CASE REPORT

Pulmonary haemorrhage and pleural effusion in an elderly patient with Henoch-Schönlein purpura (IgA vasculitis): A case report

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Abstract

Henoch-Schönlein purpura (HSP) also known as IgA vasculitis is a systemic small vessel vasculitis mainly affecting the skin, kidneys, joints, and gastrointestinal tract. However, the disease can affect any organ system of the body. The classic tetrad of presentation in Henoch-Schönlein purpura includes palpable purpura, joints pain, abdominal pain sometimes associated with bleeding, and renal disease. Pulmonary and pleural involvement in HSP is a rare manifestation and occurs more commonly in adults than in children. We report the case of a 67-year-old woman with diabetes and HSP complicated by pulmonary haemorrhage and pleural effusion. She was managed initially with pulse Methylprednisolone and Cyclophosphamide followed by a course of oral steroids. An excellent outcome was achieved.

Keywords: Henoch-Schönlein purpura, IgA vasculitis, Pleural effusion, Pulmonary haemorrhage, Case Report.

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Introduction

Henoch-Schönlein purpura (HSP), also known as IgA vasculitis, is a small vessel vasculitis that can affect people of all ages but occurs most commonly in children under 10 years of age.[1] HSP is characterised by a non-thrombocytopenic purpuric rash, abdominal pain with or without gastrointestinal bleeding, arthralgia or arthritis, and nephritis. However, it can affect any organ system of the body.[2] Pulmonary, pleural, and nervous systems and scrotal involvement in HSP are rare manifestations and occur more commonly in adults than in children.[3-9] Diagnosis is largely clinical and determined by history and physical examination findings but when the presentation is atypical, tissue biopsy may be helpful. The revised criteria for HSP diagnosis were developed by the European League Against Rheumatism, the Paediatric Rheumatology International Trials Organisation, and the Paediatric Rheumatology European Society (EULAR/PRINTO/PRES) and were published in 2010 (Table 1). These are considered as gold standard for the diagnosis of HSP.[10]

Table-1: Diagnostic criteria for Henoch-Schönlein purpura as per EULAR/PRINTO/PRES.

Criterion	Description
Mandatory criterion	Purpuric or petechial rash with lower limb predominance
Minimum 1 out of 4 criteria	1. Diffuse abdominal pain with acute onset 2. Histopathology showing leukocytoclastic vasculitis or proliferative glomerulonephritis, with predominant immunoglobulin A (IgA) deposits 3. Arthritis or arthralgia of acute onset 4. Renal disease in the form of haematuria or proteinuria

EULAR/PRINTO/PRES (The European League Against Rheumatism, The Paediatric Rheumatology International Trials Organisation and the Paediatric Rheumatology European Society).

We present the case of a 67-year-old female with diabetes and HSP complicated by pulmonary haemorrhage and pleural effusion who was successfully managed.

Case report

A 67-year-old woman with diabetes presented to PAF Hospital, Islamabad, on June 13, 2024, with four-day history of severe colicky abdominal pain and one-day history of erythematous rash over her feet and legs, dyspnoea, and haemoptysis. There was no complaint of fever, melaena, haematochezia, haematemesis or haematuria. She had coronary artery bypass surgery in 2022 due to triple vessel coronary artery disease. Her regular medications included Aspirin 75mg, Bisoprolol 5mg, Sitagliptin 50mg, and Metformin 500mg, all once daily. On clinical examination she was afebrile; her blood pressure was 200/110mmHg. Respiratory rate was 26 breaths/min. Oxygen saturation was 85% at room air. There was an erythematous, palpable, and non-tender purpuric rash over both feet and legs (Figure 1). Abdominal examination showed mild, diffuse tenderness. Chest examination revealed coarse crackles

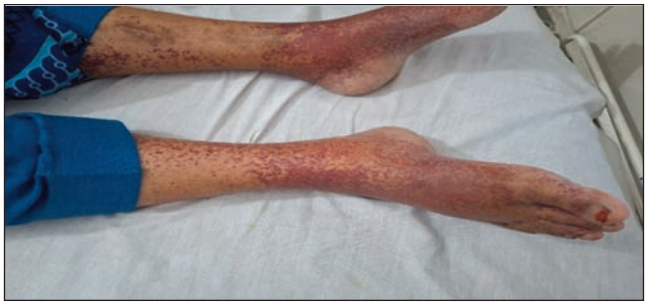

Figure-1: Palpable purpuric rash on lower limbs.

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Table-2: Results of relevant blood, urine and stool investigations.

Test	Result	Normal	Test	Result	Normal
Total leukocytes	7.6×10^9/L with 85% polymorph	$4\text{-}11\times10^9$/L	APTT	33 seconds	19-32 seconds
Haemoglobin	8.3 gm/dl	12-16 gm/dl	ANA test	Negative	Negative
Platelets	277×10^9/L	$150\text{-}350\times10^9$/L	Anti-ds DNA Ab	Negative	Negative
ESR	52 mm/hr	3-15 mm/hr	Anti-GBM Ab	Negative	Negative
CRP	56 mg/L	<5 mg/L	c-ANCA test	Negative	Negative
Serum urea	70 mg/dl	15-40 mg/dl	Complement C3	0.6 gm/L	0.81-1.57 gm/L
Serum creatinine	1.7 mg/dl	0.57-1.11 mg/dl	Complement C4	0.12gm/L	0.13-1.39 gm/L
Serum electrolytes	Normal	-	IgA level	5.5 gm/L	0.8-4.5 gm/L
INR	1.0	<1.1	Serum albumin	36 gm/L	35-50 gm/L
Serum ferritin, B12 & Folate levels	Normal	-	Urinalysis	Microscopic haematuria & +2 proteinuria	Negative for haematuria & proteinuria
Blood sugar random	187 mg/dl	<200 mg /dl	24 hour proteinuria	0.8 gm	<150mg
HbA1C	6.8%	<5.7%	Faecal occult blood	Positive	Negative
LFTs	Normal	-			

(ANA= antinuclear antibody; Anti-ds DNA Ab= anti double-stranded deoxyribonucleic acid antibody; Anti-GBM Ab= antiglomerular basement membrane antibody; APPT= activated partial thromboplastin time; c-ANCA= cytoplasmic anti neutrophil cytoplasmic antibody; CRP= C reactive protein; ESR= erythrocyte sedimentation rate; HbAIC= haemoglobin A1C [glycated haemoglobin]; IgA= immunoglobulin A; INR= international normalised ratio; LFTs= liver function tests;).

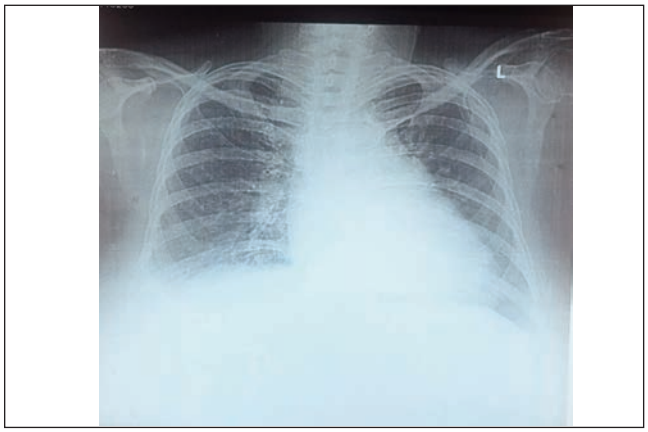

Figure-2: Chest X-ray showing blunting of right & left CP angles.

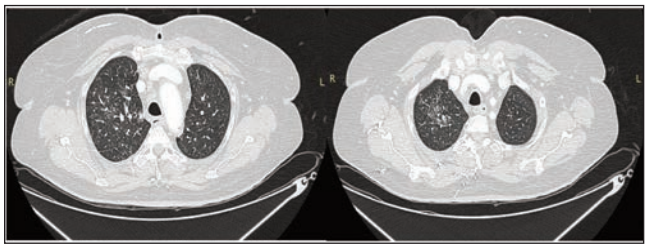

Figure-3: CT scan of the chest showing bilateral, multiple alveolar opacities and densities.

bilaterally and dull percussion note and absent breath sounds over the right lower chest.

Results of relevant blood and urine investigations are given in Table 2. X-ray of the chest (PA view) showed blunting of both the costophrenic angles (Figure 2). Ultrasonography of the chest revealed Grade-I hepatic steatosis and mild right- and left-sided pleural effusion. Computed tomography (CT) scan of the chest showed bilateral

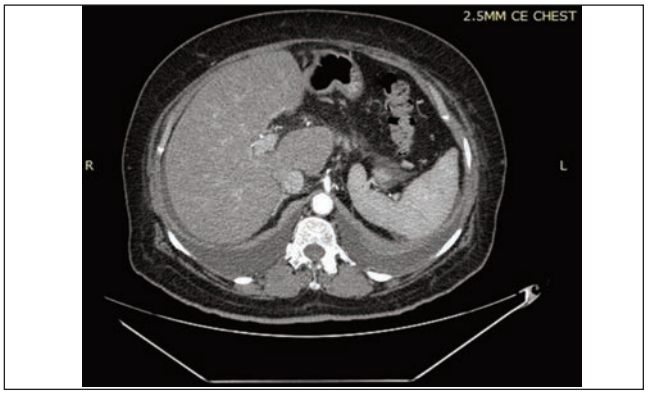

Figure-4: CT scan of the chest showing bilateral pleural effusion.

multifocal subtle alveolar opacities and densities suggestive of pulmonary haemorrhage (Figure 3). Bilateral mild pleural effusion (right more than left) was also noted (Figure 4). Stool samples were negative for various bacterial pathogens. Urine, sputum, and blood samples did not grow any micro-organisms (all cultures remained sterile). The stool for occult blood was, however, positive. Arterial blood gas analysis was relevant for type-1 respiratory failure. Skin biopsy showed leukocytoclastic vasculitis in post capillary venules with IgA deposition. A diagnostic pleural fluid aspiration was done under ultrasound guidance which showed haemorrhagic pleural fluid (erythrocyte count 3.2×10^9/L). Bronchoscopy with bronchoalveolar lavage (BAL) revealed a reddish coloured BAL fluid consistent with pulmonary haemorrhage. BAL fluid smear was negative for acid fast bacilli and cytology was negative for any malignant cells. The echocardiogram documented normal ventricular functions.

She was diagnosed as a case of Henoch-Schönlein purpura

with pulmonary haemorrhage and bilateral pleural effusion as per EULAR/PRINTO/PRES criteria.[11] The patient was initiated on intravenous pulse Methylprednisolone 1000mg daily plus Cyclophosphamide (500mg single dose) along with IV Omeprazole. Supplementary oxygen was given via face mask to keep oxygen saturation above 95%. Parenteral analgesics were administered as needed for abdominal pain. Injection Meropenem 500mg 08 hourly by intravenous infusion was also started to prevent any secondary infection. Blood pressure was managed with a combination of Amlodipine & Valsartan.

The patient showed good response to the treatment and improved clinically. Her abdominal pain settled down within three days. Within a week, her complaint of dyspnoea and haemoptysis subsided. There was no need of supplemental oxygen. Bilateral pleural effusion and pulmonary haemorrhage had been resolved. Serum urea and creatinine became normal with a decrease in the severity of proteinuria. However, microscopic haematuria persisted till the 10th day of post-corticosteroid treatment. Repeat stool examination for occult blood was negative. The intravenous pulse Methylprednisolone regimen was continued for three days followed by oral Prednisolone in a dose of 60 mg once daily with an intent to taper it off gradually over 12 weeks.

Discussion

This report describes the case of a 67-year-old woman with comorbidities, who had typical features of HSP and developed pulmonary haemorrhage along with bilateral pleural effusion. This is the first reported case of HSP from Pakistan in an elderly patient, complicated by pulmonary haemorrhage and pleural effusion. In HSP, classically, patients present with palpable purpura without thrombocytopenia or coagulopathy, arthralgia or arthritis, abdominal pain with or without gastrointestinal bleeding, and renal disease in the form of urinary abnormalities and/or raised serum creatinine levels. Purpura occurs in almost every patient and usually precedes other manifestations. Symptoms and signs may develop over days or weeks. In older children and adults other organ systems like lungs, heart, brain, and testes may less commonly be involved.[3,4,12] Pulmonary involvements due to vasculitis of alveolar capillaries are often overlooked. It can manifest with pulmonary haemorrhage (also termed diffuse alveolar haemorrhage), usual interstitial pneumonia, pulmonary infarction or interstitial fibrosis. Pleural effusion may be due to diffuse allergic vasculitis with an increase in the permeability of the pleural capillary network. Patients with HSP may present with pulmonary renal syndrome (co-occurrence of rapidly progressive glomerulonephritis and diffuse pulmonary haemorrhage) without typical manifestations such as purpura, abdominal pain, or arthralgia.[9,13]

This case of HSP is considered to be rare. The patient was an elderly lady of 67 years of age, whereas HSP mostly occurs in children below 10 years of age. According to EULAR/PRINTO/PRES, the current patient had the mandatory criterion, i.e. non-thrombocytopenic purpura with lower limb predominance. She had three out of four optional criteria, i.e. 1) diffuse abdominal pain with acute onset, 2) renal involvement, and 3) Histopathology of skin biopsy showing leukocytoclastic vasculitis. In HSP patients, skin rash precedes the development of other manifestations. The current patient presented with abdominal pain and skin lesions appeared later. CT of the chest showed bilateral multifocal subtle alveolar opacities and densities suggestive of pulmonary haemorrhage and bilateral mild pleural effusion. Bronchoscopy with bronchoalveolar lavage (BAL) was performed which showed an increasing reddish coloured return consistent with active pulmonary haemorrhage.

Differential diagnosis of this patient included other vasculitic diseases that can cause pulmonary haemorrhage and multi-organ involvement, e.g. systemic lupus erythematosus (SLE), Wegener's granulomatosis (granulomatosis with polyangiitis), microscopic polyangiitis, Churg-Strauss syndrome (eosinophilic granulomatosis with polyangiitis), antiglomerular basement membrane disease (Goodpasture's syndrome), and polyarteritis nodosa.[14] These alternative diagnoses were excluded on the basis of history, clinical examination, and investigations. The patient was negative for ANA and anti-ds DNA antibodies and did not fulfil the other diagnostic criteria of SLE. Wegener's granulomatosis can be difficult to differentiate from HSP early in the disease course. However, the current patient did not have any upper respiratory tract symptoms and signs and was negative for antineutrophil cytoplasmic antibodies (ANCA). Churg-Strauss syndrome was excluded by the absence of upper respiratory tract symptoms, late onset asthma, eosinophilia and negative test for ANCA. She was negative for anti-glomerular basement membrane antibodies making antiglomerular basement membrane disease highly unlikely. Her clinical picture was not consistent with Polyarteritis nodosa (PAN). Although there is no specific laboratory test that can confirm a diagnosis of PAN, histological examination of involved tissues can be helpful. In the current patient skin biopsy findings of leukocytoclastic vasculitis with IgA deposition in post capillary venules excluded this diagnosis. Several other conditions were considered unlikely, such as pulmonary infection because of absence of fever, cough with purulent

sputum, and leucocytosis; meningococcal sepsis because of the absence of pyrexia, leucocytosis, and neurological symptoms; thrombocytopenia, because the platelet count was normal; and coagulopathy because her coagulation profile was normal.

Patients with HSP are treated differently based on the severity of their illness and the presence of systemic involvement. In patients who have systemic involvement or life-threatening complications like pulmonary haemorrhage, corticosteroids alone or in combination with immunosuppressive drugs, Rituximab, Plasmapheresis or IV immunoglobulins are used.[15] The same treatment regimen can be used for pleural effusion if it occurs alone. In the current patient, treatment with corticosteroids in combination with Cyclophosphamide was effective in controlling the disease with excellent outcomes.

Conclusion

This case report aims to enhance our knowledge of rare manifestations of HSP in elderly patients. The patient demonstrated that HSP can present with pulmonary haemorrhage and pleural effusion. In the presence of uncommon complications, patients may be misdiagnosed with other diseases. Physicians need to maintain a heightened awareness of potential complications and atypical presentations of HSP. This will ensure timely administration of suitable therapeutic approaches whenever required.

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Author Contribution:

MT: Concept, data acquisition, analysis, interpretation, literature review, drafting, final approval and agreement to be accountable for all aspects of the work.

MZT: Data acquisition, analysis, literature review, critical revision, final approval and agreement to be accountable for all aspects of the work.

MZT: Concept, data collection, literature search, drafting, final approval and agreement to be accountable for all aspects of the work.

AS: Data collection, interpretation, critical revision, final approval and agreement to be accountable for all aspects of the work.