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Henoch-Schönlein Purpura in Adults in the Emergency Department

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ABSTRACT

A report of an unusual and easily missed diagnosis of Henoch-Schonlein purpura in an adult Caucasian male who presented in the emergency department. The patient had widespread rashes, positive urine dipstick (haematuria, proteinuria and ketonuria) and responded well to steroid and fluids treatment in the emergency department. He was admitted to the acute medical unit.

Key words: Henoch-Scholein Purpura, Palpable rash, Nephritis

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INTRODUCTION

Henoch-Schönlein purpura is an autoimmune complex mediated systemic vasculitis of small vessels that occur most commonly in children aged 2-14 years in 90% of cases and young adults.[1] The peak incidence is between 4 and 6 years of age (70 per 100,000 children), with a male to female ratio of 1.5-2.1.[2-4] In the United Kingdom, the annual incidence is 6-22 cases out of 100,000 population.[3,5] In adults the incidence is 1.3 per 100,000 population.[6] The incidence in the United States and Sweden is 14 cases out of 100,000 population,[7,8] similar to that seen in other North American countries. [2,9] In Norway, the prevalence is 3.3 cases per 100,000 population.[10] The syndrome is associated with streptococcal infection and certain drugs (e.g., penicillin, quinines, erythromycin) however, the actual aetiology remains unclear. It occurs most commonly during spring and fall.[7,11]

CASE REPORT

A 51 year old man was brought to the emergency department by the Ambulance crew on account of one day history of groin pain and worsening rash over the buttocks, abdomen, forearms and lower legs. There were also swellings of both hands and feet. The patient had seen his general practitioner a day before because of a 5 day history of groin ache, of which he arranged for a scrotal ultrasound, and administered diclofenac. Despite the anti-inflammatory medication, the pain became more intense with widely spreading rash. Henoch-Schönlein purpura was suspected and the patient was admitted into the medical unit after urine dipstick showed proteinuria, haematuria and ketonuria. An electrocardiogram performed showed ventricular ectopics. Chest X-ray was unremarkable. Blood investigations showed mild hyperglycaemia. He was commenced on intravenous fluids, hydrocortisone, and antibiotics.

RESULTS

Henoch-Schönlein purpura been uncommon in adults and can easily be missed because of its masquerading features. The clinical presentation often is preceded by an infection (i.e., herpes zoster, varicella, mycoplasma or streptococci), vaccinations, insect bite, neoplasms or drug administration. General malaise and rash usually develops early and evolves from morbilliform to symmetrical and palpable purpuric forms i.e., slightly raised "palpable" haemorrhagic skin lesions.[12,13] The legs and buttocks are affected most commonly (Figures 1 & 2). However, in adults more than in children the upper extremities are more commonly affected.[14] These presentations are non-specific and can be features of any viral illness or other small vessels diseases such as Churg-Strauss syndrome, Wegener's disease, and microscopic polyangitis.

These patients may erroneously be reassured that the illness is a self-limiting viral exanthema or may have been treated with antibiotics for suspected bacterial infection.

Diagnostic criteria of Henoch-Schönlein purpura[12]

The presence of two or more of the following criteria

- Age of disease onset (twenty years or younger)
- Palpable purpuric rash
- Acute abdominal pain and
- granulocytic infiltrate in the walls of arterioles or venules

In older adults the renal (nephritis) or gastrointestinal involvements tend to be more severe, prolonged in duration, and of poor prognosis, although the natural history is unclear.[14-16]

DISCUSSION

Henoch-Schönlein purpura can cause damage to the kidneys including haematuria, proteinuria, glomerulonephritis and acute renal injury. In adults, this damage may be severe leading to end stage kidney disease,[17] particularly in those whose renal biopsy show glomerular crescents.[18] In addition, the gastrointestinal symptoms can be severe in adults causing abdominal pain, vomiting, bloody diarrhoea, and in some cases small bowel obstruction[17,19] and non-occlusive mesenteric ischaemia.[20] A more recent 10-year observational study of 160 patients in South Korea shows that in adults

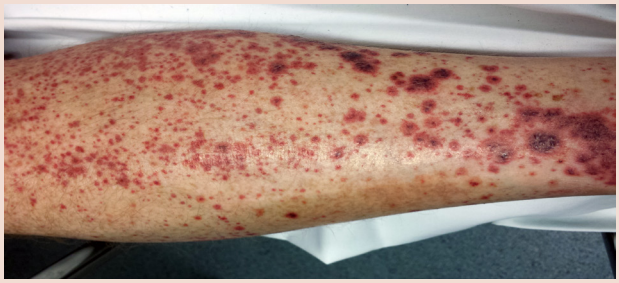

Figure 1: Palpable purpuric rashes on the anterolateral aspect of the leg.

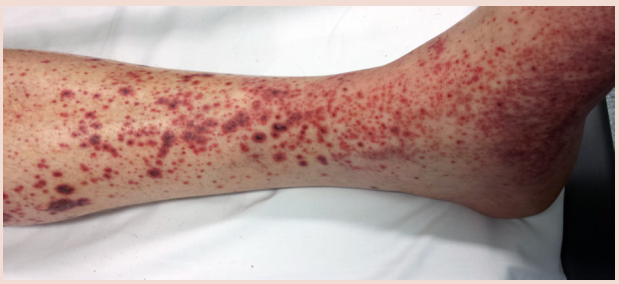

Figure 2: Palpable purpuric rashes on the anteromedial aspect of the leg, ankle and foot.

drug exposure and underlying malignancy (adenocarcinoma, lymphoma, and myeloma) were commoner in adults.[14] In the elderly population, affectations of the cardiovascular and respiratory systems although rare can occur, causing congestive heart failure, pleural effusion and pulmonary haemorrhage.[21]

There is need for early diagnosis particularly in older adults in whom diagnosis have wide range of management implications, and improve prognosis and reduce premature mortality. Delays in appropriate diagnosis may lead to unnecessary abdominal surgeries, bowel infarction and in some cases myocardial infarction. The mortality rate is in the range of 1-3percent.[22]

The diagnosis of Henoch-Schönlein purpura is based on high index of clinical suspicion. Henoch-Schönlein purpura is characterised by skin lesions (Figure 1) in form of palpable purpura (in patients without coagulopathy or thrombocytopaenia), abdominal pain from bowel lesions, arthralgia or arthritis, and renal disease. The skin lesions are often symmetrical and palpable and are usually found on the extensor surfaces of distal extremities (Figure 2) and buttocks.[13]

However, the presence of any two of the criteria identified by the American College of Rheumatologist in 1990 (Box) is diagnostic with a sensitivity and a specificity of 87.1% and 87.7% respectively.[12,23] The consensus criteria developed by the Paediatric Rheumatology European Society and the European League against Rheumatism was brought into clinical practice in 2005 for the paediatric age group.[24] Arthritis and/or arthralgia occur in 66% of patients[15] particularly the knees (38% of cases) and ankles (85% of cases).[25] Transient migratory but inflammatory arthritis is mild and non-deforming. Polyarticular arthralgia, gastrointestinal bleeding (bloody diarrhoea) and abdominal pain may dominate the presentation.[13] Renal disease occurs in 20-80% of the cases usually in the first one month of the disease, manifesting as haematuria and/or proteinuria. Bloody diarrhoea increases the chances of renal disease. In a small percentage of patients, initial renal impairment will progress from mild renal impairment and acute nephritic syndrome to end stage renal failure. Other organs may be affected. These include scrotum (scrotal swelling and torsion), central and peripheral nervous system (behavioural changes, seizures, headaches and mononeuropathies), eyes (visual impairment), cardiovascular system (congestive heart failure, myocardial infarction) and the respiratory tract.[21] Older adults who present with features suggestive of the criteria for identifying Henoch-Schönlein purpura should be referred urgently for evaluation. This should be differentiated from other conditions with similar symptoms such as acute abdomen, meningitis, idiopathic thrombocytopaenia, drug reactions, Wegner's granulomatosis and systemic lupus erythromatosus.

The presence of positive laboratory findings as well as clinical diagnostic criteria aid in the diagnosis. Anaemia may be present particularly in those patients with gastrointestinal bleeding. Leucocytosis and neutrophilia predominate, with elaboration of inflammatory markers. The absence of thrombocytosis is helpful diagnostically. Blood investigations may

show evidence of electrolyte derangement as a result of gastrointestinal haemorrhage, giving a positive stool guaiac test. Coagulation screen may be normal. Urinalysis invariably will show either a microscopic or macroscopic haematuria. Blood urea nitrogen and creatinine may be deranged. Abdominal pain can masquerade acute abdomen, however, abdominal ultrasound helps in the diagnosis. Endoscopy may show mucosal oedema and bleeding.[13] Angiography may be beneficial.

Skin or organ biopsies are the main diagnostic tool. Henoch-Schönlein purpura is confirmed by demonstrating immunoglobin A (IgA) deposition within and around blood vessel walls, i.e., invariable immune-fluorescent positivity for IgA in the mesangium.[26,27] Perivascular inflammatory lesions with serosanguinous leakage into the skin submucosa, and serosa are the hallmark of the disease.[12] In addition there tends to be leucocytoclastic infiltrates and intravascular fibrin deposits within arterioles, capillaries and venules. Clinical nephritis affect 30% of patients but almost all patients have an abnormal kidney biopsy. These kidney abnormalities range from segmental to focal mesangial changes with IgA deposits in the mesangium.[27]

Although no specific therapy is uniformly helpful, hospital admission is paramount. Most cases of Henoch-Schönlein purpura resolve spontaneously over a period of days to weeks following bed rest and supportive care. Non-steroidal anti-inflammatory drugs can abate arthralgia and other joint lesions. Evidence from randomised controlled trials reported a beneficial effect of steroid administration.[28,29] Intravenous corticosteroids alone are effective for gastrointestinal and joint involvements but nephritis usually requires treatment with both steroids and immunosuppression such as intravenous cyclophosphamide or azathioprine.[30,31] Prognosis is good except in 5-10% of patients who develop glomerulonephritis. Plasmapheresis treatment is controversial but may be useful in those with unusual features particularly renal and central nervous systems, in delaying the progression of the disease. Recurrence occurs in 50% of patients, while relapse occurs in 35% of patients.[11] Although, the outcome of renal involvement in the majority of patients is good, nephrologist referral is indicated in those with severe renal involvement as long-term follow-up is mandatory. In addition, surgical, gastrointestinal and dermatologist referral may be indicated based on manifestations.

CONCLUSION

The case report describes unique presentations of Henoch-Schönlein purpura in adults. Although it is uncommon in adults; consider the diagnosis in adults with a tetrad of palpable purpuric rashes, polyarthralgia, renal disease and abdominal pain. Treatment is usually symptomatic; however, admission is paramount. Clinical nephritis affect 30% of patients, therefore nephrology referral is indicated in those with severe renal involvement.

CONFLICT OF INTERESTS

I declare no potential financial conflict of interests in this research.

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ABBREVIATIONS USED

IgA: Immunoglobulin

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