

CONTINUING MEDICAL EDUCATION ΣΥΝΕΧΙΖΟΜΕΝΗ ΙΑΤΡΙΚΗ ΕΚΠΑΙΔΕΥΣΗ

Medical Imaging Quiz – Case 7

A 53-year-old patient, who had a history of asthma and drug abuse, had been treated for two months in the ICU for meningitis and acute respiratory failure. On transfer to a general ward he was examined and showed pulmonary infiltrates (fig. 1) and lower extremity flaccid paralysis with absent reflexes (brain CT and lumbar spine CT were normal). On blood count, WBC was elevated with eosinophilia (20%). The WBC had been elevated for the duration of the patient's stay in ICU with consecutive eosinophil counts of 8%, 12%, 22%, 32% and 30%. Consecutive

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radiographs showed patchy bilateral pulmonary infiltrates, mostly peripheral in distribution, which proved to be transient over a period of a few months (figures 2–4). Chest CT findings included

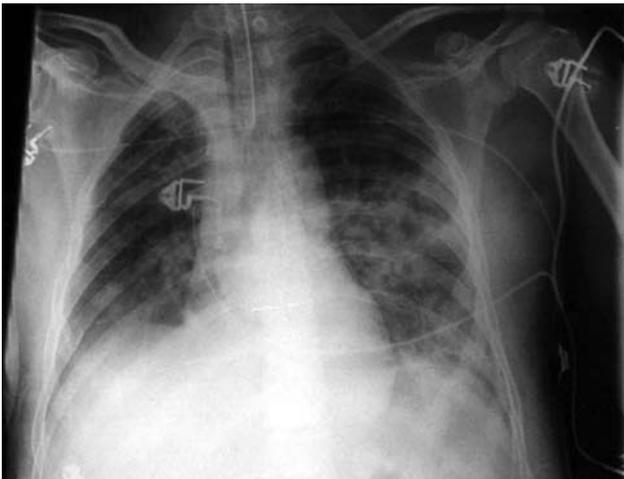


Figure 1. Chest x-ray on admission to the ICU has shown bilateral pulmonary infiltrates in the lower pulmonary zones mostly on the left.



Figure 3. Chest x-ray two months after admission has shown that the infiltrates are less extensive.

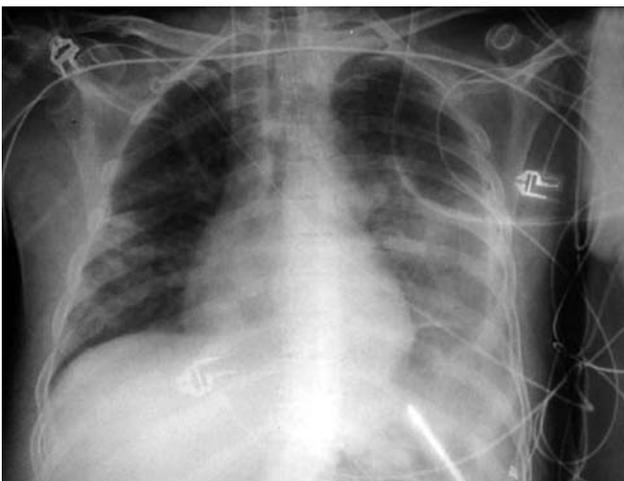


Figure 2. Chest x-ray one month later has shown that the lesions not only persist but appear to be more abundant.

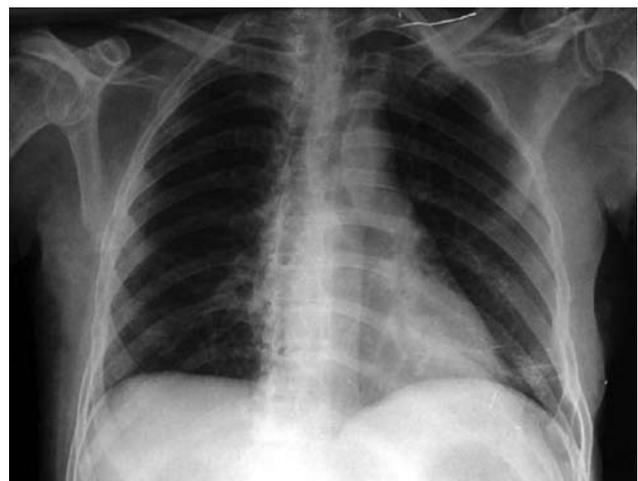


Figure 4. A few linear atelectasis can be seen, mostly on the left in the chest x-ray three months after admission.

bilateral lung infiltrates (figures 5–7).

Skin tissue biopsy revealed subacute dermatitis with vasculitis and the presence of granulomas, while skeletal muscle biopsy showed mild vasculitis of the small and medium-sized vessels. The final diagnosis, according to the American College of Rheumatology criteria, was Churg–Strauss syndrome. The patient was treated with corticosteroids and had a good outcome with as far as a 2-year remission period.

Comment

Churg–Strauss (CS) syndrome or allergic angiitis and granulomatosis affects mostly the middle age with men being affected twice more

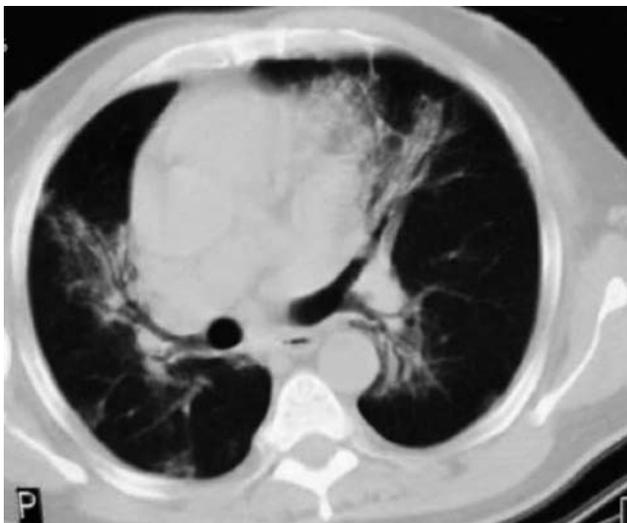


Figure 5. Chest CT two months after admission has shown that bilateral pulmonary infiltrates are present.

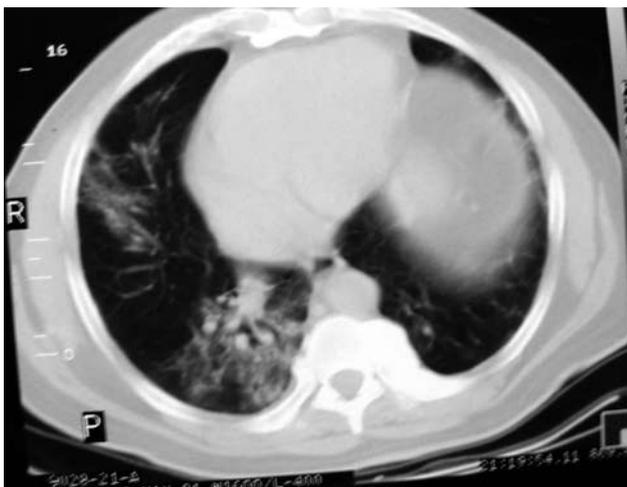


Figure 6. Pulmonary infiltrates are seen in chest CT two months after admission.

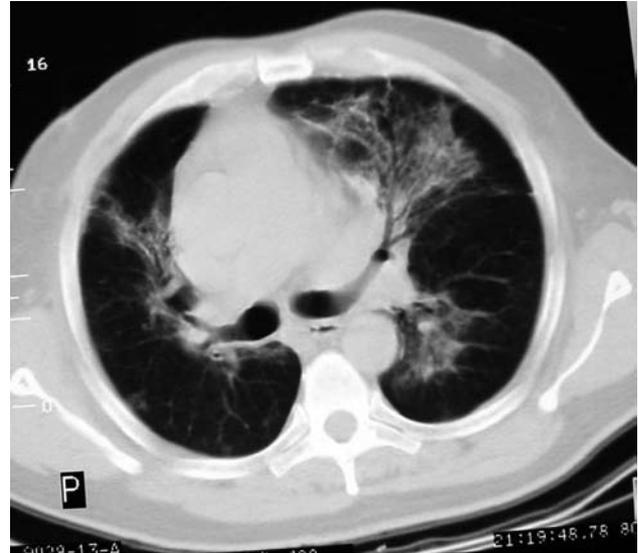


Figure 7. Pulmonary infiltrates are seen in chest CT two months after admission.

frequently than women. It is a form of angiitis, with extravascular or perivascular eosinophilic granuloma formation and parenchymal necrosis. It affects small arteries and veins and less common medium size vessels, which are infiltrated by eosinophils. Most commonly affected organs are the lungs, the heart, the gastrointestinal tract, the skin, the nervous system, joints and muscles.

The etiology is unknown. Asthma, eosinophilia and elevated levels of serum IgE suggest that there is an allergic or immune pathogenesis of the disease.^{1,2} Often there is a history of asthma and allergy which precede the angiitis phase. The onset of asthma is late compared to asthma in general population.

Clinically there are three distinct phases:

- *A prodromal phase that may persist for many years consisting of asthma often preceded by allergic rhinitis*
- *A second phase of marked peripheral blood eosinophilia and eosinophilic tissue infiltrates resembling Löffler syndrome, or chronic eosinophilic pneumonia which may recover over a period of years*
- *A third, life-threatening vasculitic phase. Common manifestations include fever, loss of weight, dyspnea, multiplex mononeuritis, myalgias, arthralgia, paranasal sinusitis, diarrhea, gastrointestinal bleeding.²*

According to Lanham et al, the diagnosis is based on the following criteria: Asthma, peak peripheral blood eosinophil counts of >1,5¹⁰/L and systemic vasculitis involving two or more extrapulmonary organs.³ According to the American College of Rheumatology, the diagnosis can be made with four or more of the six criteria: Asthma, eosinophilia >10% on differential WBC count, mononeuropathy or polyneuropathy, transient and fleeting pulmonary infiltrates on radiography, paranasal sinus abnormality and biopsy containing a blood vessel with extravascular eosinophils.⁴

The lungs are affected most commonly by transient diffuse or patchy pulmonary infiltrates. Less often the infiltrates are nodular.

Not so common is pleural effusion in thoracic involvement.

On chest radiographs the predominant finding is patchy multifocal peripheral consolidation while multiple nodular lesions with rare cavitation or diffuse interstitial pattern are less found.

On CT, findings include subpleural consolidation with lobular distribution, centrilobular perivascular densities with diffuse centrilobular nodules <5 mm in diameter often within ground glass lesions, or multiple larger nodules with rare cavitation. In the multiple larger nodules pattern the halo sign is quite frequent where the nodule appears to be within a ground glass opacity. The bronchial wall thickening with or without bronchial dilatation and hyperinflation is related to asthma while the interlobular septal thickening is related to interstitial pulmonary edema due to cardiac and pericardial involvement. Pleural effusion is not very common.^{2,5}

For these radiologic patterns with eosinophilia, differential diagnosis includes chronic eosinophilic pneumonia, acute eosinophilic pneumonia (Löffler syndrome), drug-induced eosinophilic pneumonia, allergic bronchopulmonary aspergillosis, hypereosinophilic syndrome, Wegener's granulomatosis.

Other clinical manifestations of the syndrome include: Cardiomyopathy leading to heart failure, acute pericarditis and constrictive pericarditis, gastrointestinal involvement with mesenteric vasculitis and submucosal granulomas leading to diarrhea, bleeding, abdominal pain and perforation, skin involvement with macular or papular erythematous rash, purpura, urticaria and subcutaneous nodules, renal involvement with focal segmental glomerular proliferation. Common manifestation is mononeuritis multiplex and less common sensomotor polyneuropathy while CNS involvement is rare.¹

Treatment of CS syndrome includes the use of corticosteroids, plasma exchanges and immunosuppressive agents (azathioprine,

cyclophosphamide) in variable combinations with corticosteroids being the basic treatment.¹

In patients with asthma, eosinophilia and CT or radiographs with transient diffuse parenchymal opacification or pulmonary nodules we should always keep in mind Churg-Strauss syndrome as a possible diagnosis.

References

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