Limited Systemic Sclerosis in a Young Boy

Abstrak:

Systemic Sclerosis is a chronic disorder characterized by diffuse fibrosis of the skin and internal organs. The cause of systemic sclerosis is unknown, but immune responses against unknown antigens have been implicated. Symptoms usually appear in the third to fifth decades, and women are affected three times more frequent than men. A 13 year old boy presented with hardening of left leg skin since 1 year before admittance. He was unable to both hands. The laboratory results showed slightly decreased hemoglobin, normal leucocytes and platelets, increased ESR, normal kidney and liver function tests, positive ANA test (weak), negative ENA and anti Scl-70. Other examination results showed normal X-Ray, Esophagogram, Schirmer test. Thyroid function test showed an euthyroid state. Systemic Sclerosis is established based on history of illness, physical examination and laboratory tests. ESR and CRP are increased in systemic sclerosis. Anemia in scleroderma can be due to various causes such as chronic disease, iron deficiency by gastrointestinal bleeding, B12 deficiency and folic acid deficiency. ANA test (IIF Hep-2) is positive in 60 â€“ 90%. Specific autoantibodies are Scl-70 and anticentromer. The anti Scl-70 can be used for prognosis.

Keyword:

Systemic Sclerosis, ANA test, ENA test