Late onset Job syndrome With Growth Retardation

A 9-year-old girl presented with severe eczematous lesions and multiple infections since age of 6 year with growth retardation and raised serum IgE levels, suggestive of Job syndrome. The unusual late onset of clinical manifestations of the disease is highlighted.

Keywords: Hyper IgE syndrome, Recurrent infection, Eczema.

Job syndrome or hyper-IgE syndrome is characterized by eczema, recurrent skin and pulmonary infection, and elevated serum IgE levels (>2000 IU/mL) [1]. It is mostly sporadic with an incidence of one in 500,000 and has an early onset in life [1]. We report a child with uneventful early childhood and disease onset at 6 years of age.

A 9-year-old girl, product of a non-consanguineous marriage, presented with history of scaling over scalp and itchy red lesions with oozing and pus discharge in the inguinal region, trunk and lower limbs for past 4 months. There was history of discharge from the right ear and the left eye for past 2 months. The child was treated for tubercular cervical lymphadenitis two years ago. Subsequently child developed repeated episodes of bronchitis and wheezing. Birth and developmental history of the child were uneventful. Her growth parameters were apparently normal till 6 years of age. The siblings were all healthy and there were no similar complaints in parents or close relatives. On examination, the child had hypertelorism, broad and flat nose with increased inter alar distance. Her height was 121 cm (3rd – 10th percentile) and weight 20 kg (3rd – 10th percentile). There were thick adherent yellowish greasy scales all over scalp with sparsening of hair, hemorrhagic crusts and serous exudate over few areas. Left eye showed mucopurulent discharge, crusting and erythema of eyelid margins with matting of eyelashes. Left ear had features of chronic otitis media. Trunk, buttocks and lower limbs showed scaling and erythema along with foul smelling purulent discharge from the erosion over the inguinal folds. Vulvovaginal candidiasis and chronic paronychia of right thumb and left index finger were present. Cervical lymph nodes were enlarged. Skeletal examination revealed scoliosis in dorsolumbar spine. No dental
abnormalities were noted and intelligence quotient was normal for age.

Serum IgE level was elevated [4624 IU/ml (0-175 IU/mL], hemogram revealed eosinophilia (7%), and chest X-ray showed calcified opacities in the left hilum and the right paratracheal region suggestive of healed pulmonary tuberculosis. Thyroid function tests, serum cortisol, vitamin D and parathyroid hormone levels were normal. Pus culture showed *Staphylococcus aureus*. Needle cytology from cervical lymph nodes revealed reactive lymph node hyperplasia. She received topical antibiotics and oral and topical antifungals. Her skin lesions resolved in two weeks and scalp scales cleared over a period of one month with hair growth.

Most patients of Job syndrome present early in life with severe skin and lung infections [1,2]. Sporadic and autosomal dominant Hyper IgE syndrome have additional features like scoliosis, retained primary teeth, hyper extensibility and moderate eosinophilia. Autosomal recessive form lacks these features and presents with recurrent viral infections and severe eosinophilia [1]. The index patient probably had the sporadic form.

Most cases that have been reported so far had a very early onset of disease [1-3]. Wu, et al. [3] reported onset of disease before two years in 85.7% of patients. Antoniades, et al. [4] reported an overlap of Job syndrome and Dubowitz syndrome unlike the index patient who had no particular features of Dubowitz syndrome to explain her growth retardation. Investigations to rule out on endocrine cause were also normal. Repeated immunological stimulation, infections and prolonged drug intake could be a reason for her growth retardation. We report this case to highlight that Job syndrome should be kept as a differential in patients presenting late with multiple infections and growth retardation.

**SINU ROSE MATHACHAN* AND POOJA ARORA**

*Department of Dermatology, Venereology and Leprosy, Dr Ram Manohar Lohia Hospital and Post Graduate Institute of Medical Education and Research, New Delhi, India.*

*sinurose@gmail.com*

**REFERENCES**


---

**Subsultus Tendinum in a Child with Typhoid Fever**

A 5-year-old boy, residing in a semi-urban settlement of Vellore town, presented to the community clinic in the area with a history of three days of fever associated with sore throat, malaise, cough, nausea, and headache. The highest temperature recorded during the episode was 103.8 °F. As a part of the SEFI (Surveillance for Enteric Fever in India) protocol, a blood culture is performed for all study children who present with three or more days of fever, and hence the child’s blood culture was sent to the laboratory [3]. Blood culture grew *Salmonella enterica* serovar Typhi. Following the culture result, oral azithromycin (20 mg/kg body weight) was initiated and continued for 10 days.

Fever abated on the third day following the initiation of azithromycin. Four days following fever defervescence, the child was brought back with pain in the left side of the neck and shoulder spreading down to the scapular region, with no swelling, warmth or tenderness. Two days later, the child developed twitching movements over the left shoulder and scapular region.