardiac complications were reported and have been associated with a poor prognosis.

Lower motor neuron facial palsy sometimes occurs in children with severe arterial hypertension [6,7]. As a consequence, in Henoch–Schönlein purpura peripheral facial palsy may result from a vasculitic neuropathy and severe hypertension.

Vasculitides may cause inflammation in the vasa nervorum walls inducing critical ischaemia to nerves. However, lesions in some cases may be due to compression by haematoma or localized oedema [8].

In reviewing the literature, 2 cases were reported by Özcakar ZB et al. [6] presented with severe hypertension due to HSP nephritis and led to neurological manifestations such as headache, decreased level of consciousness and visual changes. MRI finding was mainly posterior subcortical oedema. In both patients, controlling hypertension was followed by a full remission of the CNS dysfunction [7].

There are other neurologic manifestations reported in HSP including altered mental status, emotional instability, irritability, apathy, hyperactivity, tendency to sleep, seizures (partial, complex partial, generalized, or status epilepticus), and focal neurological deficits eg, aphasia, ataxia, chorea, cortical blindness, hemiparesis, paraparesis, or quadriparesis. Polyradiculoneuropathies as brachial plexus neuropathy or Guillain-Barré syndrome may result. Mononeuropathies e.g. facial nerve, femoral nerve, ulnar nerve or sciatic nerve may be also seen [9].

Table 1 shows the neurological manifestations reported in association with HSP (L. Garzoni et al.) [10].

Table 1. Neurologic manifestations reported in association with Schoenlein-henoch purpura

	No of cases reported (%) (N = 79*)
CNS involvement	
Headache	7 (8.9%) #
Mental status changes	
Behavioral	18 (23%)
Depressed state of consciousness	38 (48%)
Seizures	
Partial	9 (11%)
Partial and generalized	7 (8.9%)
Partial complex	2 (2.5%)
Generalized	35 (44%)
Status epilepticus	3 (3.8%)
Focal neurologic deficits	
Aphasia	6 (7.6%)
Hemiparesis	11 (14%)
Paraparesis	1 (1.2%)
Quadreplegia	1 (1.2%)
Cortical blindness	4 (5.1%)
Chorea	1 (1.2%)
Ataxia	2 (2.5%)
Peripheral nervous system	
Mononeuropathy	
Facial nerve	1 (1.2%)
Ulnar nerve	1 (1.2%)
Femoral nerve	1 (1.2%)
Sciatic nerve	2 (2.5%)
Peroneal nerve	1 (1.2%)
Polyneuropathy	
Guillian Barre	1 (1.2%)
Polyradiculoneuropathy	2 (2.5%)
Brachial plexopathy	1 (1.2%)

** No of case reports reviewed includes present cases.*
 # Incidence is probably underestimated

Although the long-term prognosis of Henoch–Schönlein purpura is mostly related to the kidney disease [1,2], some extrarenal features may produce substantial morbidity and mortality but they are rare. In Henoch–Schönlein purpura, a clinically relevant neurologic disease is very rare and mostly affects the patients with either a severe kidney disease or uncommon features. Two case studies [11,12], including 39 unselected children affected by HSP with normal neurological examination and blood pressure, headache was reported in 11 (28%). More importantly, transient abnormalities in the electroencephalogram including focal or generalized slow wave activity, focal attenuation of the voltage activity, sharp waves, and sometimes paroxysmal activity occurred in 21 (55%) of the 39 cases, indicating that mild cerebral involvement is more common in Henoch–Schönlein purpura [13].

Although corticosteroids and cyclophosphamide are not advised for management of Henoch–Schönlein patients with a peripheral or a cranial neuropathy as these conditions mostly will have full spontaneous recovery [14], there are some studies which recommend the use of steroids for moderate to severe Bell's palsy due to its anti inflammatory effect within the first 72 h after the onset and also for 20 % of patients in whom the palsy progresses [15].

4. CONCLUSION

Signs of nervous system dysfunction are uncommon in Henoch–Schönlein syndrome, but clinically relevant. It may present with seizures, confusion or stroke. Peripheral mononeuropathy as facial palsy is another presentation which is not rare.

CONSENT

It is not applicable.

ETHICAL APPROVAL

It is not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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