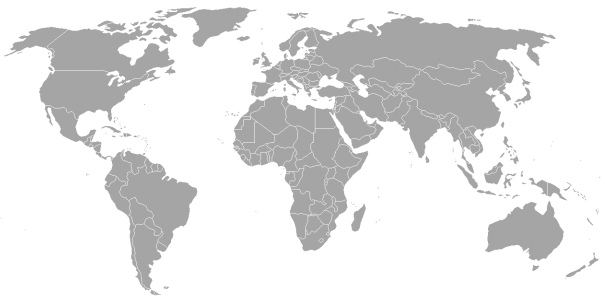
Inverse Bat Wing



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ABSTRACT

CEP accounts for up to 2% of all cases of interstitial lung diseases and seem more frequently in females. Symptoms are generally nonspecific. Wheezing, haemoptysis and respiratory failure are rarely seen. Extra-thoracic system involvement if present, excludes a diagnosis of CEP. Diagnosis is based on; respiratory symptoms of more than 2 weeks duration, alveolar (>40% on Broncho-alveolar lavage) and/or blood eosinophilia (0.1 X109/L), pulmonary infiltrates with usually a peripheral predominance on chest imaging and exclusion of other causes of eosinophilia. The chest X-ray as in the index case, described as photographic negative of pulmonary oedema is characteristic of CEP and is seen in up to one fourth of patients.

Key words: *Chronic Eosinophilic Pneumonia (CEP)-Interstitial Lung disease-idiopathic hyper-Eosinophilic syndrome-Corticosteroids-Broncho-alveolar Lavage*

30-year Caucasian female resident of Isle of Man presented with progressive worsening dyspnoea of 6 weeks duration. It was associated with intermittent fever not accompanied by chills and rigors. She also gave history of bilateral musculoskeletal chest pain since the onset of breathlessness. She denied history of cough, expectoration, haemoptysis, orthopnea, paroxysmal nocturnal dyspnea, palpitations, pedal oedema, joint pains, skin rashes, blueness of extremity, abdominal pain, muscle aches, decreased urine output, haematuria or weight loss. She was house wife with no addictions. She denied any recent change in furnishings of the house or change of home place or recent travel to Asia, Africa or Latin America. She owned a cat for past 5 years. There was no personal or family history of allergies, respiratory or autoimmune diseases. She was not on any medications including complementary and alternative drugs prior to onset of breathlessness. The patient was febrile with a temperature of 38.5 degree Celsius with a respiratory rate of 23 per minute, a blood pressure of 130/70 mm Hg, and a heart rate of 110/minute. General physical examination and systemic examination was unremarkable. Oxygen saturation was of 100% on room air. Investigations revealed haemoglobin of 15.6 gm/dl, White cell count of 15000/mm3 (N 78% L14% M1% E7%) with an absolute eosinophilic count of 1050/mm3, platelet count of 193,000/mm3 and.CRP of 18 IU. No blasts were seen on peripheral blood smear. Liver and renal functions were normal. Chest X-ray showed bilateral peripheral alveolar infiltrates having an `appearance (Figure 1). Bilateral patchy peripheral consolidations were seen on contrast enhanced computed tomogram of chest (Figure 2). Blood cultures were sterile. Serology for atypical organisms (Legionella, Chlamydia and Mycoplasma) was not contributory. No ova, cyst or parasite was detected on stool microscopy. Urine examination was normal. She tested negative for HIV, antinuclear antibodies (ANA) and anti-neutrophilic cytoplasmic antibodies (ANCA). Her serum IgE levels were normal and Aspergillus antibodies were not detected. No abnormality was seen on echocardiogram, nerve conduction velocities and. spirometry. Patient did not give consent for bronchoscopy which was planned. Peripheral alveolar infiltrates having the appearance of

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the photographic negative of pulmonary oedema (inverse of classical description of pulmonary oedema as batwing) characteristic of chronic eosinophilic pneumonia was seen on chest X ray.1 This was further confirmed by clinical presentation, CT Chest and blood eosinophilia. Eosinophilic pneumonia's are commonly seen with drugs (antibiotics , NSAIDS, antidepressants, contraceptives, leukotriene inhibitors, anticonvulsants, cocaine and many other drugs), foods, infection with parasites (strongyloidiasis, ascariasis, paragonimiasis, schistosomiasis, dirofilariasis, ancylostomiasis, trichomoniasis, clonorchiasis, and visceral larva migrans) and fungi (allergic broncho-pulmonary aspergillosis), asthma, autoimmune diseases (Churg Strauss disease) and malignancies (lymphomas, eosinophilic leukaemia's)2. Idiopathic syndromes known to cause eosinophilia include chronic eosinophilic pneumonia (CEP) and idiopathic hypereosinophilic syndrome (IHES).

CEP is twice as common in women as in men.3, 4, 5 The exact prevalence is difficult to determine but it is estimated that CEP is seen in up to 2.5% of cases of interstitial lung diseases.6 The triggering stimulus still remains elusive but by some undefined mechanism, the selective migration of Th2 cells to the lungs leads to chronic release of IL-5 and related cytokines in the alveolar space from Th2 cells, along with eotaxin, resulting in eosinophilic accumulation in the lungs. CEP is characterized by gradual onset of non-productive cough, fever, dyspnoea, fatigue, musculoskeletal pains and weight loss. Wheezing, night sweats, chest pain, and, occasionally, haemoptysis have been reported3, 5 Respiratory failures is occasionally reported3, 4, 5 Extra-thoracic system involvements is not seen in CEP. If extra-thoracic features are present, it is an indicator of other disease process for example IHEP, Churgh Strauss syndrome or rarely association of CEP with some other disease. Laboratory investigations show blood eosinophilia in excess of 1000/mm3. In the absence of significant blood eosinophilia, a diagnosis is supported by the demonstration of eosinophilia (≥ 40%) in broncho-alveolar lavage. Acute phase reactants, Erythrocyte sedimentation rate and C-reactive protein level are usually raised. Immunoglobulin E (IgE) levels are elevated in about half of the cases. Pulmonary function tests can be normal or show a restrictive/ obstructive pattern. Diffusion tests often show a reduced carbon monoxide transfer factor (DLCO). Hypoxemia is generally present on arterial blood gas analysis. Though histopathology remains the gold standard for diagnosis, it is not necessary for confirmation of diagnosis. When performed, lung biopsy shows an interstitial and alveolar inflammation with predominantly eosinophilic infiltration. Diagnosis is based on respiratory symptoms of usually more than 2 weeks duration, alveolar and/or blood eosinophilia, pulmonary infiltrates with usually a peripheral predominance on chest imaging, and exclusion of other causes of eosinophilia. Absence of involvement of any other organ system except respiratory system in presence of above features is again suggestive of CEP. In addition to above, a rapid response to steroids is considered as sine non queue of CEP.5,7 In contrast IHES is defined with absolute eosinophil counts more than 1500/mm3 pres-

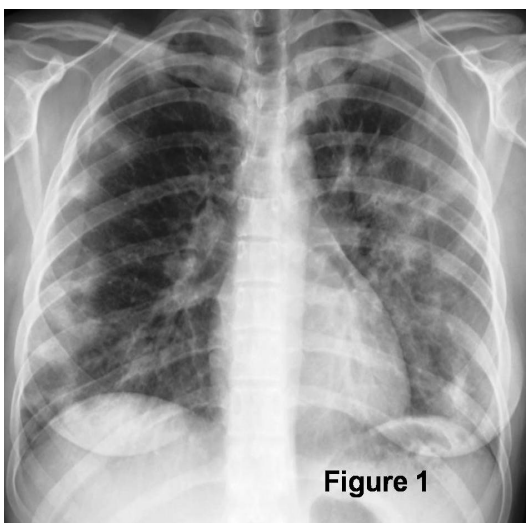

***Figure 1**. X-ray chest showing peripheral opacities described as photographic negative of pulmonary oedema*

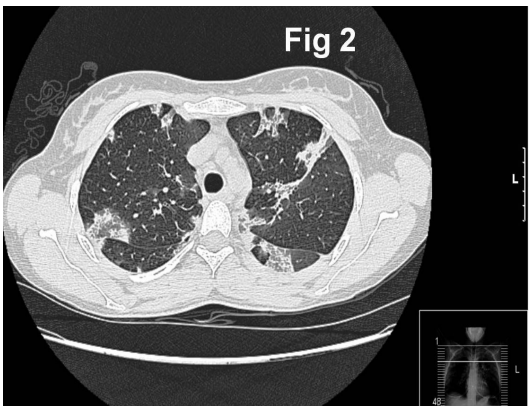

***Figure 2**. CT Chest showing peripheral consolidation*

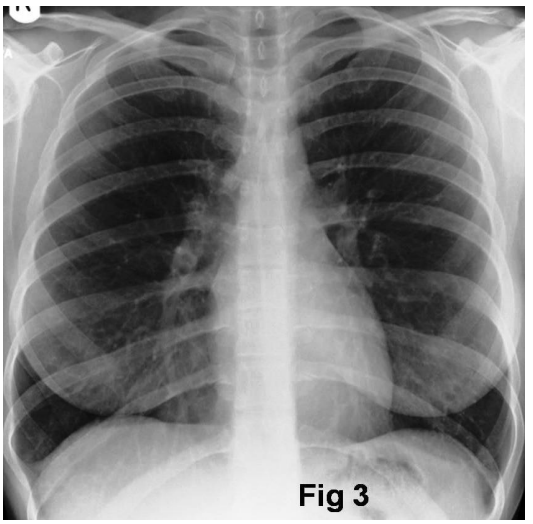

***Figure 3**. Chest X-ray showing complete clearance of opacities after treatment.*

ent for more than 6 months with involvement of other organ system (cardiac, peripheral nervous system, gastrointestinal and genitourinary system). Only one fourth of patients with CEP have the characteristic chest X ray described as photographic negative of pulmonary oedema as in the index case.1,5 Very unusually sarcoidosis, cryptogenic organising pneumonia and drug induced eosinophilia can present with similar radiology.5 CT chest demonstrates typical non-segmental areas of airspace consolidation with peripheral predominance6. The mainstay of treatment of CEP is corticosteroids. However the dose and duration still remain controversial based upon anecdotal reports and small series. Most authors however prefer to use 20-60 mg/day for the first few weeks, followed by maintenance of low dose (5-10 mg/day). Within hours of starting steroids patients start feeling better with a radiological clearance. This rapid response to steroids is diagnostic for chronic eosinophilic pneumonia. The duration of treatment is still controversial and most patients will require treatment for minimum of 6 month.1, 5 Though response to corticosteroid treatment is dramatic always leading to complete resolution, relapses are commonly seen in up to 50% of the cases both during and after treatment. These relapses respond to steroids and therefore some patients may require long term and even lifelong steroids. Inhaled corticosteroids have been proposed in order to prevent relapses and avoid systemic side effects of steroids. Up to one third of patients with CEP may develop asthma. Our patient was started on 40 mg of oral prednisolone for two weeks. She had a dramatic clinical and radiological response with clearing of opacities (figure 2). Prednisolone was tapered to maintenance dose of 10 mg and is currently doing well since past 4 months with no relapse.

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