

## Henoch–Schonlein purpura in mesothelioma

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### Keywords

Henoch–Schonlein purpura, mesothelioma, pleura, renal failure, vasculitis.

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### Abstract

Henoch–Schonlein purpura (HSP) is primarily a childhood immunoglobulin A (IgA)-mediated illness. When adults are affected, malignancy can be associated. We present a rare case of HSP in a 75-year-old man with malignant pleural mesothelioma. He presented with episodes of dizziness and subsequently developed non-palpable purpura across his legs, arthralgia, hematuria, proteinuria, and acute renal impairment. HSP was diagnosed based on clinical and histological findings on biopsy specimens from the skin and kidney that showed a leukocytoclastic vasculitis and mesangioproliferative glomerulonephritis with IgA deposits, respectively. He was treated with high-dose oral steroids with resolution of the skin and renal manifestations of the disease. HSP is rare in adults but has been linked to cancers. This is the first report of HSP in a patient with known malignant pleural mesothelioma.

## Introduction

Henoch–Schonlein purpura (HSP) is an immunoglobulin A (IgA)-mediated small-vessel vasculitis that can affect most organs, especially the kidneys, skin, joints, and gastrointestinal tract. It is primarily considered a childhood illness; the annual incidence in adults is as low as eight cases per million [1].

Malignant pleural mesothelioma (MPM) is an uncommon and universally fatal cancer with a median survival of 9 months. Australia has one of the highest incidences of MPM in the world.

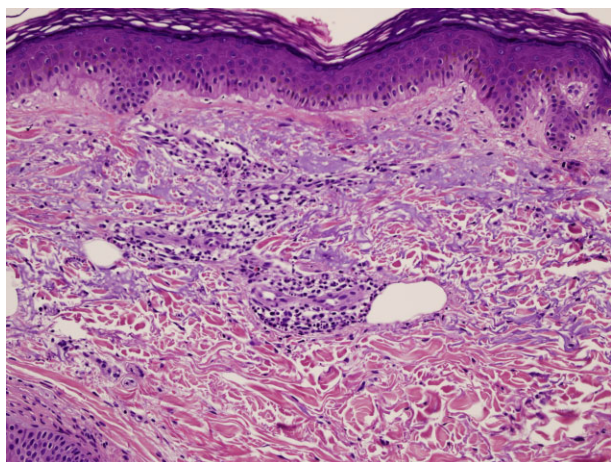
Although limited data exist, adult-onset HSP has been associated with various cancers, especially lung carcinoma [2]. The present study reports a rare case of HSP associated with MPM.

## Case Report

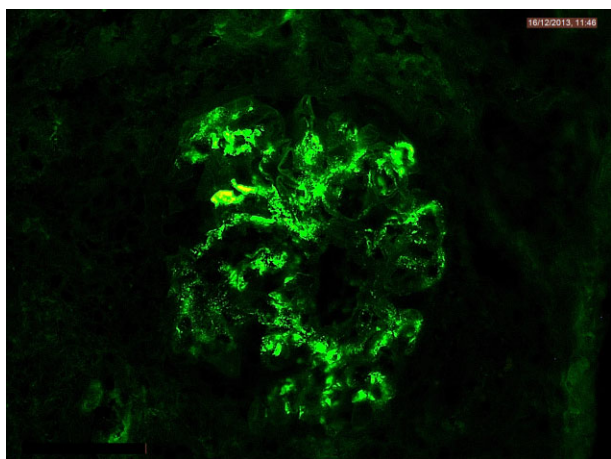
A 75-year-old Caucasian man presented with dizziness, a non-palpable petechial rash, and acute renal failure. He was

known to have a right-sided MPM diagnosed 10 months before but had never received chemo-irradiation. He was an architect with significant occupational asbestos exposure. He was a non-smoker with no other major comorbidity and no regular medications. His malignant effusion was initially treated with an indwelling pleural catheter, which was removed when his fluid production ceased. He had constitutional symptoms, diarrhea, and arthralgia but no abdominal pain.

Clinical examination showed signs of a right pleural effusion and thickening. He had an extensive, non-palpable, petechial rash covering both lower limbs. Peripheral blood showed a normocytic normochromic anemia (hemoglobin 83 g/L) and new onset renal impairment (creatinine 124 vs. 84  $\mu\text{mol/L}$  a fortnight prior). Spot urine analysis revealed proteinuria (protein-creatinine ratio of 474 mg/mmol) and hematuria (>100 erythrocytes per high-power field). He had a raised C-reactive protein (150 mg/L) and erythrocyte sedimentation rate (58 mm/h). The titers for antinuclear antibody titer (20 IU/mL) and antineutrophil cytoplasmic antibody (5 IU/mL) were raised. Tests for hepatitis B



**Figure 1.** Skin biopsy (x20): perivascular inflammatory infiltrate comprising neutrophils, lymphocytes, and histiocytes with moderate amount of leukocytoclastic debris.



**Figure 2.** Microphotograph of a histological section from the renal biopsy under immunofluorescence showing immunoglobulin A deposits in glomerulus (x40).

surface antigen, hepatitis C virus, and human immunodeficiency virus antibody were negative. Subsequent computed tomography scans showed signs of his known mesothelioma but no other cranial or abdominal (especially renal tract) abnormalities.

A biopsy of his skin rash (Fig. 1) revealed perivascular inflammation with leukocytoclastic debris. His renal biopsy showed mesangio-proliferative glomerulonephritis and, on immunofluorescence studies, strong granular IgA deposition in the mesangium of all glomeruli associated with moderate C3 and fibrinogen deposition (Fig. 2). This confirmed an IgA nephropathy, and a clinical diagnosis of HSP was made.

His rash subsided but the renal impairment and systemic inflammation failed to improve with conservative therapy after 3 weeks. Oral steroid (prednisolone 1 mg/kg/day) was initiated and gradually weaned over 4 months. His symptoms and renal impairment gradually resolved over 8 weeks. Urinalysis performed 3 months after presentation showed no proteinuria or hematuria.

The patient died 5 months after the initial presentation in a hospice from progression of mesothelioma without recurrence of HSP or renal impairment.

## Discussion

HSP, also known as IgA vasculitis, is characterized by a tetrad of clinical features: purpura, arthralgia, abdominal pain, and renal impairment. The pathophysiology involves deposition of IgA and complement components in small blood vessels and in the skin, joints, gastrointestinal tract, and kidneys, causing bleeding and other symptoms. Most (> 90%) cases occur in the pediatric population; however, adult patients often develop a more severe clinical syndrome and renal impairment [3]. The cause of HSP is unknown although bacterial infections, allergens, and drugs have been recognized as triggers. The course is generally self-limiting.

As many as 43% of adult patients with HSP have an underlying malignancy. Retrospective studies suggested that cancer patients >40 years of age are more likely to develop HSP, but no temporal correlations between the onset of HSP and the cancer diagnosis have been defined [2].

Although lung carcinoma is among the most common cancer type associated with HSP, mesothelioma has rarely been linked with HSP. One reported case described an 84-year-old patient with IgA-positive leukocytoclastic vasculitis who was subsequently diagnosed with MPM [4]. The onset of HSP preceded the mesothelioma diagnosis by 6 months. In contrast, our patient developed HSP 10 months after the diagnosis of MPM. Mitsui et al. suggested that HSP in patients with known malignancies might herald the occurrence of new metastases or tumor growth [2].

Renal manifestations of HSP are more frequent and more severe in adults [3]. Most patients with HSP receive only supportive therapy with analgesia and hydration. Steroid therapy may be beneficial in patients with advanced renal involvement [5].

In summary, we report the first case of HSP that developed during the course of MPM. Corticosteroid therapy was implemented with evidence of clinical and biochemical benefits.

## Disclosure Statements

No conflict of interest declared.

Appropriate written informed consent was obtained for publication of this case report and accompanying images.

## Acknowledgment

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