HENOCCH SCHONLEIN PURPURA

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Introduction

During the months of April-May 1996, Indo Bhutan Friendship Hospital, (Indian Military Training Team) and the paediatric department of 'Jigme Dorji Wang Chuck National Referral Hospital, Thimpu Bhutan' diagnosed five patients with Henoch Schonlein; of them two were males and three females, ranging in age group 2 to 8 years. All these patients with variable presentation, were investigated systematically for achieving the final diagnosis and the salient features of these cases are enumerated below. Varied presentation of Henoch Schonlein purpura is highlighted. In 1895 Henoch was the first to draw attention to the important association of renal disease with the Syndrome. Osler pointed out the similarities of the condition to serum sickness, attributed it to an anaphylactic response to foreign antigens and coined the term 'anaphylactoid purpura' a term commonly used to refer to a group of non-thrombocytopenic purpura characterized by a septic inflammation of the vessels of the skin and other tissues [1] in association with other allergic features such as oedema and urticaria.

Case No. 1

A male child of 2 years presented with fever, purpuric lesions over both lower limbs and arthralgia involving bilateral knee joints. On examination he was febrile with palpable purpuric lesions over both lower limbs mainly on the extensor aspect. Both knee joints were tender and swollen. Laboratory investigations revealed Hb of 11.2g%, normal ESR and polymorphonuclear leucocytosis; platelet counts were normal. BT and CT were normal and ASO titre was normal. Entertaining a diagnosis of Henoch-Schonlein he was started on low dose aspirin for his arthritis. Arthritis subsided after 3 days, though new crops of skin lesions kept appearing for 2 weeks. The patient was asymptomatic on discharge.

Case No. 2

An 8-year-old female child presented with generalised oedema and haematuria of 1 week duration. Two months back she had symptoms of upper respiratory tract infection followed by reddish raised lesions over both lower limbs which subsided after 3 weeks. On general examination she had pallor and anasarca. There were

no skin lesions. Her blood pressure and systemic examination were normal. Investigations revealed low Hb (5.6 g%) bleeding parameters were normal. Total serum proteins decreased. Serum cholesterols and triglycerides were normal. Renal parameters normal. Urine microscopy revealed high RBC count and albumin 2+; USG kidneys normal.

ASO titre negative. Kidney biopsy done at CMC Vellore (where the patient was referred to for the same) revealed Mesangio proliferative glomerulonephritis. Electron Microscopy and Immunofluorescence showed mesangial IgA deposits. The patient received transfusion and steroids following which she improved.

Case No. 3

8-year-old boy referred from a district hospital with history of arthritis of both knee joints and cutaneous lesions over both lower limbs. Examination revealed symmetrically distributed palpable purpuric lesions over both lower limbs. All his investigations were normal, he responded to a short course of steroid therapy.

Case 4 and 5

Essentially presented with bilateral knee swelling, symmetrical raised purpuric rashes over both lower limbs. Both responded to aspirin.

Discussion

The Schonlein Henoch Syndrome is an acute condition, chiefly of children, in which a characteristic exanthem is associated with various combinations of painful periarticular swelling, GIT symptoms and haematuria. The skin lesions are an invariable feature. Renal involvement can be demonstrated at some time during the course of the disease in nearly 20-60% of the patients. In the majority this is asymptomatic and completely reversible but in 5 to 10% the picture of chronic or subacute glomerulonephritis develops [2,3]. Two-thirds of the patients have a history of upper respiratory infection preceding 1-3 weeks. Suspected though not proven inciting antigens include Group A β haemolytic streptococci, viruses, drugs [3], food [4] and insect bites.

The exanthem has been described by Gardner [5]. It is fairly symmetrical, usually urticarial at onset, later turning to a red maculo-papular rash which may become confluent before fading to a brown colour. New

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lesions may continue to appear for 2-4 weeks. The rash typically occurs in successive crops. Chiefly on the buttocks and extensor surfaces of lower legs and arms. These purpuric rashes are palpable. Subcutaneous oedema of scalp and eye lid, dorsum of hands and feet can be a striking feature [6] frequently painful.

The knees and ankles are joints most frequently affected. Intraarticular effusions seldom occur. Generally there is complete resolution and no residual deformity.

Acute colicky abdominal pain is common; serohaemorrhagic effusion into the bowel wall occurs. Nausea and vomiting may occur. Melena is seen in half of the patients and occult blood in stool is seen in 25% of patients. Sometimes intussusception [7] may endanger life.

Renal manifestation occur in 50 percent patients [8]. 90 percent of patients show urinary abnormality within 4 to 6 weeks of onset. Gross haematuria is seen in 20 percent of cases. Renal involvement is usually detected after the onset of skin and joint complaints. Moderate azotemia, hypertension, Oliguria and hypertensive encephalopathy may occur. Proteinuria with or without casts may be seen during first few weeks of illness. A nephrotic picture is seen in 10 percent children with Henoch's Purpura. Rare manifestations include CNS involvement with seizure, paresis and coma. Testicular swellings have been reported [9].

Laboratory tests are not diagnostic. There may be leucocytosis and ESR may be raised. Platelet counts and clotting studies are invariably normal. Immunoglobulin concentrations are normal with exception of Serum IgA, which may be elevated in 50 percent of these patients [10]. Immuno fluorescence studies of dermal vasculitis promise diagnostic clues. At present IgG, IgM, IgA and several components of complement have been demonstrated in and about the vessel. Renal biopsy shows mesangio-proliferative glomerulonephritis. IF and electron microscopy demonstrate deposits of IgA and IgM in glomerular mesangium. A rarer variety is IgM nephropathy which is often associated with nephrotic proteinuria [10].

There is no specific therapy. In rare instances when

the specific allergens can be proved, avoid the antigens. Salicylates will often alleviate symptoms of arthritis, fever, malaise. For GIT symptoms, prednisolone may bring dramatic response. Corticosteroids are indicated for the rare patients with CNS manifestations. Patients with systemic disease respond well to short course (3 months) of treatment with steroids [1].

The prognosis is excellent in the absence of significant renal disease. Recurrence of original symptoms is common for the first 3 months. 3-4 percent patients develop end-stage renal disease within a few months.

In our study, patients, ranged from 2-8 years. All presented with characteristic, skin lesions, 4 cases had arthritis, 2 cases, had abdominal pain and one had nephritis. Three of the patients responded to low dose aspirin and two to steroid therapy. Variable presentation of Henoch-Schonlein purpura with dramatic response to treatment is highlighted. Emphasis is made on the skin lesion as an invariable feature of the condition despite all types of varied presentation.

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