Henoch-Schönlein purpura and Hodgkin's disease

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Summary: A 39 year old man who presented with classical Henoch-Schönlein purpura was shown to have mediastinal nodular sclerosing Hodgkin's lymphoma. The Henoch-Schönlein purpura resolved after treatment of the lymphoma.

Introduction

Henoch-Schönlein purpura (HSP) as a presenting feature of carcinoma has been described.^{1–4} We report the first case of Hodgkin's lymphoma presenting with HSP and the association of the two diseases is discussed.

Case report

A previously healthy 39 year old single male civil servant presented to hospital with a one week history of abdominal pain, bloody diarrhoea, arthralgia and a progressive purpuric rash which started over the legs. He was not on any medication. Physical examination revealed a florid pleomorphic purpuric rash over the legs and buttocks. A single small node 1½ cm in diameter was noted in the left supraclavicular fossa. There was no hepatosplenomegaly. A clinical diagnosis of HSP was made.

Investigations revealed haemoglobin 13.2 g/dl, white blood count $21.4 \times 10^9/l$, (84% neutrophils), platelet count $513 \times 10^9/l$, ESR 40 mm/hour, serum albumin 27 g/l (normal 36–52) and proteinuria 1 g/24 hour; serum electrolytes, urea and creatinine and coagulation screen were normal. Hepatitis B surface antigen and antibody for human immunodeficiency virus and stool cultures were negative. Serum immunoglobulin levels were as follows: IgG 6.2 g/l (normal 7.0–19.0), IgA 2.6 g/l (normal 0.9–5.0) and IgM 0.58 g/l (normal 0.45–1.8). Screening test for circulating immune complexes was positive although they were not quantitated. Chest X-ray

showed a mediastinal mass, and computerized tomographic (CT) scan of the chest revealed fairly extensive mediastinal lymphadenopathy (Figure 1) extending from the level of the aortic arch to the root of the neck.

Although the node in the left supraclavicular fossa appeared innocuous at first, in view of the mediastinal mass the node was excised. Histology showed nodular sclerosing Hodgkin's disease. A skin biopsy was also performed and histology showed an established leucocytoclastic vasculitis with features of an anaphylactoid purpuric reaction in keeping with HSP. The patient was treated with chemotherapy and radiotherapy and there has been no recurrence of his HSP since he achieved remission from his Hodgkin's disease.

Figure 1. CT scan of the chest showing marked mediastinal lymphadenopathy (arrows).

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Accepted: 26 May 1988

Discussion

HSP is considered to be an IgA-mediated immune complex vasculitis⁵ which may involve the skin, joints, gastrointestinal tract and kidneys. Serum IgA levels are increased in about 50% of cases,⁶ although they were normal in our patient. Immunofluorescence studies⁶ on renal or skin biopsy characteristically show capillary granular deposits of IgA, IgG and complement C3. Although these studies were not performed in our patient, we felt that there was enough clinical and laboratory evidence to make a diagnosis of Henoch-Schönlein purpura.

Although no aetiological factor can be identified in the majority of cases of HSP, bacterial infection or drug ingestion have been documented in some patients. Moreover there are reports of HSP occurring in association with bronchial¹⁻³ and prostatic⁴ carcinomas. Recently, the case of a 63 year old man with non-Hodgkin's lymphoma and HSP was described.⁷ The patient, however, had a 30-year history of HSP with over 85 documented exacerbations before he eventually developed the lymphoma. There has also been a report⁸ describing two patients with Hodgkin's disease, who both presented with pruritic excoriations rather than purpura and skin biopsy in both cases showed

histological features of leucocytoclastic vasculitis. Although the skin manifestation of HSP is a form of leucocytoclastic vasculitis, the full clinical picture of HSP with skin, joints, gastrointestinal tract and renal involvement as a presenting feature of Hodgkin's disease has not been previously documented.

It is possible that in the case of HSP occurring in association with malignancy, the tumour itself may be the source of the antigenic stimulus leading to the immune complex reaction. Circulating immune complexes have been detected in patients with malignant conditions and tumour-specific antibodies and antigens have been isolated in these complexes.⁹⁻¹² We speculate that the Hodgkin's lymphoma in our patient triggered off the immunological reaction which led to the development of HSP. Tumour antigens such as Ki-1 and Leu-M1 have been described on the Reed-Sternberg cells of Hodgkin's disease.⁸ Although it would have been nice to isolate tumour-specific antibodies and antigens in circulating immune complexes in our patient, this investigation was not pursued.

Acknowledgement

We would like to thank Mrs S. Dodd for typing the manuscript.

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