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Jobs Syndrome – Hyper IgE Syndrome

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Abstract

Autosomal dominant hyper-IgE syndrome (AD-HIES), formerly known as Job syndrome, are mainly due to elevated serum IgE levels and recurrent skin infection. The common manifestation is anomalies of dentinogenesis. Diagnosis can be made by laboratory and radiological investigations. If identified early it can be treated. This patient presented to us and diagnosed as Job syndrome and now he is doing well in the follow up.

Keywords: NIL

INTRODUCTION

Autosomal dominant hyper-IgE syndrome (AD-HIES), formerly known as Job syndrome, is a condition that affects several body systems, particularly the immune system. Recurring pneumonia often results in the formation of air-filled cysts (pneumatoceles) in the lungs. Mutations in the STAT3 gene cause most cases of AD-HIES. A shortage of functional STAT3 blocks the maturation of T cells (specifically a subset known as Th17 cells) and other immune cells. Frequent skin infections and an inflammatory skin disorder called eczema are also very common in AD-HIES. Anomalies of dentinogenesis are possible manifestations. Decreased resorption of the roots of the deciduous teeth may result in prolonged retention of the deciduous teeth, preventing the appearance of definitive teeth. About 70% of the patients with Job syndrome have been reported to retain three or more primary teeth.

Case Report:

18-year-old boy, reported to us with increased cough and shortness of breath for the past 1 month, which was present since childhood. On examination bilateral rhonchi and crepts were observed. Chest x ray showed multiple thick-walled cavities. CT showed multiple lung abscesses. Patient provided us with a history of recurrent skin infections in childhood.

Diagnosis:

Immunoglobulin levels were checked and total IgE levels was found to be 57000, Eosinophils 24%.

Management:

Patient was treated with antihistaminic and intravenous steroids and antibiotics after which he drastically improved.

Clinical Implication:

Job Syndrome (Hyper-IgE syndrome) is a rare, primary immunodeficiency distinguished by the

clinical triad of atopic dermatitis, recurrent skin staphylococcal infections, and recurrent pulmonary infections. Thanks to the antibiotics, and if the diagnosis is made early, patient survival can be increased. Bone-marrow transplantation has been associated with mixed results in these patients. Although AD-HIES is associated with high morbidity and mortality, advances in medical care, close monitoring and patient compliance have led to improved prognosis, with survival up to 50 years or more.

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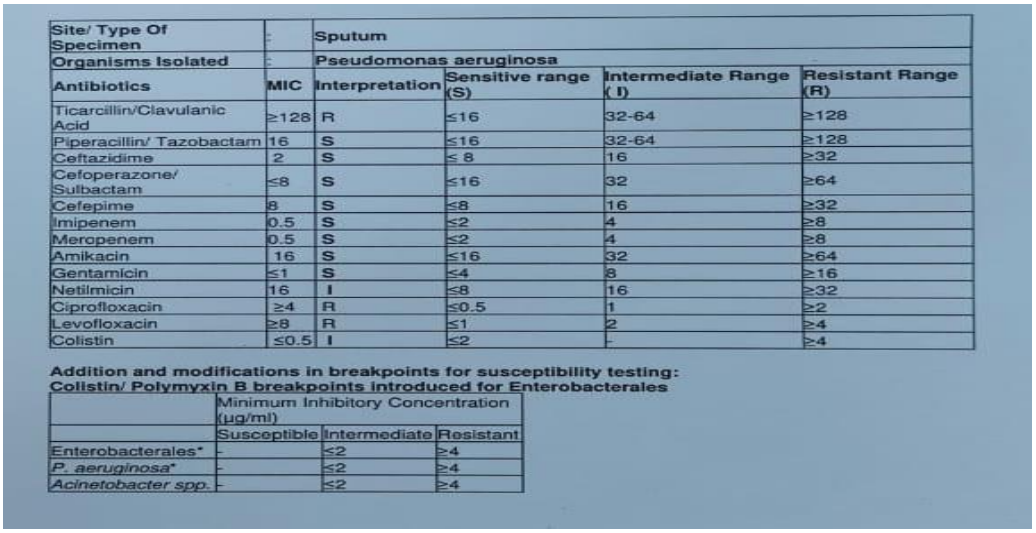
Figure 1: Sputum gram stain and culture sensitivity showing growth of Pseudomonas



Site/ Type Of Specimen	:	Sputum			
Organisms Isolated	:	Pseudomonas aeruginosa			
Antibiotics	MIC	Interpretation	Sensitive range (S)	Intermediate Range (I)	Resistant Range (R)
Ticarcillin/Clavulanic Acid	≥128	R	≤16	32-64	≥128
Piperacillin/ Tazobactam	16	S	≤16	32-64	≥128
Ceftazidime	2	S	≤ 8	16	≥32
Cefoperazone/ Sulbactam	≤8	S	≤16	32	≥64
Cefepime	8	S	≤8	16	≥32
Imipenem	0.5	S	≤2	4	≥8
Meropenem	0.5	S	≤2	4	≥8
Amikacin	16	S	≤16	32	≥64
Gentamicin	≤1	S	≤4	8	≥16
Netilmicin	16	I	≤8	16	≥32
Ciprofloxacin	≥4	R	≤0.5	1	≥2
Levofloxacin	≥8	R	≤1	2	≥4
Colistin	≤0.5	I	≤2	-	≥4

Addition and modifications in breakpoints for susceptibility testing:
Colistin/ Polymyxin B breakpoints introduced for Enterobacterales

	Minimum Inhibitory Concentration (µg/ml)		
	Susceptible	Intermediate	Resistant
Enterobacterales*	-	≤2	≥4
*P. aeruginosa**	-	≤2	≥4
Acinetobacter spp.	-	≤2	≥4

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Figure 2: Chest X-ray PA view showing multiple thick walled cavities

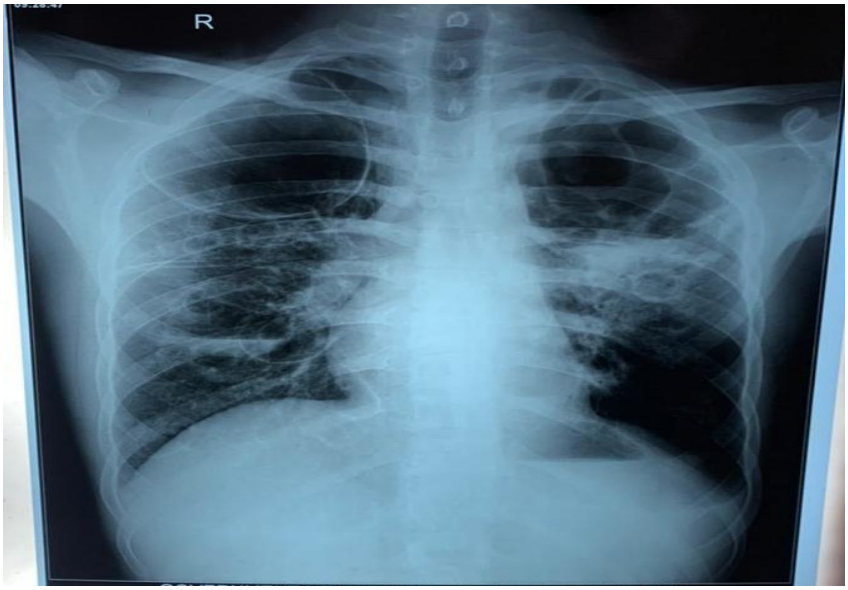

Figure 3: X-ray of teeth revealing retained primary dentitions

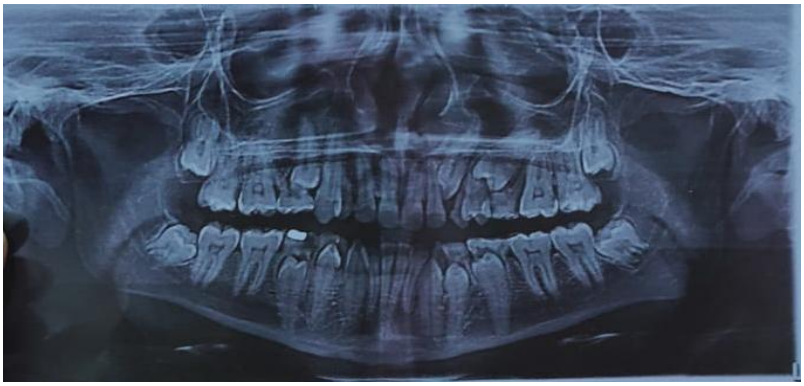

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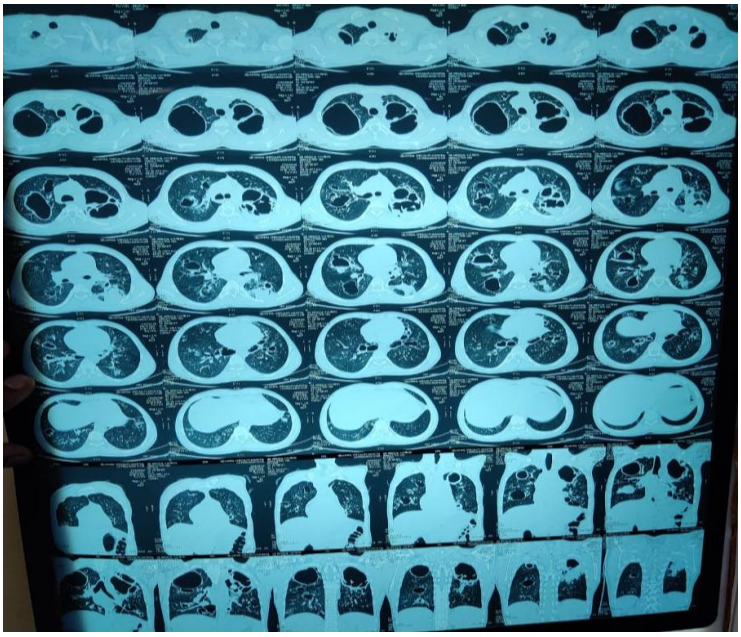
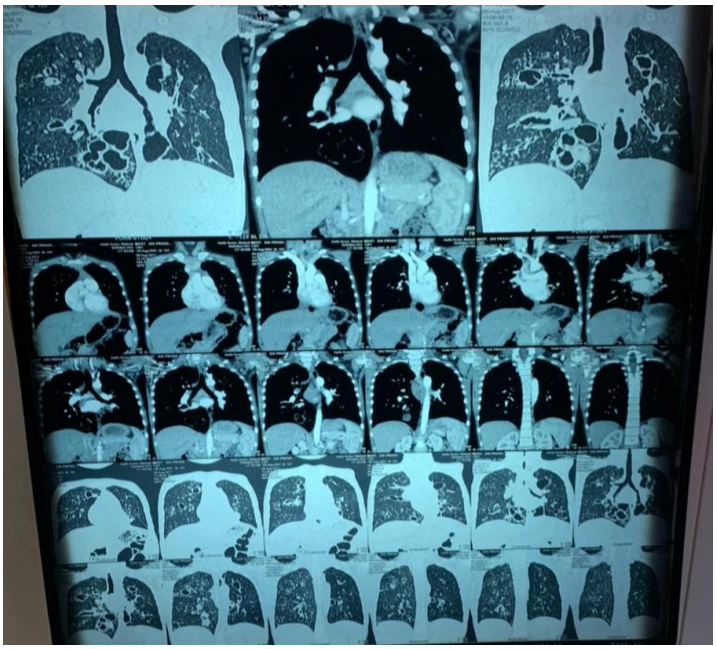
Figure 4: High resolution CT thorax showing multiple lung abscesses





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