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CASE REPORT

Hyper Immunoglobulin E (IgE); an immune deficiency (Job Syndrome)

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A B S T R A C T

Hyper-IgE Syndrome (HIES), also known as Job Syndrome. HIES is a rare genetic disorder that affects the immune system and can lead to various health complications. The text mentions that the cornerstone of HIES therapy is proper skin care, infection prevention, and aggressive infection treatment. Treatments for HIES can include antibiotics that target S. aureus, typically used to reduce the incidence of pneumonia and the risk of lung damage. Skincare routines can involve bleach baths or chlorhexidine washes, oral anti-staphylococcal prophylactic antibiotics, and other treatments to avoid abscesses. Antifungal prophylaxis may be beneficial for HIES patients who have chronic Candida infections. Commonly occurring hypertension should be treated because it may be related to vascular problems. Defects in antibody synthesis can vary, making it difficult to provide general recommendations for immune globulin replacement in HIES. However, anti-staphylococcal prophylaxis is probably necessary for all individuals.

Keywords: *Hyper-IgE Syndrome, Splenic Abscess, STAT3 mutation, Eczema, Recurrent Infections*

Introduction

Eosinophilia, eczema, and recurring skin and lung infections are the hallmarks of the immunological deficit known as Job Syndrome, also known as High Immunoglobulin E (IgE) syndrome. Job syndrome is uncommon; it affects about one in one million people annually.[1] IgE levels are also high in primary childhood and begin early when two patients with eczema, recurring pulmonary infections, and cold lung abscesses were first diagnosed with "Job syndrome" in 1966, David et al. Later, in 1972, Buckley et al. found a connection between this syndrome and elevated serum immunoglobulin E (hyper-IgE) levels and behavioral characteristics known as HIES (Hyper Immunoglobulin E Syndrome). Almost two-thirds of individuals with job syndrome have a STAT3 gene mutation.

Case report

A 9-year-old male boy who had previously experienced bleeding wounds on the left anterolateral side of his chest and belly came to see us. Also, there was a six-month history of abdominal pain, fever, cough, vomiting, and various skin abscesses that flared up and down. History of therapy at many facilities without receiving ongoing care. A large infected ulcer measuring 20 x15 cm was discovered during an examination, with serous and purulent drainage, necrotic cheese debris, and the anterolateral side of the left chest and belly.

There were visible scars from previously treated abscesses on the hands and forearms. X-rays of the chest and abdomen showed haziness in the lower left lung. An uneven necrotic fluid attenuated region is visible in the CECT abdomen and chest. After receiving conservative

care and multiple debridements, the patient's wound began to heal. She was then discharged and gradually resumed oral feeding, but a few months later, she presented with nearly the same complaints. This time, CECT abdomen and immunoglobulin levels were checked because immunodeficiency was suspected, and the results revealed high IgE levels along with normal IgG, IgA, and IgM levels.CECT abdomen revealed a splenic abscess and localized collection at the perigastric region. A hyper IgE with skin lesions was diagnosed with job syndrome. IVIG was used to treat the patient. An exploratory laparotomy was planned. The peri-gastrin necrotic tissue was removed, a jejunostomy was made, and the splenic abscess was cleaned up. The culture of pus and necrotic tissue showed a fungus infection which was handled appropriately. The whole area healed without needing a skin graft, thanks to multiple wound debridements and dressings with vinegar, coco powder, and ginger.

The patient was advised to take care of his skin, use antimicrobial prophylaxis, and seek quick infection treatment. Pneumococcal vaccination was also administered to the patient. The patient's abdominal scar is healthy, with no new skin infections over the past year.

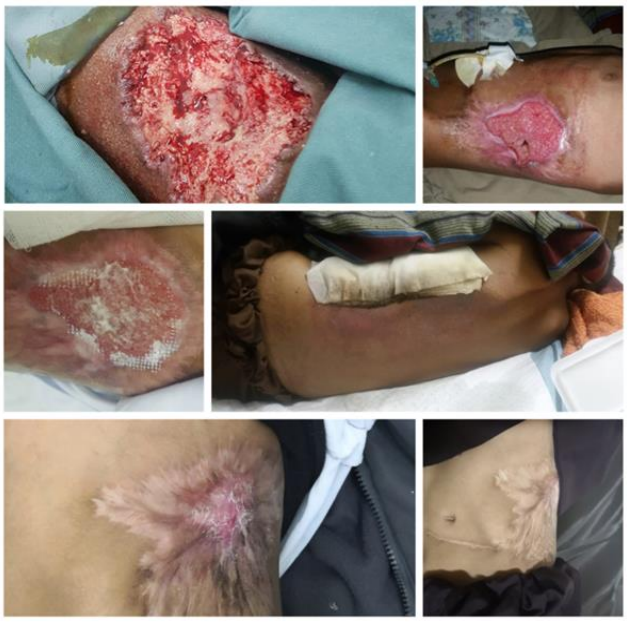

Figure 1: A Journey to Recovery: A Patient's Triumph over Job Syndrome Skin Infection

Discussion

An extremely rare immunodeficiency known as the hyper-IgE syndrome with recurrent infections is characterised by recurring cutaneous and lung abscesses and serum IgE levels that are abnormally raised. There have been identified skeletal and facial characteristics. Because it frequently results in abscesses, eczematoid rashes that last for a long time, and extremely high blood IgE levels, the hyper IgE syndrome has been predominantly classified as an immunodeficiency condition.[3]

A specific autosomal dominant genetic mutation in the STAT3 gene causes Job syndrome. The immune system and healing processes both involve STAT3. This gene's mutation will increase the amount of immunoglobulin E B cells produce, reduce their ability to be modulated by IL-6, IL-10, and IFN-gamma, and impair neutrophil chemotaxis. IL-10's lack of anti-inflammatory properties likely causes the inadvertent inflammatory response in Job syndrome patients. The absence of IL-6 causes a shortage of Th17 cells in Job syndrome because IL-6 is essential for developing Th17 cells. In the fight against the CD4 + Th17, cells are crucial. Autosomal-dominant and autosomal-recessive are two distinct variants of the condition that have been identified. Several skeletal, connective tissue, facial, and dental abnormalities are associated with the autosomal-dominant variant of the condition but not with the recessive form.

Chronic eczematoid eruptions, recurring skin and bacterial lung infections, and mucocutaneous candidiasis are the main immunological symptoms. These patients frequently develop cold staphylococcal skin abscesses with minimal to no inflammation. The skin, mucous membranes, and nails are frequently affected by candida infections. The most frequent side effects are upper airway infections, which manifest as paranasal sinusitis, exudative otitis media, otitis externa, mastoiditis, or respiratory tract infections. S. aureus, including methicillin-resistant strains, and rarely H. influenzae and Streptococcus pneumoniae are frequently to blame for severe reoccurring respiratory infections. The leading cause of death in HIES is pulmonary sequelae, which continuously contribute to the development of chronic respiratory insufficiency. Non-immunological Characteristics include distinctive face

characteristics that first show in early adolescence or earlier.[2, 4]

Reviewing documented lymphoma cases in people with Job's syndrome reveals a higher relative risk, particularly for class C Hodgkin and mature B cell lymphomas. [5] Proper skin care, infection prevention, and aggressive infection treatment are the cornerstones of HIES therapy. Although HIES patients may not exhibit the typical symptoms of infections, such as fevers, chills, or rigours, it is critical to take a thorough medical history, perform a physical examination, and use the right imaging to detect infections.

To reduce the incidence of pneumonia and the risk of parenchymal lung damage, prophylactic antibiotics targeting S. aureus are used (such as trimethoprim/sulfamethoxazole). To treat eczematoid dermatitis and avoid abscesses, skin care routines typically involve bleach baths or chlorhexidine washes, oral anti-staphylococcal prophylactic antibiotics, and these other treatments. Antifungal prophylaxis may benefit HIES patients with onychomycosis or other chronic Candida infections despite not being commonly administered. Aspergillus-treating antifungal medications. Commonly occurring hypertension should be treated because it may be related to vascular problems.[6, 7]

Defects in antibody synthesis can vary. 8 It has been difficult to provide general recommendations for immune globulin replacement in HIES due to this inherent variance in the B cell repertoire. Receivers of immune globulin are said to have fewer infections in certain publications, which is to be expected in some circumstances. Tests on antibody responses and replacement options in cases where deficiencies are evident seem reasonable. Anti-staphylococcal prophylaxis, however, is probably necessary for all individuals.[9-11]

Conclusion

A good collaborative effort of surgeons, physicians and pathologists can overcome any problematic diagnosis and management challenge.

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