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Year : 2010 | Volume : 56 | Issue : 2 | Page : 143-145

Tricuspid endocarditis in hyper-IgE syndromeS Gupta¹, A Mittal², S Gupta³, Jagdish¹¹ Department of Medicine, MM Institute of Medical Sciences and Research, Mullana, Ambala, India² Department of Radiodiagnosis, MM Institute of Medical Sciences and Research, Mullana, Ambala, India³ Department of Dermatology, MM Institute of Medical Sciences and Research, Mullana, Ambala, India

Date of Submission	13-Sep-2009
Date of Decision	30-Dec-2009
Date of Acceptance	10-Jun-2010
Date of Web Publication	8-Jul-2010

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Source of Support: None, Conflict of Interest: None

DOI: 10.4103/0022-3859.65293

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Hyper-IgE syndrome is a congenitally acquired primary immune deficiency condition. We report a case of possible hyper-IgE syndrome who presented with multiple cold skin abscesses and chest infection due to *Staphylococcus aureus* and hyper-IgE findings. Patient also had tricuspid valve acute bacterial endocarditis with purulent

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pericarditis which is very rare. This case is presented to highlight that early diagnosis and treatment in such cases decreases the mortality and morbidity in phagocytic disorders.

Keywords: Endocarditis, hyper-IgE syndrome, tricuspid

How to cite this article:

Gupta S, Mittal A, Gupta S, Jagdish. Tricuspid endocarditis in hyper-IgE syndrome. J Postgrad Med 2010;56:143-5

How to cite this URL:

Gupta S, Mittal A, Gupta S, Jagdish. Tricuspid endocarditis in hyper-IgE syndrome. J Postgrad Med [serial online] 2010 [cited 2010 Jul 30];56:143-5. Available from: <http://www.jpgmonline.com/text.asp?2010/56/2/143/65293>

The hyper-IgE syndrome (HIES or Job's syndrome) is a rare disorder of immunity and connective tissue characterized by dermatitis, boils, cyst-forming pneumonias, elevated serum IgE levels, retained primary dentition, and bone abnormalities.^[1] There are two forms of HIES: a dominant form caused by mutations in STAT3, and a recessive form, for which a genetic cause is unclear. These two different syndromes have distinct presentations, courses, and outcomes and share very little in terms of pathogenesis other than the IgE elevation. The dominant form is characterized by non-immunologic features including skeletal, connective tissue, and pulmonary abnormalities in addition to recurrent infections and eczema. In contrast, the recessive form lacks the somatic features and has marked viral infections and neurological complications.^[2]

:: Case Report

A 32-year-old male, laborer by occupation presented with generalized weakness for two months with loss of appetite, and joint pains for the last one month. The patient also gave history of dry cough with chest pain and dyspnea, New York Heart Association (NYHA) Grade IV for seven days. There was also history of multiple swellings over the extremities off and on since childhood. Patient additionally gave history of recurrent eczema since childhood.

General examination revealed a febrile patient with pulse rate of 126/min and blood pressure of 120/80 mm of Hg. There was severe pallor, mild icterus, mild pedal edema, and a raised jugular venous pressure. The patient had coarse facial features with thickened skin and prominent chin and forehead. On systemic examination, there was bilateral wheeze with crepitations in the left fourth and fifth intercostal spaces on chest auscultation. In cardiovascular examination, the heart sounds were muffled, and no murmur was detected, the liver was palpable three fingers below costal margin and spleen was just palpable suggestive of hepatosplenomegaly. There was no tenderness and consistency was soft.

There were multiple subcutaneous swellings over the left thigh, right leg and right forearm, each of size approximately 5x4 cm which were non-tender, cystic to firm in consistency with normal temperature, normal overlying skin.

Routine investigations showed Hb-6g%, total leukocyte count (TLC)-20000/mm³ -

with polymorphs 60%, lymphocytes 25%, eosinophils 10%, monocytes 2%. Renal function tests were normal. Liver function tests revealed mildly raised bilirubin. Electrocardiography (ECG) showed low-voltage complexes. Chest radiograph revealed an enlarged cardiac shadow with multiple well- and ill-defined nodular opacities of various sizes in both the lung fields. Both the costophrenic angles were blunted [Figure 1].

Blood culture showed *S. aureus*. Ultrasonography showed hepatosplenomegaly with mild ascites. Echocardiography showed massive pericardial effusion with septations and thickened pericardium. Tricuspid valve showed large vegetation with tricuspid regurgitation [Figure 2]a and b. Mitral and aortic valves were normal.

Cold abscesses were aspirated and frank pus was drained from the subcutaneous swellings. Pus was sent for culture and sensitivity which showed *S. aureus*. Pericardiocentesis was done and that fluid also showed *S. aureus* sensitive to Methicillin, Vancomycin, Linezolid and resistant to penicillin.

In view of disseminated *S. aureus* bacteremia with cold abscesses, acute bacterial endocarditis, and purulent pericardial effusion, patient was evaluated for immunodeficiency disease but history and all examinations did not reveal diabetes mellitus, tuberculosis, acquired immune deficiency syndrome (AIDS), steroid intake, drug abuse, and any indwelling device. Then patient was evaluated for primary immune deficiency syndrome which revealed; IgG-279.7mg/dl (708-1605), IgA-477mg/dl (70-405), IgM-107 mg/dl (50-230), IgE-2511.40 IU/ml (158 IU/ml). Raised IgE levels with past history of recurrent skin infections with cold abscesses, bacterial endocarditis, and purulent pericarditis confirm the diagnosis of Job's syndrome. Retrospectively, he was evaluated for skeletal and dental abnormalities but no abnormality was found. Patient gave history of delayed shedding of primary teeth at age 10-12 years. There was no significant family history.

Patient was managed with intravenous vancomycin 1g b.d. and amikacin 50 mg b.d. along with supportive management of cardiac failure and anemia. Within one week patient responded to treatment and chest became clear, pericardial effusion very minimal, with no pedal edema. Vancomycin was given to patient for six weeks.

:: Discussion



Job's syndrome (hyperimmunoglobulin E syndrome) is a congenital/primary immunodeficiency characterized by extremely high levels of serum immunoglobulin IgE, chronic eczema, and recurrent *Staphylococcal aureus* infections. [3] Initially believed to be inherited as an autosomal dominant (AD) trait, autosomal recessive inheritance and sporadic cases have been described. In autosomal dominant and sporadic cases, hyperimmunoglobulin E syndrome (HIES) is part of a syndrome that affects multiple organ systems including skeleton, connective tissue, and dentition. The immunological features of HIES are characterized by recurrent skin abscesses, cystic lung infections with pneumatoceles (*Staphylococcus*, *Candida*), chronic mucocutaneous candidiasis, eczematous dermatitis, eosinophilia and elevated IgE levels. Infective lung lesions were also seen in our case on chest radiograph. Non-immunological features of AD HIES include bone fractures and other skeletal abnormalities, coarse facial features, joints' hyper-extensibility, and retained primary dentition. In contrast patients with recessive HIES are susceptible to viral infections characterized by severe *Molluscum contagiosum* and may develop severe neurological complications for unknown reasons. These patients lack dental, skeletal and cystic

lung changes. [4]

Recently, STAT3 gene mutation has been determined as a predominant cause of sporadic and familial hyper-IgE syndrome. All mutations associated with the hyper-IgE syndrome identified to date are restricted to the DNA-binding or SH2 regions of STAT3. These mutations are likely to preserve protein levels, phosphorylation, and nuclear localization. STAT3 is a major signal transduction protein involved in diverse pathways including wound healing, angiogenesis, immune pathways, and cancer. [1],

[2] We have not done molecular genetics in our case, so we cannot confirm and classify hyper-IgE syndrome genetically.

Patients with Job's syndrome appear to have inadequate inflammatory response that delays recognition of infection. Some postulate that decreased neutrophil chemotaxis accounts for poor inflammatory response. T-cell abnormalities including decreased production of T_H 1 cytokine and interferon-gamma (IFN- γ) and increased production of T_H 2 cytokine (IL-4) present with elevated IgE. Decreased production of T_H 1 cytokine and interferon-gamma (IFN- γ) reduces ability of macrophages to kill bacteria. Increased IgE levels cause histamine release which blocks certain aspects of the inflammatory process, so patients develop cold abscesses.

The IgE level in Job's syndrome exceeds 2000 IU/L and may be within normal limits (normal-0.1-90 IU/L) in 20% of cases. [5],[6]

One case of acute bacterial mitral valve endocarditis in Job's syndrome was reported.

[3] Tricuspid valve acute bacterial endocarditis is common in intra-venous drug abusers and patients who have had central indwelling catheters and right-sided heart instrumentation. The organisms responsible for tricuspid valve endocarditis include *Pseudomonas* or *S. aureus* in intra-venous drug abusers and *S. aureus* in patients with peripherally inserted central catheter lines/ central lines or right-sided heart cardiac instrumentation. [7] We are reporting here a case of *S. aureus* tricuspid valve endocarditis without any history of intravenous drug abuse or indwelling catheter in a patient of hyper-IgE syndrome which has not been reported till date.

Staphylococcus endocarditis is the most common cause of infective endocarditis and is potentially fatal with a mortality rate 25-47%. The course of bacterial endocarditis is fulminant with aortic or mitral valve involvement and metastatic spread that result in death in approximately 40% patients. [8]

Myocardial abscesses (with conduction disturbances), purulent pericarditis and valve ring abscesses are more common in *S. aureus* acute endocarditis than in subacute endocarditis. Peripheral suppurative foci (e.g. lung, brain, spleen, kidneys) are common and occur in more than 40% of these patients. Extravascular site involvement may offer clues to early diagnosis. In tricuspid valve endocarditis suppurative foci spread to lung fields. Mortality rate of tricuspid valve involvement is lower. [9],[10]

Our patient had history of recurrent skin infections since childhood but till now he had not been evaluated for immune deficiency. Recurrent skin infections led to bacteremia and bacterial endocarditis in our patient and he responded well to treatment.

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Figures

[Figure 1], [Figure 2]

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Figure 1: Chest radiograph PA showing enlarged cardiac shadow, bilateral pleural effusion and multiple nodular opacities in bilateral lung fields

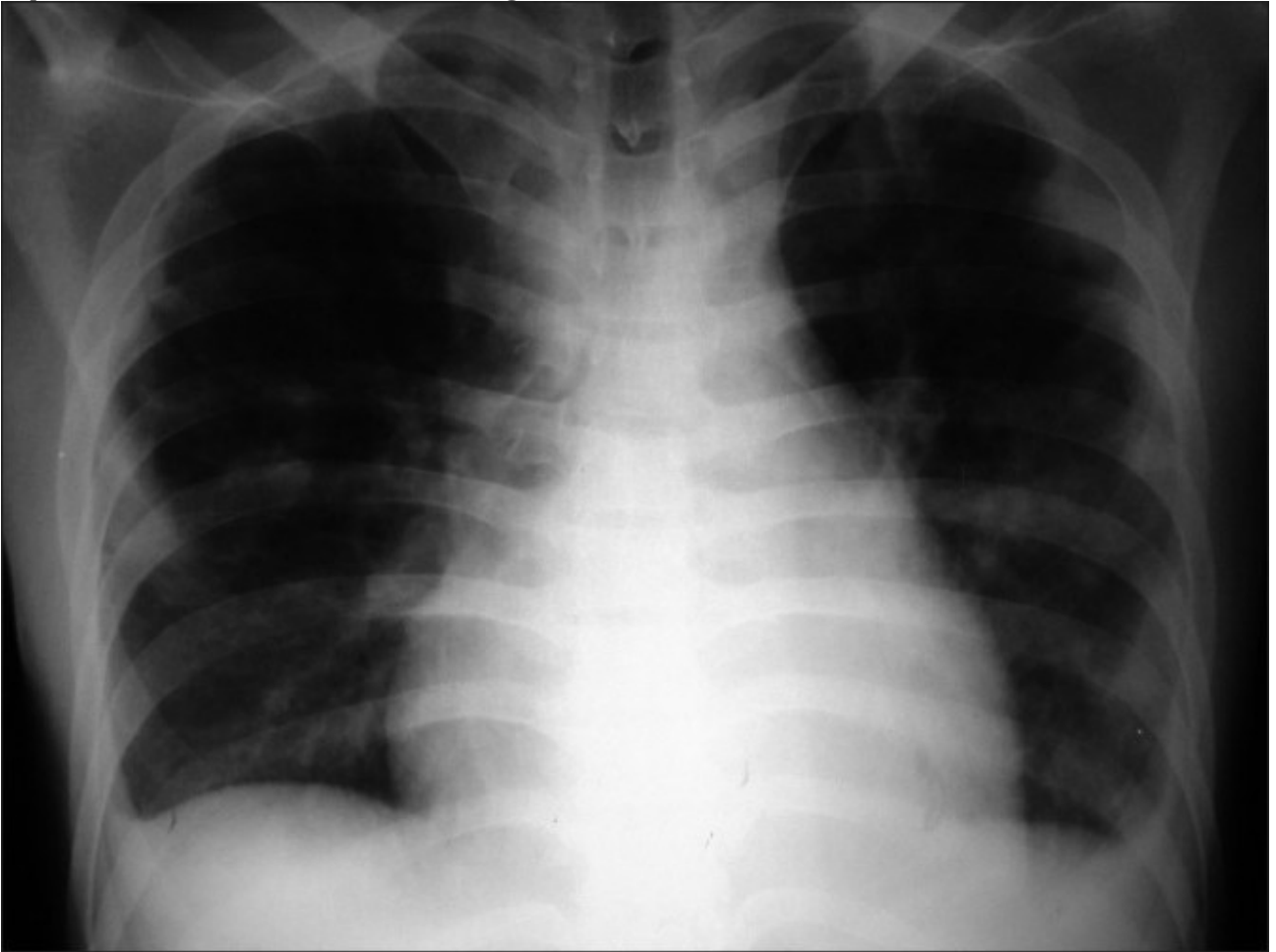


Figure 2a and b: Echocardiography is showing thick vegetation of tricuspid valve and pericardial effusion

