Research Paper Medical Science

Job's Syndrome

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ABSTRACT

Job's syndrome is a rare syndrome presented with diffuse dermatitis, deep seated pyogenic infection and hyper IgE.

KEYWORDS Hypereosoniphilia, recurrent infection

INTRODUCTION :

It is a rare syndrome (incidence 1 in 10^6)[1], found equally in males and females. Most cases are sporadic or Autosomal dominant inheritance with variable expressivity. AR inheritance has also been described. It presents in first few months to 1st year of life[2]. It id due to impaired regulation of IgE function and deficient neutrophil chemotaxis.

CASE SUMMARY :

We here report a case of 8 year male child presented to out patient department with history of multiple pyoderma and itching. Patient was normal at the time of birth but with in 2 weeks he developed multiple pyodermas on and off. He also had a history of fever which is mild, remittent and relieved by medication. He also had a history of recurrent pneumonia.

Patient was delivered normally. He has two siblings, one is alive and normal but another one was expired in early infancy. His vaccination has been completed.

Patient had no history of any bleeding tendencies, diarrhea, polyurea, exfoliation of skin.

On general examination, mild growth retardation was present but mile stones were normal.

On systemic examination, no splenomegaly, hepatomegaly or lymphadenopathy.

on cutaneous examination : seborrheic like dermatitis present on face. Excoriated papules, pustules and excoriation marks were present on upper chest and back, neck, axillae, forearm


Fig. 1


Fig.2


Fig 3


Fig.4


Fig.5

Teeth were normal and nail dystrophy was present.

LAB INVESTIGATIONS:

On blood examination, hyper IgE was detected (2129.6) and hypereosinophil count was present (584)

RBS, WBC count and platelet count were normal.

Xray spine, Xray chest and XRay skull was normal.

DIFFRENTIAL DIAGNOSIS :

-atopic dermatitis

-Wiscort-aldriech syndrome

-Digeorge syndrome

-LCH

DISCUSSION :

This syndrome is also known as

-Buckley's Syndrome

-HIES (Hyper eosinophilic syndrome)

Only 150 patients has been described uptill now[3].

The gene for HIES located on chromosome 4Q21[4], although underlying cause is not known.

Classic triad of Jobs syndrome[5] :

-Recurrent staphylococcal skin abscesses

-Pneumonia

-High serum levels of IgE

Other common features include atopic dermatitis, scoliosis, fractures and dental abnormalities.

It is a multisystem disorder affect skin, soft tissues, skeletal system and dentition.

SKIN: excoriated papules, pustules, furuncles and abscesses (30% cold)[6] on scalp, neck, axilla, groin and periobital regions.
Paronychial infection
Eczematous dermatitis present in flexural region, postauricular and hair line areas.
SINOPULMONARY:
Recurrent bronchitis, lung abscesses, pneumonia and pneumatocele with bacterial/fungal super infection.
Recurrent otitis media and sinusitis.

CRANIOFACIAL :
Coarse facies with broad nasal bridge and prominent nose are universally present by the age of 16 years[7].

DENTAL :
It includes retained primary teeth and lack of development of secondary teeth.

MUSCULOSKELETAL :
Osteopenia (57%)[8] is common with at least 3 pathological fractures especially of long bones, pelvis and ribs.
Scoliosis (63%)[8]
Hyperextensiblity of joint (68%)[8]

DIAGNOSIS :
Classic triad for diagnosis is
Recurrent staphylococcal skin abscesses, pneumonia with pneumatocele formation and high S.IgE.
AR disease is characterized by recurrent bacterial and viral infection, autoimmunity and neurological complications that are often fatal in childhood.
AR HIES shows no tendency to form pneumatocele and had no skeletal and dental abnormalities. Eosinophilia and IgE levels tends to be more severe.

TREATMENT :
Antistaphylococcal antibiotics are effective for cutaneous infections.
Pulmonary abscesses require I & D and partial lung resection.
Ivlg therapy and cyclosporine has been successfully used.
Bone marrow transplant has been attempted in two cases of dominant HIES.

CONCLUSION:
Collobrating the history, examination and lab investigation, the diagnosis of JOB'S syndrome was made. It's a rare case which presented with classic triad of HIES.

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