Letter to Editor

From:
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Dear Editor,

Hyper IgE syndrome also known as Job's Syndrome is a rare primary immunodeficiency characterized mainly by eczema, recurrent cutaneous cold abscesses, recurrent sinopulmonary and skin infections and elevated IgE level (>2000 IU/ml), along withconnective tissue and skeletal abnormalities [1].

A 16 months old child born of non-consanguineous parentage, and a normal perinatal and family history with one healthy sibling was admitted to us with multiple skin abscess with minimal signs of inflammation along with bronchopneumonia. Pus was drained and it grew *Staphylococcus aureus*. Child was given Injection Amoxiclav and upgraded to vancomycin as per culture report and was discharged on oral Linezolid. 2 months later child was again admitted with similar complaints. Because of repeated episodes child was investigated for tuberculosis, diabetes, HIV and peripheral smear was sent for abnormal cells, but work up was normal except for eosinophilia in the blood count.

In view of repeated cold abscess serum IgE levels were sent and value of 3500 IU/ml was obtained. Due to financial constraints and non-availability in the institute confirmatory genetic studies could not be done. A diagnosis of Hyper IgE syndrome was made clinically and after treating the abscess patient was discharged on prophylactic antibiotics.

First described in 1966, the Hyper IgE Syndrome is rare multisystem immunodeficiency disorder. Incidence cannot be determined due to rarity of the disease. Autosomal dominant variant is caused by mutation in STAT3 gene, whereas genetic cause of autosomal recessive variant is unclear [2].

Chronic and recurrent lung infections lead to complications like bronchiectasis. Skeletal anomalies include scoliosis, osteopenia, minimal trauma fractures, hyperextensiblity and degenerative joint disease. Failure of exfoliation of primary teeth, cardiac and brain aneurysms and malignancies are other associations. Septicemia is main cause of death [3].

Treatment is aggressive skin care, prophylactic antibiotics. Immunomodulatory therapy and bone marrow transplantation is being explored as therapeutic option [4].

References

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Letter to Editor

Reply to letter

To

Dr Sunil Kumar,
Senior Resident, Department of Pediatrics,
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Sub: Reply to letter to Editor

Dear Dr. Kumar,

We are pleased to receive the communication related to suspected case of Job's Syndrome from you. However, we share important concerns regarding the aforementioned letter, as under:

1. The case report is very abridged. The phenotypic features of child whether fitting into those described for children with Job's Syndrome, detailed family history of other affected/suspected family members and evaluation for other features of Job's Syndrome is missing.

2. Vast knowledge of genetic defects associated with both Autosomal Recessive (at least four) as well as Autosomal Dominant forms of Job's Syndrome have been reported in Literature, unlike your draft mentioning no such clear data available for Autosomal Recessive forms.

3. No mention of eosinophilia, if detected, in the patient.

4. Many differential diagnosis and confounding factors need to be considered and evaluated for in such presentation.

5. A scoring System devised by US National Institute of Health (NIH) is available and helps in diagnosing possible, probable and definitive cases of Job's Syndrome. A more detailed description of associated symptoms and signs in child is required for this.

6. The References referred by you are quite old and newer updated References need to be referred to.

Regards,

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