Case 11384

Idiopathic Hypereosinophilic Syndrome manifest with Deep Vein Thrombosis and Pulmonary Embolism

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Section: Chest imaging
Area of Interest: Lung
Procedure: Diagnostic procedure
Imaging Technique: Digital radiography
Imaging Technique: CT
Imaging Technique: CT-Angiography
Special Focus: Pathology Chronic obstructive airways disease Embolism / Thrombosis Case Type: Clinical

Authors: Theodorou DJ, Theodorou SJ, Pallada S, Mantzoukis F, Tzimas T, Batsoulis A, Mastora M, Akritidis N.

Patient: 39 years, male

Clinical History:

Patient presented with abdominal pain and fever (38.2 °C). Medical history was insignificant. Laboratory tests showed elevated leukocyte count: 13,170/mm³ with 43% eosinophils, 0.1% basophils, and decreased platelet count: 55,000/mm³ [3]. Erythrocyte sedimentation rate and C-reactive protein level were elevated. On the second hospital day, he experienced left leg swelling.

Imaging Findings:

Chest radiograph showed bilateral patchy opacities, in middle and lower zones, with no clear pattern of distribution (Fig. 1). There was no pleural effusion. Axial chest CT scan revealed peripheral left upper lobe infiltrates (arrowheads) and right upper lobe ground-glass opacity (asterisk) tending towards consolidation (Fig. 2). Lower extremity venous ultrasonography revealed thrombosis of the common and superficial femoral veins as well as the popliteal vein. In addition, CT pulmonary angiogram revealed an intraluminal filling defect in the right main pulmonary artery, consistent with pulmonary thrombus (Fig. 3).

Discussion:

Idiopathic Hypereosinophilic Syndrome (IHS), a rare disorder, has been defined as persistent idiopathic eosinophilia greater than 1500 cells/mm³ [3] for more than 6 months or death within 6 months; absence of parasitic, allergic, or other known causes of eosinophilia; and signs or symptoms of organ damage related to eosinophilic infiltration [1]. Onset usually occurs in the third or fourth decade of life, with a male-female ratio of 7:1 [2]. Cardiac involvement including mural thrombosis, endocardial fibrosis, which may lead to restrictive cardiomyopathy is the major cause of morbidity and mortality in IHS [2]. Pulmonary involvement is observed in 40% of patients. Pleural effusion is seen in 50% of affected patients [3, 4]. Thromboembolic disease, peripheral neuropathy, and involvement of the GI tract, kidneys, joints, and skin have also been reported [1, 2]. Although biopsy is generally not
required for the diagnosis of IHS, histopathologic analysis demonstrates eosinophilic infiltration of involved organs with associated disruption of organ architecture and necrosis [2]. Radiographic findings in IHS are often non-specific and consist of focal or diffuse interstitial or alveolar non-lobar opacities, with most pulmonary opacities being related to severe cardiac failure, although lesions from the eosinophilia itself may be seen [1]. CT shows single or multiple nodules with or without surrounding ground-glass opacity and focal or diffuse areas of ground-glass opacity or interstitial infiltrates. Treatment of IHS is empirical based on systemic corticosteroids. Currently, there are no randomised controlled trials to establish dosing regimens. Our patient was treated with per os prednisone (25 mg four times a day) and subcutaneous heparin. Clinical improvement was rapid, with normalisation of body temperature within four days after initiation of treatment. Lung infiltrates subsided and he was discharged on the eighth hospital day. Prednisone dose was tapered down over the next eight weeks. There was no recurrence of symptoms during tapering of steroids, or upon discontinuation of therapy at a 2-year follow-up.

Differential Diagnosis List: Idiopathic Hypereosinophilic Syndrome, Infectious diseases (invasive pulmonary aspergillosis, Mucormycosis, Candidiasis, Wegener granulomatosis, Primary and metastatic hemorrhagic tumors, Bronchioloalveolar carcinoma, Pulmonary lymphoma

Final Diagnosis: Idiopathic Hypereosinophilic Syndrome

References:

**Figure 1**

Description: Chest radiograph shows bilateral patchy opacities, in middle and lower zones. Origin: GenHospIoannina
Description: Axial chest CT scan obtained the same day as the chest radiograph, shows peripheral infiltrates in the left upper lobe (arrowheads), and right upper lobe ground-glass opacity (asterisk) tending towards consolidation. Origin: GenHosp Ioannina
Figure 3

Description: Sequential CT image at the level of carina shows thrombosis of right pulmonary artery (arrow). Origin: GenHosploannina
Description: Transverse thin-section CT scan (mediastinal window) shows a filling defect in the right main pulmonary artery consistent with pulmonary thrombus (arrowheads). Origin: GenHospIoannina