A Case of Hypereosinophilia with a Fulminant Course Shortly After Starting Steroids Therapy

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Abstract: Hypereosinophilia is a challenging diagnostic dilemma that requires extensive work-up. Here, we report a case of an immunocompetent young man with a short and tragic course. He presented to us with chronic bouts of abdominal pain and Hypereosinophilia. Later, after starting steroids, he suffered from complications related to all major organs including multiple small bowel perforations resulting in an uncontrolled intraabdominal sepsis leading to his demise.

Key words: Hypereosinophilia, Hypereosinophilic syndrome, Intestinal perforation, Myopericarditis, CMV enteritis.

1. Introduction

Hypereosinophilia is a markedly increased in blood eosinophilia count, ≥1,500 cells/μL, whether discovered incidentally or found with signs and symptoms, commands diagnostic evaluation and often therapeutic interventions[1]. Hypereosinophilia irrespective of the cause can involve any organ, commonest being the skin, heart, lungs, GI, and nervous system. The mucosal surfaces of the gastrointestinal tract and the respiratory surface and the skin are rich with eosinophils, the reason behind that it protect against possible organisms invasion, as it has phagocytic activities against pathological organisms, but not as strong as neutrophils[2][3][4]. We report here a case of a young male with chronic abdominal complaints and hypereosinophilia that turned for the worse and involved all major organs soon after initiation of steroids therapy.

2. Case Presentation and Hospital Course

A 19-year-old male who is known to have bronchial asthma presented to us with history of recurrent diffuse colicky abdominal pain for four weeks, and constipation and vomiting of three days duration. He was in his third week post open appendectomy performed in another hospital for almost the same abdominal pain he is having without significant relief afterwards. He complained of anorexia and significant weight loss during the one-month period. He also reported history of fever which was not measured before without chills nor night sweats. There was no diarrhea, bleeding, nor jaundice. Also he denied any urinary or rheumatological complaints. He gave us a minor history of bronchial asthma during this period that was relieved by PRN inhalers. Had no family history of note and no tuberculosis contact.

On physical exam, the patient was ill looking, lethargic, and in pain. Febrile with a temperature of 38.6°, pulse rate of 120/min, but otherwise maintaining his pressures and saturation. Had a BMI of 16. Cardiac exam was only remarkable for soft heart sounds. His chest was clear and he had no lymphadenopathy.

On abdominal examination, his surgical scar was closed and looked well healing. Bowel sounds were audible and normal and his abdomen was soft and lax but tender all over even to superficial palpation with no significant rebound phenomenon. The rest of his examination including neurological exam was normal.

Investigations revealed leukocytosis of 18,800 cells/μL (P1.5 L1.2 E 6.6 M0). Repeated blood count on the next day also showed marked eosinophilia (P3.4 L1.2 E 8.8 M0) with total eosinophil count of 8700 cells/μL. The platelet count, renal, and hepatic panels were normal apart from an albumin of 18 g/L. Peripheral blood smear was normal and had elevated inflammatory markers with an ESR of 79 and CRP 236. Chest x-ray suggested pericardial effusion (see Figure 1)
Figure 1: Chest x-ray showing subtle globular heart shadow with otherwise normal lung parenchyma.

Figure 2: Electrocardiogram showing sinus tachycardia and low voltage in limb leads with some non-specific t wave changes.

Electrocardiography showed low voltage waves (see Figure 2), Echocardiography was done which showed large pericardial effusion (see Figure 3) and global impaired systolic function with ejection fraction of 40% suggestive of myopericarditis. His troponins were measured 3.3. He also underwent an enhanced abdominopelvic CT scan which showed diffuse wall thickening of the bowel with total collapse distal to the ascending colon. No thrombosis was noted in the abdominal vessels. HRCT done and showed bilateral small peripheral ground-glass opacities.

Figure 3: Apical 4 chamber view showing large circumferential pericardial effusion.

Pan-cultures were taken and the patient was started on Ceftriaxone, Metronidazole along with PRN Buscopan and bowel rest. He was admitted to our medical in-patient unit after being put on conservative management by the surgical team and he opened his bowel the next day. Stool examination of the specimen did not show any ova or cysts. There was yet no improvement in his abdominal pain or anorexia. Had an abdominal U/S which confirmed the previous finding of diffuse bowel wall thickening and showed mildly enlarged liver (16 cm) with normal spleen size. Diagnostic Pericardiocentesis was carried and drained 800 cc of clear yellow fluid. Later analysis of the fluid revealed an exudative eosinophilic effusion (LDH 997, fluid protein 77) with negative bacterial, fungal, and tuberculous studies (stain, culture, and PCR). Autoimmune panel including ANA, C and P ANCA, and anti-CCP were negative except for an elevated rheumatoid factor of 84. Hepatitis and HIV serology along with his pan-cultures were all negative. Echinococcus and Schistosoma serology were normal.

Hematology service was consulted for his unexplained eosinophilia. Bone marrow aspiration and biopsy was performed and reported back normal cellularity with increased eosinophils and eosinophil precursors. In view of the persistently elevated eosinophils count with multiorgan involvement, a diagnosis of hypereosinophilic syndrome was considered and intravenous methylprednisolone initiated at 1 mg/ kg/BID. Within two days of starting steroids his eosinophil count dropped to normal with total white count of 12,600 cells/μL (P₂ 2 L₂ 8 E₄ 4 M₃ ) and his symptomatology and abdominal pain improved dramatically and he asked to go out on pass for an important family issue to attend; however, he returned the next day with recurrence and worsening of his abdominal pain. Repeated ESR and CRP 50, 149 respectively.

Gastroenterology were contacted as well for both upper and lower GI scopes but the procedures were delayed on multiple occasions due to patient’s and logistical issues. OGD was carried out a week later and showed extensive esophageal candidiasis (see Figure 4), gastritis, duodenitis, and small white dots covering the second part of the duodenum (confirmed by histopathology to be candida as well) (Figure 5) and he was started on Fluconazole. Colonoscopy was attempted but never performed.

Figure 4: OGD showing extensive esophageal candidiasis.
4 days after starting steroids, he remained to have normal eosinophil count but his clinical condition deteriorated with the development of overt features of congestive heart failure with bilateral pulmonary crackles, elevated neck veins, tender hepatomegaly, and lower limbs edema. Repeated echocardiography revealed worsening global hypokinesia with ejection fraction of 15% and picture of fulminant myocarditis. He was started on diuretics and ACE-i. Cardiac MR showed mildly dilated left ventricle with global hypokinesia and an ejection fraction of 23%. Patchy areas of delayed myocardial enhancement and few areas of subendocardial enhancement were also noted all in keeping with a non-ischemic pattern of myocardial fibrosis such as that found in hypereosinophilic syndrome. No intracardiac thrombosis was reported.

9 days after starting steroids, he developed severe weakness. Neurological exam revealed profound weakness along with asymmetrical sensory and motor deficits in both upper and lower extremities in the form of hypertonia in the left upper and lower limbs with a spastic catch. Hypotonia in the right lower limb. Depressed power all over with 3/5 in upper limbs and 2/5 in lower limbs along with generalized depressed reflexes except for an exaggerated left knee reflex. Had flexor plantar response bilaterally. Sensory exam revealed patchy areas of fine and pain sensations loss along with diffuse loss of vibration and joint position sensations. MRI of the brain and spine did not show any pathology and metabolic work-up along with B12 level was normal. The patient was booked for nerve conduction studies but never had them because of his rapidly deteriorating course.

14 days into steroids therapy, his abdominal pain became severe and he developed peritoneal signs. Portable x-ray revealed pneumoperitoneum (see Figure 6). On the same night, laparotomy was performed and showed generalized fecal peritonitis along with two areas of perforation in the small bowel. 8 impending areas of perforation were also noted. Primary closure and washout were done with drains placement and biopsies. Examination of the samples revealed full thickness infarction with necrotic slough. No evidence of infection or malignancy was reported. Post-operatively he stayed in the intensive care unit for the remainder of his hospital stay and suffered from recurrent episodes of intraabdominal sepsis requiring two other surgical interventions and numerous courses of antibiotics. Biopsies from following operations revealed many CMV inclusion bodies which were not present previously and he was given Ganciclovir. He passed away from his illness a month later from his first operation.

3. Discussion

Hypereosinophilia entails a large list of possible etiologies. Initial efforts should be focused on ascertaining that eosinophilia is not secondary to other underlying disease processes, including helminthic parasite infections, varied types of adverse reactions to medications, and other eosinophil-associated syndromes, such as eosinophilic gastroenteritis, eosinophilic pneumonias, and Churg-Strauss syndrome vasculitis. Elimination of secondary causes is essential in making the diagnosis of hypereosinophilic syndrome. To make the actual diagnosis of HES 3 criteria were suggested by old literature: eosinophil count greater than 1500 cells/mL persisting longer than 6 months and single or multiple organ system dysfunction attributable to cytotoxic injury by eosinophils, without an identifiable etiology to explain the eosinophilia.[3]

In our case here after reasonable work-up we were able to fulfil two of these criterion, but the patient presentation did not meet the time period requirement. These criteria were found not useful or practical in the clinical practice, as new advances in diagnostic methods has evolved since the introduction of the criteria in 1975[4][5]. The duration can be shorter and no need to wait for the organ damage if the other causes is excluded[7].

HES disease spectrum has a heterogeneous presentation with a predilection for multisystem affection that can range from asymptomatic disease with elevated eosinophil count only to possible life-threatening organ involvement[6][8]. It is life threatening when it involves the cardiac or pulmonary system. Our case here illustrated many
of the features of HES and GI tract involvement specifically played a major role in his clinical outcome.

HES is rare and it’s not known the prevalence of it[10]. Due to the rarity and under-diagnosis of HES there are no published data on epidemiology of the disease and with the limited experience with this entity in our community, dealing with such a case created a challenge in reaching the diagnosis without the aid of the suggested cytogenetic studies and functional approaches.

Review of the literature for treatment of HES showed that steroid is the main initial modality in treating HES with usual good initial response[11]. Although our patient showed excellent early hematological response, unexpected and peculiar rapid clinical deterioration followed. The possible explanation for what happened to our patient that steroid led to tissue redistribution and deterioration in his organ involvement.

4. Conclusion

HES is a multisystem disorder secondary to eosinophilic infiltration of unknown etiology and it is reported to show a good initial response to high doses of steroids. What is interesting about the case presented here is the unusual and unfortunate response to therapy and the deterioration that quickly ensued soon after initiation of steroids with severe multisystem affection.

Competing Interests

The authors declare that they have no competing interests.

Authors’ Contributions

Both authors wrote and approved the final manuscript.

References


