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A REVIEW ON HENOCH-SCHÖNLEIN PURPURA

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ABSTRACT
Henoch-Schönlein purpura (HSP) is a leukocytoclastic vasculitis involving small vessels with the deposition of immune complexes containing IgA commonly affecting joints, gastrointestinal tract and kidneys. Although HSP is typically a disease of children, adult cases have been described. HSP can affect multiple organs with a characteristic purpuric rash present in all patients. Most cases resolve with symptomatic treatment, but serious complications can occur such as renal failure. Physicians should be well aware of the disease because the true incidence is probably underestimated.

KEYWORDS: Henoch-Schönlein purpura, Purpuric rash, renal disease.

INTRODUCTION

Henoch-Schönlein purpura (HSP) is a leukocytoclastic vasculitis involving small vessels with the deposition of immune complexes containing IgA commonly affecting joints, gastrointestinal tract and kidneys.[1,2] This was first reported by Heberden in 1801,[1] who described the association of macroscopic haematuria with a purpuric rash, colicky pain, blood stools and arthralgia.[3] In 1837 Schonlien described association of purpuria and joint pain and termed this as "peliosis rheumatic". In 1874 and 1899, Henoch added GIT and renal manifestations to that description respectively.[1]

The etiology is unclear but is associated with bacteria (streptococci, staphylococcus aureus, h. pylori), virus (hepatitis A, herpes simplex adenovirus, HIV, etc), vaccinations (pneumococcal, influenza, meningococcal, hepatitis B, etc), medications (quinolones, clarithromycin, acetaminophen, codein, etc), tumors (lymphoma, multiple myeloma, prostate cancer, etc), genetic (alpha-1 antitrypsin deficiency, HLA-B35, etc) and parasites like toxocara canis.[4,5] Some studies have suggested that, infection, mainly upper respiratory tract infection, triggers the bouts of HSP, which can leads to skin and urine abnormalities after few days of infection.[6]

HSP mainly occurs in children, about 100 times more than adults. It has been reported that 6 to 24 per 100 000 children younger than 17 years will develop HSP, depending on the ethnic background of the child. In Asia, the incidence is as high as approximately 70 cases/100 000 children per year, [7] high in males when compared to females.[8] HSP occurs predominantly in winter seasons of the year usually between June and December.

Pathophysiology

It has been reported that all patients with HSP have small molecular mass IgA1-containing circulating immune complexes. The IgA1 molecule has a hinge region containing up to six O-linked glycan chains consisting of N-acetylgalactosamine, usually with an attached β1, 3-linked galactose. The primary defect leading to the production of such abnormally glycosylated IgA1 is probably heritable. These aberrantly glycosylated IgA1 molecules have been shown to form immune complexes with IgG antibodies specific for galactose-deficient IgA1.[7] This formation results in an increased amount of IgA immune complexes in circulation. These antigen antibody complexes deposit in the small vessel walls and activate the alternate complement pathway which leads to neutrophil accumulation resulting in inflammation and vasculitis without a granulomatous reaction. This can involve multiple systems including skin, gastrointestinal tract, kidney, and joints but it can involve any organ system. Vasculitis causes extravasation of blood and its components into the interstitial spaces resulting in oedema and hemorrhage.[4] Researchers have found elevations in the serum levels of immunoglobulin (Ig)A, IgA-containing circulating immune complexes and IgA rheumatoid factors in patients with HSP. Therefore, it is generally believed that HSP is an immune complex mediated disease. Because HSP is frequently reported to follow respiratory tract infections, a variety of viral and bacterial pathogens have been implicated as triggers of the disease.[7]

Clinical manifestations

HSP is a systemic vasculitis with multi-organ involvement. It is characterised by a classic tetrad of non-thrombocytopenic palpable purpura, arthritis or arthralgias, gastrointestinal and renal involvement, and rarely, other systems (lungs, central nervous system, and genitourinary tract).[4]

Skin

Skin was the most frequently affected organ system and characterized by purpuric rash. In severe cases it can cause large areas of raised purple patches and skin ulcers.[6] Recurrence of purpura might be associated with severe renal involvement in 25% of children with HSP.[9] Subcutaneous oedema involving scalp, face, extremities and penoscrotal area are reported in some studies.[10]

Arthritis/ arthralgia

The frequency of joint involvement is known to be similar in adults and children/adolescents.[11] Joint involvement is usually oligoarticular with large joints of the lower extremities (knee, ankle, and hip) most commonly affected,[9,12] and upper extremities may be affected too. Symptoms may include pain, swelling and decreased range of movement.

Renal involvement

The renal disease, which occurs in about 40 to 50% of patients, often presents as mild glomerulonephritis,[13] which produces proteinuria, microscopic hematuria, and often red blood cell casts. Most of the HSP nephritis resolves rapidly, only 5% of cases progress to end stage renal disease.[13,4] Other less common clinical manifestations of HSP include cerebral vasculitis, scrotal or testicular haemorrhage, and interstitial pulmonary haemorrhage.[9]

Gastrointestinal (GI)

Abdominal pain is the most common symptom of gastrointestinal involvement, which is colicky on nature and gets worse with food. Other symptoms include nausea, vomiting, hematemesis, melena and hematochezia, are secondary to vasculitis involving the splanchnic circulation.[4] Intussusception is also a rare complication but important to exclude as delay in management can result in ischemic bowel.[12]

Other symptoms

Other symptoms include neurological symptoms like nonspecific headache followed by subtle encephalopathy with minimal changes in mental status, such as labile mood, apathy and hyperactivity. Subdura haematoma, subarachnoid haemorrhage, cerebellar haemorrhage, intraparenchymal bleeding and infarction have also been documented in case reports.

Pulmonary involvement in HSP is rare. The most common and severe manifestation is diffuse alveolar haemorrhage (DAH) but it may present as interstitial pneumonia or interstitial fibrosis. Orchitis is a relatively common finding in boys. Genital swelling may also be due to more generalised oedema secondary to hypoalbuminaemia from renal or gastrointestinal involvement.[9,12,14]

Diagnosis

The presence of palpable purpura with the other components of the tetrad makes the diagnosis straightforward. However, other systemic autoimmune diseases must be considered, such as other forms of vasculitis or juvenile rheumatoid arthritis.[13] HSP should remain in the differential diagnosis of any patient presenting with abdominal pain with or without a skin rash. It is important to consider other causes of non-blanching rash, viz, disseminated intravascular coagulation, other vasculitides (Cutaneous small vessel vasculitis, Systemic lupus erythematous), causes of thrombocytopenia, hypersplenism. Other investigation includes blood pressure monitoring and dipstick urinalysis. If the diagnosis is uncertain in the patients, complete blood count (CBC) to exclude thrombocytopenia and autoimmune cytopenias, electrolytes, urea and creatinine, if the urinalysis demonstrates proteinuria, hypertension, oliguria or fluid overload; coagulation studies, auto-antibodies, blood culture, renal tract ultrasound and early morning urine protein and creatinine ration should be monitored to exclude orthostatic proteinuria and quantify pathology preoteinuria in the child with dipstick positive for protein.[15,17]

Treatment

There is no specific treatment for HSP. Only symptomatic treatment is recommended for the patients with the main goal of relieving the symptoms such as joint pain, abdominal pain and swelling. The treatments modalities were divided into four groups: general treatment, glucocorticoid therapy, immune inhibitor treatment and anticoagulant therapy. The supportive therapy includes proper hydration and administration of simple analgesics, like, paracetamol, NSAIDS. Glucocorticoid therapy was used to improve abdominal pain and joint symptoms, in case of infection, the antibiotics were used accordingly. First choice glucocorticoids were, oral prednisone 30 mg once daily and I.V dexamethasone 5~10 mg daily infusion. Severe symptoms were treated with I.V hydrocortisone 100~200 mg daily for 3 to 5 days, which on improvement was changed to oral prednisone. Kidney patients were added with immunosuppressive agents (such as azathioprine 2 to 3mg/kg/d cyclophosphamide 3 mg/kg/d, or triptolide 20 mg tid).[15,16] The use of glucocorticosteroids (GCS) in HSP has been source of controversy for many years. While the suggested benefits of early GCS treatment have included shortened duration of abdominal pain, decreased risk of intussusceptions and decreased risk of surgical intervention, the quality of evidence is generally poor, having come from mostly from small studies or case reports. In clinical practice, short courses of GCS are being used in patients with severe abdominal pain,

usually with rapid symptomatic improvement.[9] HSP patients were not all treated with single drug regimen but mixed treatment was also used in some patients.

Prognosis

Although relapses are quite common they are mostly mild and self-limiting. HSP recurs in approximately one third of patients, typically within 4 months of the initial presentation. Recurrent purpura can be occasionally associated with joint complaints and episodes of gross haematuria although each subsequent episode is generally milder and shorter.(9) Usually the disease stops re-occurring as patients get older. However, adults can present with new HSP having never experienced it in their youth. As with all forms of inflammation, there may be complete resolution or there may be scars that form – on the skin or in the kidney. These are not repairable and are associated with some loss of function- this is why recurrent bouts of HSP can lead to progressive kidney damage and doctors will monitor for kidney function, presence and quantity of blood and protein in the urine. If there is a lot of protein leaking out, patient can be started with angiotensin converting enzyme inhibitors(ACE) inhibitor or an angiotensin receptor blocker (ARB), which lowers the blood pressure that are especially good at lowering protein leaks from the kidney and reducing the rate of decline in kidney function. In older patients kidney function may have been severely damaged and dialysis may be occasionally, to be initiated.[6]

CONCLUSION

HSP is the most common vasculitis of childhood, and the true incidence is probably underestimated because the number of cases is underreported. HSP can affect multiple organ systems, and the characteristic palpable purpuric rash is present in all patients. The use of isolated non-thrombocytopenic purpura as the only criterion to diagnose HSP in children might therefore lead to over diagnosis and unnecessary follow-up. Physicians should take into consideration the prognostic and therapeutic implications of their diagnosis to decide whether or not to perform a skin biopsy.

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