A patient with generalised warty lesions

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Case report

A 36 year-old male, presented with intermittent low-grade fever, night sweats, productive cough with haemoptysis and diarrhoea of two month duration. Additionally, he complained of anorexia and weight loss for the past six months. He also suffered from generalised warty skin lesions for the past five years and recurrent respiratory tract infections for the past two years. He has a past history of recurrent cellulites from the age of twenty.

On examination, he was febrile, emaciated and depressed. There were warts covering all parts of the body and oral mucosa which showed

evidence of Candida infection. He did not have jaundice, lymphadenopathy, clubbing, peripheral stigmata of endocarditis or features suggestive of chronic liver disease. Abdominal examination revealed an enlarged liver (liver span of 16 cm) and spleen. In addition, he had coarse crepitations in apical region of the right lung.

After the initial clinical evaluation, the following possibilities were considered; disseminated tuberculosis or lymphoreticular malignancy in the setting of immune deficiency state.

His full blood count showed low white cells ($<1000/\text{mm}^3$), anaemia (Hb 8.2 g/dL) and

thrombocytopenia ($90 \times 10^9/L$). ESR was 120 mm in 1st hour while blood picture showed normochromic normocytic red blood cells with marked leucopenia and low platelet count. Excessive rouleaux formation was noted. His chest X-ray showed right apical non-confluent shadow suggestive of consolidation. Examination of sputum for Acid Fast Bacilli was negative on three consecutive days and Mantoux test was also negative.

Ultrasound scan of the abdomen confirmed mildly enlarged spleen and there were multiple circumscribed hypoechoic lesions within the spleen. Some lesions showed central hyperechogenicity. There was moderate hepatomegaly. No ascites or enlargement of para-aortic lymph nodes were seen.

His liver and renal functions were normal while liver biopsy showed normal liver tissue. Bone marrow examination was normal and bone marrow culture was negative for bacteria or Acid Fast Bacilli. His blood, urine and stool cultures were sterile. HIV screening also was negative.

Since the above investigations did not point to a definitive diagnosis and there were features of immunodeficiency the following immunological studies were done.

Serum immunoglobulin concentrations

IgG	190 mg/dL	(200 – 1300)
IgM	70 mg/dL	(60 – 280)
IgA	100 mg/dL	(90 – 450)

Lymphocyte subsets

CD4	- 95 /L	(300 – 1400)
CD8	- 86 /L	(200 – 900)
CD3	- 194 /L	(700 -2100)
CD20	- 2 /L	(100 – 500)

The above findings were suggestive of common variable immunodeficiency and patient was treated with a combination of anti- tuberculosis therapy, broad-spectrum antibiotics and antifungal agents. While on treatment patient failed to show an improvement and subsequently

died. Postmortem findings including histology confirmed disseminated Aspergillosis involving the liver, spleen, kidneys and lungs.

Discussion

Common variable immunodeficiency is a heterogeneous disorder characterised by the following [1, 2]:

- markedly reduced serum levels of IgG (<400 mg/dL) and variably low IgA or IgM.
- recurrent bacterial infections.
- impaired antibody response despite the presence of B cells.
- normal or near-normal T-cell immunity in 60% of patients [1-3].

We arrived at the diagnosis based on reduced IgG, IgA and IgM with the

- (a) onset of immunodeficiency state after 2 years of age and
- (b) exclusion of defined causes of hypogammaglobulinaemia.

Fulfilling the third criteria which include the “absent or poor response to vaccines” was not practical in this patient.

Treatment includes intravenous immunoglobulin for life and antibiotics for the possible infections. Interleukin-2 (IL-2), Tumour necrosis factor (TNF) inhibitor and B lymphocyte stimulator (BlyS) therapy are been used as experimental therapies [2, 4].

References

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