Case Report

# Liver abscess in a boy with hyper IgE syndrome

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## Abstract

Hyper immunoglobulin-E syndrome is a rare primary immunodeficiency disease, characterized by the classical triad of recurrent staphylococcal skin abscesses, pneumonia with pneumatocele formation, and elevated levels of serum IgE, usually over 2000 IU/mL. Chronic granulomatous disease, hyper IgE, and complement deficiencies are immunopathologies known to be associated with liver abscesses. We present a 2 ½-year-old boy with liver abscess and associated hyper IgE.

**Keywords:** Hyper IgE, immunopathology, liver abscess

## Introduction

Children with liver abscesses constitute more than 79/100,000 pediatric admissions in tertiary care centers in India.[1] Among the cases of pyogenic liver abscesses, *Staphylococcus aureus* is the leading cause in most series.[2] Chronic granulomatous disease, hyper IgE, and complement deficiencies are immunopathologies known to be associated with liver abscesses.[3] We present a 2 ½-year-old boy with liver abscess and associated hyper IgE.

## Case Report

A 2½-year-old boy presented in March 2011 with fever and vomiting for 8 days along with abdominal distension for 2 days. There was no jaundice. He had no other illnesses in the past. On examination, weight was 10.7 kg and he had hepatomegaly. Other systems were normal. Investigations showed hemoglobin of 7.9 g%, white cell count of 22,800/cumm, and platelets of 64,000/cumm. Serum glutamic oxaloacetic transaminase was 148 IU/L and serum glutamate pyruvate transaminase was 98 IU/L. Ultrasound (USG) abdomen showed multiple liver abscesses with largest being 201 cc. He underwent USG-guided pus drainage, and pigtail catheter was inserted in segment VII and VIII of liver. Pus culture and blood culture did not grow any organism. He was treated with IV vancomycin and clindamycin for 14 days and then oral ofloxacin and linezolid for the next 4 weeks. His HIV ELISA was negative, nitroblue tetrazolium was 98%, serum IgG = 29.5 g/L, IgA = 0.798 g/L, IgM = 1.86 g/L, and IgE was elevated (5420 IU/ml [normal = 3–423 IU/ml]). He had complete resolution of abscess in May 2011. In July 2011, he had paronychia which responded to oral fluconazole and topical clotrimazole. In October 2011, his USG abdomen was normal and IgE was high (4832 IU/ml). He is asymptomatic and on regular follow-up.

## Discussion

Hyper immunoglobulin-E syndrome (HIES) is a rare primary immunodeficiency disease, characterized by the classical triad of recurrent staphylococcal skin abscesses, pneumonia with pneumatocele formation, and elevated levels of serum IgE, usually over 2000 IU/mL.[4] This disease was first named as hyper IgE syndrome by Buckley *et al.* upon observing an association between recurrent staphylococcal abscess formation, chronic eczema, and high level of IgE in blood circulation.[5]

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Both autosomal dominant and autosomal recessive forms of the disorder have been described. Most autosomal dominant-HIES (AD-HIES) have been found to be due to mutations in signal transducer and activator of transcription 3, whereas dedicator of cytokinesis 8 mutations have been identified in patients with autosomal recessive-HIES (AR-HIES). Patients with AD-HIES also exhibit distinct dental, skeletal, and connective tissue abnormalities not found in patients with AR-HIES.[6] The resulting immunopathology results from an imbalance of $T_H$ 1 and $T_H$ 2 responses. There is a decreased production of interferon (IFN)-γ in contrast to relatively elevated production of interleukin (IL)-4, defects in the IL-12 pathway as well as under-expression of certain chemokines, adhesion molecules, transforming growth factor β, and IFN-γ messenger RNA in circulating activated T-cells.[7]

HIES is associated with recurrent abscesses involving several organs including the liver.[8] Patients with this condition develop pyogenic abscesses in the presence of bacteremia caused by *S. aureus*.[2] Eosinophilia is present in approximately 90% of the patients, and moderate to severe eczema is nearly found in 95% of patients with hyper IgE.[9] Our patient showed an unusual presentation of hyper IgE syndrome. He had increased IgE levels that persisted even after resolution of symptoms though he did not have eczema or eosinophilia. Therapy for HIES is directed at the prevention and management of infections by using sustained systemic antibiotics and antifungals along with topical therapy for eczema and drainage of abscesses.[3]

## Conclusion

The presentation of hyper IgE is highly variable which makes it easy to confuse the diagnosis with that of severe atopy or other rare immunodeficiency disorders. This case highlights that hyper IgE should be considered as a differential diagnosis when a patient presents with clinical manifestations as mentioned above.

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## Conflicts of interest

There are no conflicts of interest.

## References

1. Sharma MP, Kumar A. Liver abscess in children. Indian J Pediatr 2006;73:813-7.
2. Mishra K, Basu S, Roychoudhury S, Kumar P. Liver abscess in children: An overview. World J Pediatr 2010;6:210-6.
3. Wang DS, Chen DS, Wang YZ, Li JS. Bacterial liver abscess in peritoneum or GIT (haemobilia), and sometimes children. J Singapore Paediatr Soc 1989;31:75-8.
4. Yong PF, Freeman AF, Engelhardt KR, Holland S, Puck JM, Grimbacher B. An update on the hyper-IgE syndromes. Arthritis Res Ther 2012;14:228.
5. Grimbacher B, Holland SM, Puck JM. Hyper-IgE syndromes. Immunol Rev 2005;203:244-50.
6. Freeman AF, Holland SM. Clinical manifestations, etiology, and pathogenesis of the hyper-IgE syndromes. Pediatr Res 2009;65(5 Pt 2):32R-7R.
7. Borges WG, Augustine NH, Hill HR. Defective interleukin-12/interferon-gamma pathway in patients with hyperimmunoglobulinemia E syndrome. J Pediatr 2000;136:176-80.
8. Buckley RH, Wray BB, Belmaker EZ. Extreme hyperimmunoglobulinemia E and undue susceptibility to infection. Pediatrics 1972;49:59-70.
9. Farmand S, Sundin M. Hyper-IgE syndromes: Recent advances in pathogenesis, diagnostics and clinical care. Curr Opin Hematol 2015;22:12-22.