Test yourself in eczema in primary immunodeficiency

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Choose only one answer:

1. Which of the following is characteristic of WAS?
   a) Eczema presenting in older age
   b) Thrombocytopenia
   c) Low B cell
   d) Normal T cell
   e) All of the above

2. Immunological features of Omenn syndrome include(s):
   a) Normal Immunoglobulin level
   b) Eosinophilia
   c) Low T cell number
   d) Good response to vaccination
   e) Abnormal T cell proliferation

3. To differentiate between eczema in an atopic patient and a patient with AD hyperimmunoglobulin E syndrome all are true EXCEPT:
   a) Site of eczema
   b) Response to treatment
   c) Presence of infection of top
   d) Elevated IgE >1000 IU/L
   e) Age of onset

4. Characteristic features in DOCK8 deficiency includes all EXCEPT:
   a) Presence of eczema
   b) Recurrent sinopulmonary infections
   c) Recurrent viral infections
   d) Low CD4+ cells and CD8+
   e) Coarse features

5. A patient aged 3 years, presenting with eczema, recurrent sino-pulmonary infections and viral infections, neutropenia. He had normal immunoglobulin level. He had mild CD4+ lymphopenia. He had mild pulmonary stenosis which was accidentally discovered. His EBV PCR was persistently positive for the last 6 months. What is your provisional diagnosis?
   a) DOCK8 deficiency
   b) AD hyperimmunoglobulin E
   c) Wiscott Aldrich Syndrome
   d) Mammalian sterile 20-like 1(MST-1)
   e) Shwachman- Diamond syndrome

(Answers on page 49)