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Case Report

A case of Henoch-Schonlein purpura presenting with pulmonary involvement

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ABSTRACT

Pulmonary involvement in Henoch-Schonlein purpura (HSP) although rare is an important treatable cause of hospital admission in patients. Its diagnosis is difficult due to the vague symptoms and non-specific radiographic findings. We present one such case of a known case of HSP presenting with pulmonary involvement and treated successfully with steroids and immunosuppressant medications after ruling out all other causes. This particular case could also have an additional component of methotrexate induced lung involvement. Overall it is an interesting case with some much needed learning points.

Keywords: Henoch schonlein purpura, Methotrexate toxicity, Steroids

INTRODUCTION

Pulmonary involvement in Henoch-Schonlein purpura (HSP) is rare and occurs primarily in adults. The most common manifestation is Diffuse alveolar haemorrhage (DAH) and occasionally Usual interstitial pneumonia (UIP) or interstitial fibrosis (IF). Diffuse alveoloar haemorrhage (DAH) is associated with high mortality. Diffuse alveolar hemorrhage is a rare but serious and frequently life-threatening condition. Although it may occur in isolation without an identifiable underlying cause, more commonly, it is the result of inhalational injury or a complication of systemic vasculitis or connective tissue disorder. Most patients with DAH present with dyspnea, hemoptysis, and bilateral alveolar infiltrates on chest radiography. However, hemoptysis may be absent in up to 33% of patients with DAH. A low or decreasing hemoglobin level may be seen. Early recognition and prompt institution of supportive measures and immunosuppressive therapy are crucial for survival. Bronchoscopy with bronchoalveolar lavage should be performed early because it may confirm the diagnosis (progressively bloody return in the bronchoalveolar lavage fluid) and help to exclude infections before initiation of immunosuppressive therapy. In some cases, in which clinical features and serologic markers support a diagnosis of a specific underlying disease or when a biopsy can be performed less invasively from an alternate site (eg, a nasal biopsy in Wegener granulomatosis), a surgical lung biopsy may be unnecessary. Usual interstitial pneumonia refers to the histopathological pattern of architectural distortion, fibrosis often with honeycombing, and scattered fibroblastic foci, with a patchy distribution and predominantly peripheral involvement of the acinus or lobule. It is the underlying histopathological lesion of idiopathic pulmonary fibrosis (IPF). However, a pattern of fibrosis indistinguishable from that seen in IPF/UIP can occur in patients with asbestosis, collagen vascular diseases, drug-induced lung disease, chronic

hypersensitivity pneumonitis, and vasculitis. Thus, a UIP (usual interstitial pneumonia) pattern can result from varying causes and may involve different pathogenetic mechanisms in these respective contexts Aggressive supportive care, including immunosuppressive therapy and mechanical ventilation, when needed, appears to be beneficial in the treatment of patients who manifest DAH associated with HSP.[1] Henoch-Schonlein Purpura is not restricted to the pediatric population and has more severe nephritic and pulmonary involvement in adults. Pulmonary involvement may warrant treatment with high dose pulse steroids.[2]

The difficulty in recognizing methotrexate (MTX) lung toxicity also relates to the nonspecific symptoms voiced by patients, including progressive dry or productive cough and dyspnea, with or without fever. Because no single test can confirm a diagnosis of MTX-induced pneumonitis, investigations serve to rule out other possible etiologies. Chest radiographs (CXR) will reveal a diffuse interstitial pattern not consistent with typical bacterial pneumonias – although Pneumocystis jirovecii and other atypical pneunomias may produce a similar radiographic pattern. Computerised tomography (CT) will show characteristic ground-glass opacities with or without foci of consolidation.[3]

CASE REPORT

We present a case of a 74-year-old known diabetic and hypertensive male with Henoch Schonlein purpura presenting with mild non remitting fever, breathlessness, cough with expectoration along with constipation and bilateral legs having purpura. He was prescribed 10 weeks of 25 mgs of methotrexate once daily 6 months back and started on steroids a day before he presented with cough. Relevant examination findings included bilateral extensive rhonchi with crepitations and an oxygen saturation of 91% without oxygen support and tachypnea. He also had pedal edema.

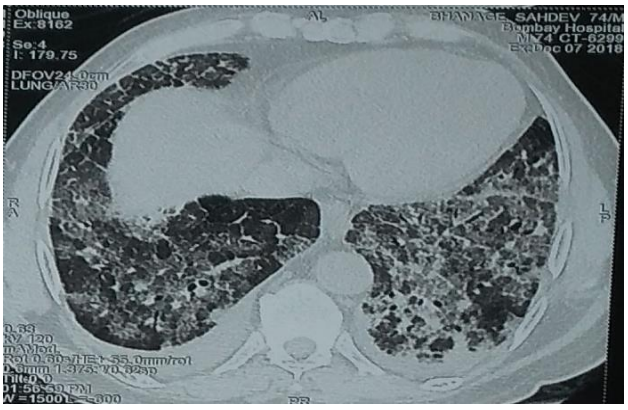

Figure 1: Ct scan image 1 of the patient

His routine investigations including hemogram, renal function tests, liver function tests and serum electrolytes were unremarkable apart from a mildly elevated leukocyte count. Ultrasonography of the abdomen and pelvis shows mild prostatomegaly and an 18.1 mm right renal calculi. Colonoscopy shows evidence of pancolitis. His c-reactive peptide (CRP) was also elevated. Sputum culture yielded methicillin resistant staphylococcus aureus (MRSA) and Candida species.

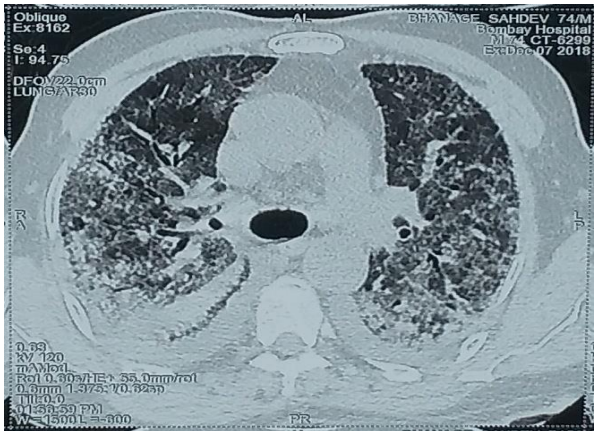

Figure 2: CT scan image 2 of the patient.

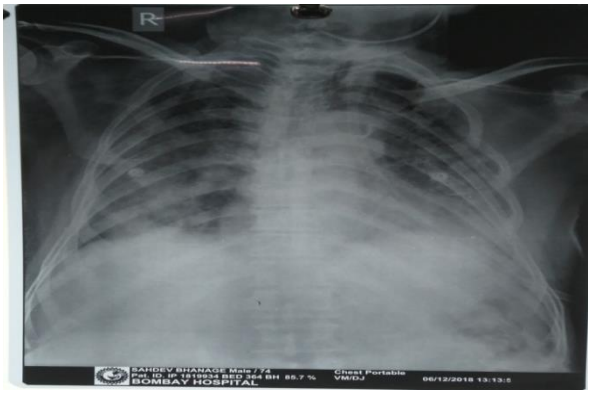

Figure 3: Chest radiograph of the patient.

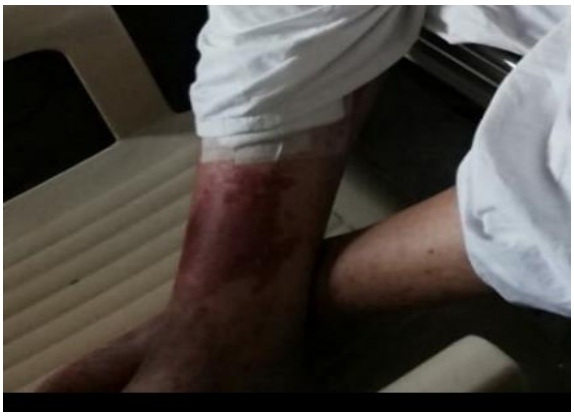

Figure 4: Skin lesions on the patient's leg.

Chest radiographs showed extensive patchy consolidation in bilateral lung fields. High resolution CT scan of the chest revealed bilateral pleural effusion, ground glass

attenuation and septal thickening. His transthoracic echocardiographic evaluation was normal.

Differential diagnosis that were considered at the stage were 1) lower respiratory tract infection 2) viral pneumonia 3) bacterial pneumonia 4) cardiogenic failure 5) autoimmune pneumonitis 6) primary colonic involvement: malignancy or inflammatory bowel disease 7) methotrexate induced lung involvement.

Treatment administered

He was treated with antibiotics, diuretics and non-invasive ventilatory support. He also required prolonged ICU admission. Bacterial infection was our primary differential, viral infection looking unlikely with an elevated leukocyte count, but however we were worried about worsening of the underlying HSP. Keeping that in mind, under antibiotic cover considering the local community antibiogram at first and later on the culture reports, steroids were continued right from admission. We started with oral prednisolone then switched to intravenous methylprednisolone when breathlessness was aggravated and when oxygen requirement was increased and then switched back to maintenance oral steroids. All the differentials were ruled out with relevant investigations including an echocardiography, 12 lead electrocardiogram, blood cultures, repeated sputum cultures, and various antigen and antibody tests.

Antimicrobial agents were adjusted according to the sensitivity pattern and pulsed steroids were also given for five days. It was a challenge to decide the time when antibiotics should be stopped but once clinical improvement ensued with an afebrile period of 72 hours we stopped antibiotics and later on steroid sparing and immunosuppressant drugs were added. He continued to improve on the immunosuppressant treatment and hence we deferred a lung biopsy or further invasive bronchoscopic studies. His diabetes and hypertension medications along with supportive treatment were continued. His oxygen support needed frequent titrations throughout the entire course in hospital. The patient was completely alright at the end of 3 weeks and was planned to be discharged. He was advised follow up at 1 week.

DISCUSSION

Nadrous et al studied 124 patients during a 6-year period at the Mayo clinic and identified 3 patients with HSP and pulmonary involvement not attributable to other causes.[1]

Setji et al reported another patient with HSP presenting with diffuse pulmonary involvement and that patient showed rapid pulmonary and renal improvement with IV high dose pulse steroids. He also stated that intravenous high dose methylprednisolone but not conventional doses of steroids has been shown to decrease the risk of persistent renal damage when given early in the disease.[2]

Cubero et al and Sim YS et al also reported a case of HSP presenting with diffuse alveolar involvement.[4,5] The mechanism of MTX-induced lung pathology remains unclear. Lung damage due to folate deficiency has been suggested but seems unlikely since MTX pneumonitis may occur after a single MTX dose and is not prevented by folinic acid treatment.

A hypersensitivity reaction is suggested by findings in lung biopsies: interstitial pneumonitis, granuloma formation and bronchiolitis, and in brochoalveolar lavage: lymphocytic alveolitis, increased eosinophils and reversed CD4/CD ratio, together with the clinical findings of fever, peripheral eosinophilia and response to corticosteroids.[7,8] The reports of spontaneous remission during MTX treatment and rechallenge of the drug without recurrence of lung pathology argue more for an idiosyncratic reaction than for hypersensitivity. A specific cellular immune reaction to the drug has been suggested by the production of a lymphokine which inhibits leukocyte migration [leukocyte inhibitor factor (LIF)] by peripheral blood lymphocytes after incubation with MTX. LIF production was observed in patients with MTX pneumonitis but not in other patients treated with MTX or healthy controls. A toxic drug reaction is suggested by the accumulation of MTX in lung tissue. the biopsy findings of alveolar and non-specific lung injury, and the resolution of pathology after stopping or lowering the drug. The fact that pulmonary pathology does not appear to be related to cumulative MTX dose argues, against this hypothesis. A great variety of chest x ray patterns have been described but bilateral interstitial (in 50% of the patients) or mixed interstitial and alveolar infiltrates (in 41% of the patients), most prominent at the lung basis are probably the most common. Unilateral infiltrates, a reticulonodular pattern and more rarely pleural effusions and transient hilar lymphadenopathy have also been reported. High-resolution computer tomography may show parenchymal ground-glass opacities (alveolitis), granulomas and fibrosis. Gallium and Tc-99 diethylenetriamine-pentacetate (DTPA) lung scintigraphy may show increased pulmonary uptake. These are sensitive but non-specific investigations in drug-induced lung disease and their value in the diagnosis and follow up of lung pathology related to MTX or other drugs has yet to be determined.[6]

Jakubovic et al reported that approximately 1% to 7% of patients receiving MTX treatment will develop pulmonary side effects. Prompt recognition of interstitial pneumonitis is essential before it progresses to irremediable pulmonary fibrosis. Part of the difficulty in recognizing MTX lung toxicity also relates to the nonspecific symptoms voiced by patients, including progressive dry or productive cough and dyspnea, with or without fever. Because no single test can confirm a diagnosis of MTX-induced pneumonitis, investigations serve to rule out other possible etiologies. CXRs will reveal a diffuse interstitial pattern not consistent with typical bacterial pneumonias–although Pneumocystis jirovecii and other atypical pneunomias may produce a similar radiographic pattern. CT will show

characteristic ground-glass opacities with or without foci of consolidation. Evidence of restrictive lung disease will be apparent in pulmonary function tests. Bronchoalveolar lavage findings are nonspecific and include increases in both CD4+ cell number and CD4/CD ratio.[8] Finally, lung biopsy (transbronchial and/or surgical) may also be performed and is generally indicated in instances of more severe or evolving respiratory disease in which cessation of MTX does not rapidly result in clinical improvement. Biopsy findings are also nonspecific, and may demonstrate evidence of acute pneumonitis with type II cell hyperplasia/dysplasia and interstitial infiltration.[3]

Arakawa et al reported that CT features of MTX-induced pulmonary injury were variable and included diffuse parenchymal opacification, reticular opacities, and centrilobular nodules. These opacities usually responded quickly to treatment; however, those patients with lung fibrosis at presentation may have worse prognosis.[7]

CONCLUSION

Henoch-Schonlein purpura is not restricted to the pediatric population. It has more severe nephritic and pulmonary involvement in adults. Pulmonary involvement with Henoch-Schonlein purpura may warrant treatment with high dose pulse steroids and later on steroid sparing agents however we need to be watchful for superimposed bacterial, viral and fungal infections. Clinical judgement is good enough to decide antibiotic duration as clinical markers can be confusing. Methotrexate (MTX) causes pneumonitis that is associated with a constellation of nonspecific findings so more often than not it can be considered a diagnosis of exclusion. A patients prescribed MTX should be advised of the potential for lung toxicity and to report the development of respiratory symptoms to their physician. This will enable timely investigations to be initiated to search for the underlying etiology and, if no alternative can be identified, MTX treatment discontinued.

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