Job’s Syndrome with Bilateral submandibular Abscess
A Very Rare Case Report

Abstract
We present a very rare case to be encountered by otolaryngologist which may be the first case report in its presentation. A 16 years old boy known case of Job’s syndrome presented to the emergency department with a history of 3 weeks bilateral painful submandibular swellings. This was accompanied by sinuses discharging purulent discharge. Physical examination and imaging study showed bilateral submandibular septated abscesses. Incision and drainage was done.

Keywords: Job’s syndrome, submandibular, abscess, bilateral.

Introduction
Job’s syndrome (hyperimmunoglobulinemia E) is an autosomal dominant immunodeficiency disease caused by defective T suppressor cell function leading to elevated levels of serum immunoglobulin E (IgE) with values reaching more than 10 times the upper limit of normal (normal: 200 IU/ml). It was first described by Davis et al in 1966 [1]. It included the triad of eosinophilia, eczema, and recurrent skin and pulmonary infections. Herein we report a case with unusual presentation which, by itself, a very rare encounter, and not reported yet as a presentation of Job’s syndrome.

Case Report
Sixteen years old Saudi male, a known case of Job’s syndrome, presented with bilateral submandibular swellings for 3 weeks which increased in size gradually with pain and purulent discharge from superficial sinuses and intermittent low grade fever with malaise and body ache. The symptoms were preceded by upper respiratory tract infection. No dysphagia or shortness of breath, night sweating, cough or weight loss. On presentation, there was bilateral, tender submandibular fluctuant swellings with very tiny sinuses draining out pus (Figure 1-A, B). The right one was 8X10 cm in size and the left one was 6X8 cm in size. He was having characteristic fancies including broad nose, deep-set eyes and coarse facial features.

Routine laboratory work up done for him and showed, slightly elevated WBC (12.65), platelet (712), and ESR (24). Also, IgE requested for him and found to be markedly elevated (38100). Computed tomography scan also done for him and showed bilateral submandibular septated collections with peripheral enhancement, Figure 2.

Patient was admitted under care of infectious diseases department and received intravenous cefazolin 2 gm IV every 6 hours and paracetamol. He underwent incision and drainage for both sides under general anesthesia. The patient improved and the drains removed on the second day post operatively. Stitches removed 1 week later after discharge from hospital. Patient was...
Figure 2: CT-neck with IV contrast showing bilateral submandibular abscesses

Figure 3: photos for the patient 1 month post operatively, showing absolute improvement
reviewed 1 month later with no recurrence or remnant, Figure 3-A, B, C. Bacteriological culture samples culture came positive for staphylococcus aurous, and histopathologically it was consistent with an abscess.

Discussion

Hyperimmunoglobulin E Syndrome (HIES), also known, as Job’s Syndrome is a rare primary immunodeficiency characterized by recurrent cutaneous abscesses, repeated respiratory tract infections and elevated serum levels of IgE. It is an autosomal dominant disorder with detected sporadic cases. Mutations on STAT3 (Signal Transducer and Activator of Transcription) gene, mapped on chromosome 4q, were described for this syndrome The most common infecting organisms are Staphylococcus aureus and Hemophilus influenza [1, 2, 3, 4, 5]. Pulmonary bacterial pneumonia and emphysema are the most frequent systemic infections and may result in pneumatoceles that become the nidus for further bacterial and fungal infection. Job’s syndrome is abscess formation without the anticipated degree of erythema and warmth and inflammatory response either locally or systemically, hence the term cold abscesses. Job’s syndrome has also been associated with scoliosis in 76% of adult patients and hyperextensibility of joints in 68% of patients [6, 7]. Patient with Job’s syndrome usually share a characteristic facial appearance, with a broad nose, deep-set eyes with prominent forehead, and generalized coarsening of the facial features with age [8, 9].

This is the first case report of bilateral submandibular abscesses as a presenting symptom for Job’s syndrome with very high serum levels of IgE. This case improved back to his usual state of life after drainage of the abscesses with systemic antibiotic within few days only. Reporting this case is important to add to the literature another view of presentation of Job’s syndrome and to consider it as one of the differential diagnoses of bilateral submandibular abscess.

References