Brock’s syndrome due to Common Variable Immunodeficiency Disorder: A case report

Dinesh Keran. S
Pre final Year- MBBS Student,
Department of Pediatrics,
Govt., Royapettah Hospital,
Chennai.
• 9 year old boy
• Born to a 2 degree consanguineous parents
  • Since 1 year of age,
• Recurrent episodes of cough, fever, ear discharge
• Frequent passage of normal stools
• Frequent skin abscesses
• Asthmatic father treated for TB 10 years ago
• Febrile fits till 5 years
On examination

- Pale and non icteric
- Sub- mandibular lymph nodes palpable
- Pot belly
- Phimosis +
- Pan digital clubbing and marked wrinkles
- Ear discharge
- Kyphoscoliosis
Systemic Examination

- **Respiratory system:**
  - **Dull note** heard over the right mammary and infra mammary region on percussion
  - **Bilateral crepitations** over infra mammary, scapular and sub scapular in both the lungs

- **Gastrointestinal system:**
  - Hepatosplenomegaly

- Other systems were normal
Differential Diagnosis

- Pulmonary TB
- Cystic Fibrosis
- Chronic granulomatous disease
- Primary humoral immunodeficiency disorder
- Immotile cilia syndrome
Investigations

- **Hematological**
  - Anemia and neutrophil leukocytosis
  - Neutrophils containing huge granules +

- **Infectious etiologies:**
  - Urine, stool, Widal, HIV, BAL: *Negative*, *Mx negative*,

- **Pus Culture:** MRSA

- **ECG:** Normal
IMAGING

- **CXR**
  - Consolidation of the lower lobe of the right lung.
  - Atelectasis of the right middle lobe of the right lung (Brock’s Syndrome)
  - Thymus prominent
- **X ray(PNS)**- Pan sinusitis
- **USG abdomen**- Hepatosplenomegaly.
• **Sweat chloride test** – Negative

• **Immunoglobulin assay**: All immunoglobulin levels are low (IgG, IgE, IgA, IgM)

• **Lymphocyte subset panel 7-**
  - No defect in absolute B cell, T cell and N K cells.
  - Relative increase in CD8 cells in relation to CD4 cells
• *Nitrozolium blue test* – Negative
  Not Chronic Granulomatous disease

• *Bone marrow aspiration:*
  • Hypercellular marrow showing striking granulomatous hyperplasia with increased reticulo-endothelial cells

PROVISIONAL DIAGNOSIS
• Common Variable Immunodeficiency Syndrome
Literature Review - CVID

- Prevalence - 1: 50,000 worldwide
- No sex predominance
- Commonly presents between 20 to 40 years
- Predisposition to autoimmune disorders
- Predisposition to malignancies
- Diagnosis by exclusion.

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