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CASE REPORT

Focal Liver Lesions in a Child with Idiopathic Hypereosinophilic Syndrome: an Unusual Finding

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Abstract

Idiopathic hypereosinophilic syndrome (IHES) is characterized by the triad of peripheral blood eosinophilia (> 1500 cells/µl) for more than 6 months, without any other discernible cause of eosinophilia in association with signs and symptoms of organ infiltration related to eosinophilia. IHES is usually seen in the middle age group of 20–50 years and is uncommon in the pediatric population. Herein, we report a case of a 12-year-old male child with IHES who had multi-system involvement (hepatic, pulmonary, and cardiac). The child showed symptomatic, biochemical, and radiological improvement with steroid therapy on follow-up.

Keywords Hypereosinophilia · Idiopathic · Hepatic · Ultrasound · Computed Tomography

Introduction

Idiopathic hypereosinophilic syndrome is uncommon in the pediatric population and is often a diagnostic challenge for clinicians. It can cause multi-system involvement (cardiovascular, skin, pulmonary, central and peripheral nervous systems, gastrointestinal tract, and eyes). These patients are often misdiagnosed and present with unremitting symptoms and can show improvement with appropriate treatment. It is imperative to rule out eosinophilic leukemia in all these cases.

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Case Presentation

A 12-year-old male child initially presented with symptoms of intermittent fever, cough, and shortness of breath for 20 days, 6 months back. On examination, the child had decreased air entry bilaterally in axillary and infra-axillary areas with the rest of the systemic examination being within normal limits. Chest radiograph revealed bilateral pleural effusion. Pleural fluid analysis was done which showed eosinophilia with elevated adenosine deaminase (ADA) levels. Peripheral blood picture also showed eosinophilia (6995 cells/mm^3) with elevated acute phase reactants; however, sputum for acid-fast bacilli (AFB) and cartridge-based nucleic acid amplification test (CBNAAT) was negative. The patient was given a short course of antibiotic therapy with no improvement. Anti-tubercular therapy (ATT) was started, given clinical suspicion of tuberculosis, along with anti-helminthic therapy for parasitic infection, and the patient was asked to follow up in a week with a repeat complete blood count to monitor response for therapy. However, the patient came 6 months later for a follow-up with unremitting symptoms of intermittent fever and complaints of chest pain and abdominal pain. On examination, he had decreased air entry on the left side with tenderness in the right hypochondriac region and a pan systolic murmur over the mitral area. On further evaluation, the child was found to have eosinophilia in the peripheral blood picture with hypoechoic lesions in the liver and bone marrow aspiration/

biopsy showing eosinophilia with no blast cells. Because of suspicion of IHES, serum IgE was done which was elevated, and troponin level was also elevated. 2D ECHO revealed mild mitral regurgitation with a negative workup for rheumatic etiology. The acute leukemia panel also came to be negative. Serial chest radiographs which were done after a gap of two and one month, respectively, revealed consolidation in the left lower zone in the initial radiograph which resolved in the subsequent radiographs, and there was the appearance of a patch of consolidation in the right lower zone. There was also the development of bilateral mild-to-moderate pleural effusion in the subsequent radiographs (Fig. 1). Ultrasound abdomen was done (due to persistent pain abdomen) which revealed a few heteroechoic lesions with irregular margins in both lobes of the liver (Fig. 2A, B). CECT thorax and upper abdomen were done from outside, and the films were reviewed which showed bilateral pleural effusion with a patchy area of consolidation in the left lower lobe (Fig. 2C, D).

Repeat CECT chest and abdomen was done as symptoms of the patient were persisting; this is to evaluate the response of ATT which revealed multiple hypo-enhancing lesions in both lobes of the liver (Fig. 3A, B) with the presence of mediastinal lymphadenopathy (Fig. 2D). Consolidation in the left lower lobe was resolved; however, there was an appearance of consolidation in the right lower lobe (Fig. 2C). To know the nature of liver lesions, sampling was considered and advised; however, parents were apprehensive and did not give consent for the same. As there were fleeting opacities in the bilateral lung parenchyma, eosinophils in pleural fluid and bone marrow aspiration also showed eosinophils; therefore, focal liver lesions were also considered to be due to hypereosinophilic syndrome.

Hence, a diagnosis of IHES with multi-system involvement was made, and the patient was started on steroid therapy. No additional treatment was given for mitral regurgitation as it was mild in severity. The patient was followed up 2 weeks and subsequently 1 month after starting steroid therapy and showed significant improvement in symptoms (no fever/chest pain/abdominal pain), and a falling trend in eosinophil count was also noticed. Eosinophil count after 6 weeks of treatment was 1545 cells per mm^3. Ultrasound abdomen was repeated after 6 weeks of treatment and did not show any residual parenchymal lesion in the liver (Fig. 4A, B). Follow-up ECHO also showed resolution of the mitral regurgitation. At present, the patient is alive, healthy, and doing well on follow-up.

Discussion

Hardy and Anderson first gave the term hypereosinophilic syndrome which is characterized by the persistent absolute blood eosinophilic count over 1500 cells/mm^3 for more than 6 months, with involvement of multiple organ systems and without an identifiable cause of eosinophilia [1]. It most commonly occurs in middle age (20–50 years), predominantly in males (85%) [2]. FIP1L1-PDGFR alpha-associated variants mostly affected males, while other variants affect both males and females. Presentation varies from localized benign form to fatal disseminated form. The patient presents with various non-specific complaints depending upon the organ of involvement; initial presentation includes weakness, dry cough, and weight loss. GI symptoms include diarrhea, vomiting, abdominal pain due to eosinophilic gastritis, enteritis, or colitis.

There is a higher predilection for the involvement of hematologic, cardiovascular, neurologic, and cutaneous systems. The cause of mortality is either cardiovascular or neurologic involvement [3].

Cardiac involvement is the most serious complication which can lead to mortality in up to 75% of untreated patients. IHES involves the left ventricular wall, papillary

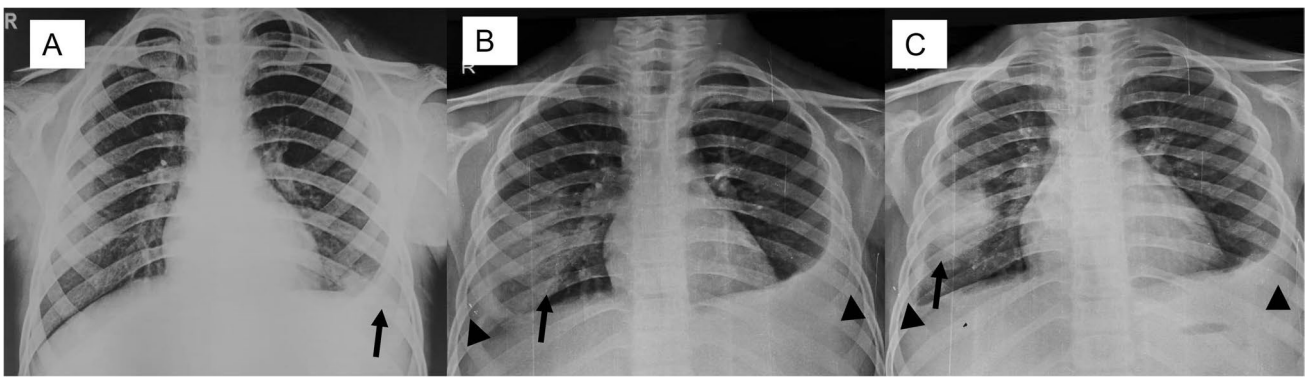

Fig. 1 (**A**–**C**) Serial chest radiograph (PA view) depicting consolidation in a left lower zone (black arrow in A) which resolved in subsequent radiographs, and there was an appearance of consolidation in the right lower zone (black arrow in B and C). Pleural effusion was also present in the subsequent radiographs which were not present initially (arrowhead in B and C)

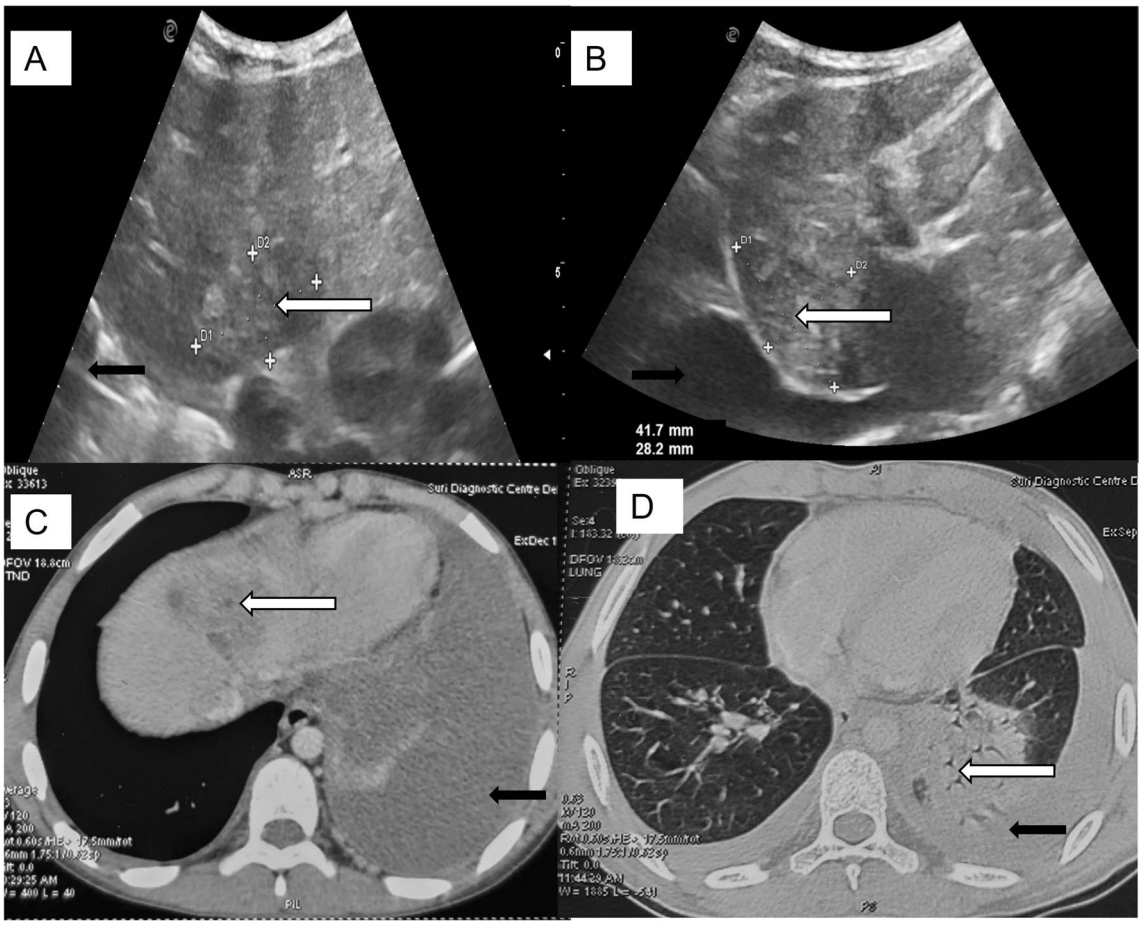

Fig. 2 (**A**, **B**) USG's abdomen transverse view through the liver shows two heteroechoic lesions in the right lobe of the liver (white arrow) and right pleural effusion (black arrow). (**C**) CECT abdomen and thorax show a hypo-enhancing lesion in segment VIII of the liver (white arrow) and left pleural effusion (black arrow). (**D**) The lung window shows an area of consolidation in the superior segment of the left lower lobe (white arrow)

muscles, or posterior mitral valve leaflet which can further lead to mitral regurgitation.

Pulmonary involvement is seen as either area of ground-glass opacities, consolidation, pleural effusion, or mediastinal lymph node enlargement. In a study by Dulohery, 31 out of a total of 49 patients had respiratory symptoms, and 21 patients revealed abnormal findings on chest radiography or CT thorax [4].

Hepatic involvement is reported in 33% of cases of hypereosinophilic syndrome and may present with chronic active hepatitis, focal hepatic lesions, eosinophilic cholangitis, or Budd-Chiari Syndrome [5]. In our case, the patient had pulmonary involvement in the form of consolidation with pleural effusion, cardiac involvement in the form of mitral regurgitation, and hepatic involvement in the form of multiple ill-defined hypo-enhancing lesions in both lobes of the liver. The other striking feature in our case was the young age of presentation in contrast with the usual presentation in middle age. Shatrey et al. reported two cases, one with a single hypodense mass in the right lobe of the liver and the other with diffuse involvement of the liver [5]. Jung et al. reported a case of a 17-month-old girl with hypereosinophilic syndrome and multiple hypodense lesions diffusely scattered in the periportal area of the liver [6]. Kim et al. also showed the presence of multiple hypodense hepatic lesions in 5 of their patients with hypereosinophilic syndrome [7]. The cause of liver damage in hypereosinophilic syndrome is substances released by eosinophils instead of a direct insult.

On histopathology, eosinophilic infiltration in the periportal and centrilobular areas will be seen. It is difficult to differentiate it from other conditions like metastases, lymphoma, or micro abscess radiologically, as the radiological findings of hepatic involvement of the hypereosinophilic syndrome are non-specific. So, a diagnosis can be made when other clinical and laboratory evidence is present.

These patients usually respond to corticosteroid therapy with the resolution of lesions.

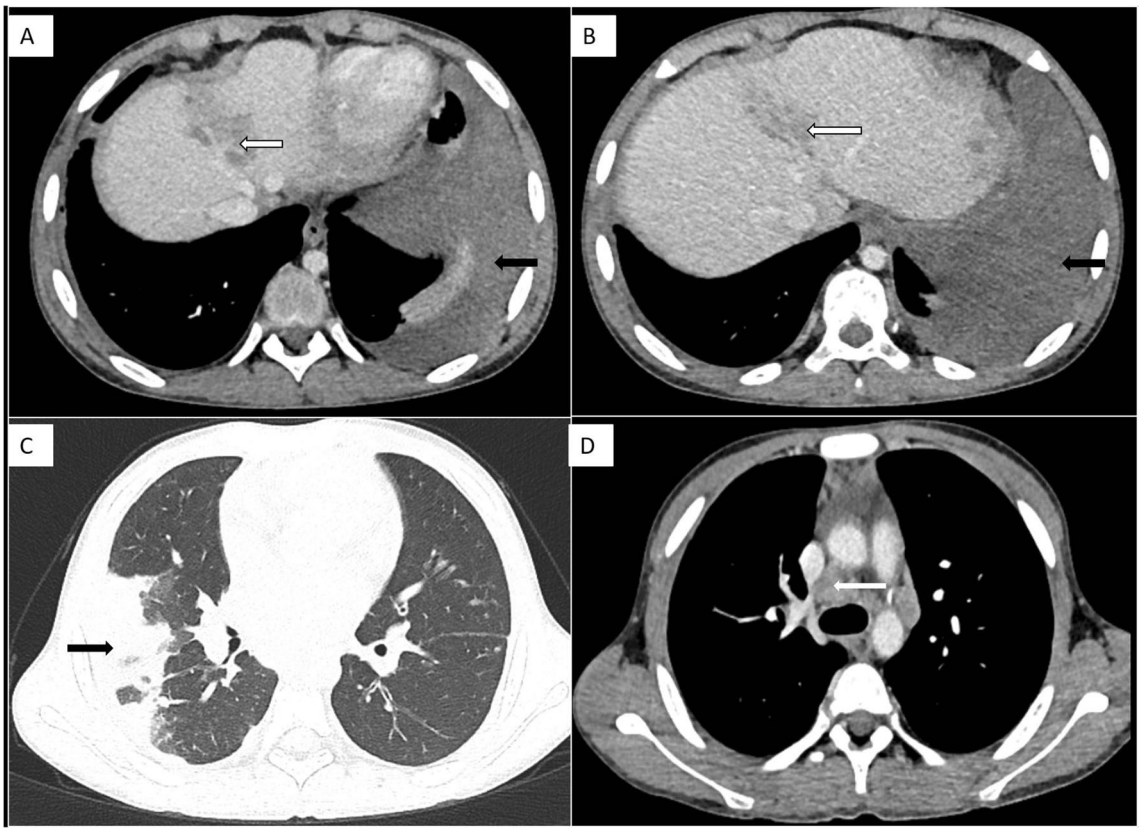

Fig. 3 (**A**, **B**) Axial CECT chest and abdomen (done after completion of treatment with ATT) which shows the persistence of hypo-enhancing lesions in the liver (white arrow in A and B) and left pleural effusion (black arrow). (**C**) The lung window shows an area of consolidation in the right middle and lower lobe (black arrow) resolution of consolidation on the left side, suggestive of fleeting areas of consolidation. (**D**) The mediastinal window shows enlarged mediastinal lymph nodes (white arrow)

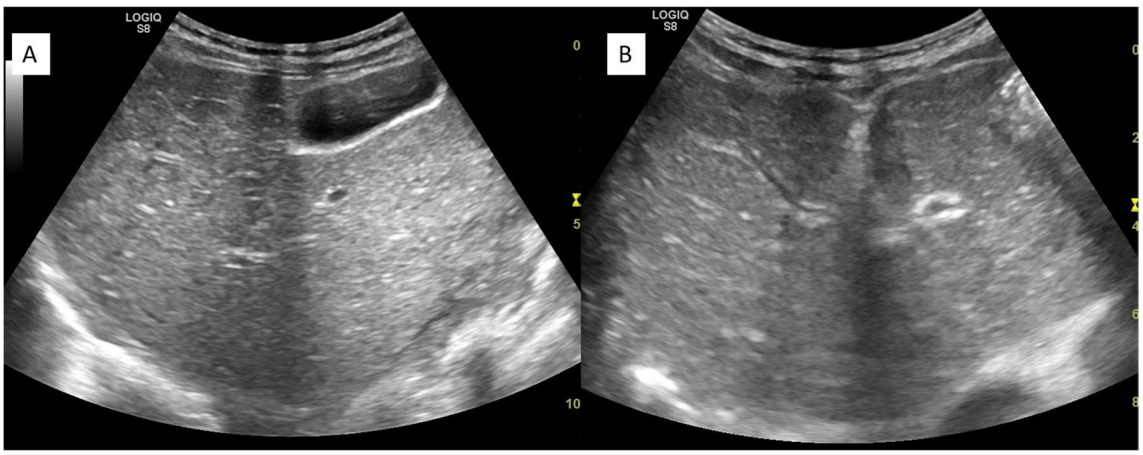

Fig. 4 Follow-up USG abdomen transverse view of the liver after treatment with steroids shows complete clearance of the liver lesions

To conclude, it is crucial to have a high clinical suspicion in patients with peripheral blood eosinophilia for a prolonged period without any obvious cause and multi-system involvement.

Learning Points/Take-home Messages

- Idiopathic hypereosinophilic syndrome (IHES) consists of a triad of Eosinophilia $> 1.5 \times 10^9$/L in at least two

instances without any underlying cause of eosinophilia (any parasitic infection, allergy) and eosinophilic infiltration of various organs leading to dysfunction.
- Imaging findings on the chest include ground-glass opacities (GGO), consolidation, pleural effusion, and mediastinal lymph node enlargement.
- Hepatic involvement in IHES is very rare and is rarer in cases of children, as seen in our case, and can be seen either as focal liver masses, chronic active hepatitis, eosinophilic cholangitis, or Budd-Chiari Syndrome.
- These patients are managed with corticosteroids, and improvement is seen as a decrease in the eosinophil count and resolution of pulmonary and liver lesions.

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Author Contribution PSH – conception and design of the work, drafting the article, critical revision of the article. SS – literature search, writing the initial draft, and figures. VS – clinical inputs, data collection. PSV – clinical inputs, drafting the article.

Data availability The datasets used and/or analyzed during the current study are available from the corresponding author upon reasonable request.

Code Availability Not applicable.

Declarations

Ethics Approval and Consent to Participate Ethical approval was taken from the institutional Research Committee.

Consent for Publication Consent of publication obtained from patients for figures included in this manuscript.

Conflict of Interest The authors declare no competing interests.

References

1. Leru PM. Eosinophilic disorders: evaluation of current classification and diagnostic criteria, the proposal of a practical diagnostic algorithm. Clin Transl Allergy. 2019;9(1):1–9.
2. Nam KJ, Jung WJ, Choi JC, Koo BS, Park BH, Lee KN, Han SY, Shin WW, Han SS. Hepatic involvement in hypereosinophilia: sonographic findings. J Ultrasound Med. 1999;18(7):475–9.
3. Inayat F, Hurairah A. Gastrointestinal and hepatic involvement in hypereosinophilic syndrome. Cureus. 2016;8(8).
4. Dulohery MM, Patel RR, Schneider F, Ryu JH. Lung involvement in hypereosinophilic syndromes. Respir Med. 2011;105(1):114–21.
5. Shatery K, Sayyah A. Idiopathic hypereosinophilic syndrome presenting with liver mass: report of two cases: idiopathic hypereosinophilic syndrome and liver mass. Hepat Mon. 2011;11(2):123–5.
6. Ran Jung M, Woo Goo H, Sook Hong S, Hyun Yoon C. A Case Report hypereosinophilic syndrome with hepatic involvement in a young child. J Korean Radiol Soc. 2003;49.
7. Kim GB, Kwon JH, Kang DS. Hypereosinophilic syndrome: imaging findings in patients with hepatic involvement. AJR Am J Roentgenol. 1993;161(3):577–80.