

# Juvenile Dermatomyositis

J. Mbutia, L. Owino

# V.A.E. History

- 4 years old male, well until age 1.5 yrs
- Two years swellings all over the body
- Swellings increasing in number and size over time
- Progressive weakness with inability to walk for over 1 year
- Previously treated for febrile illnesses and anemia
- Admitted in several hospitals with no diagnosis
- No relevant family history or consanguinity
- No relevant environmental history.
- Not on any prolonged treatment

# Clinical Examination at Admission

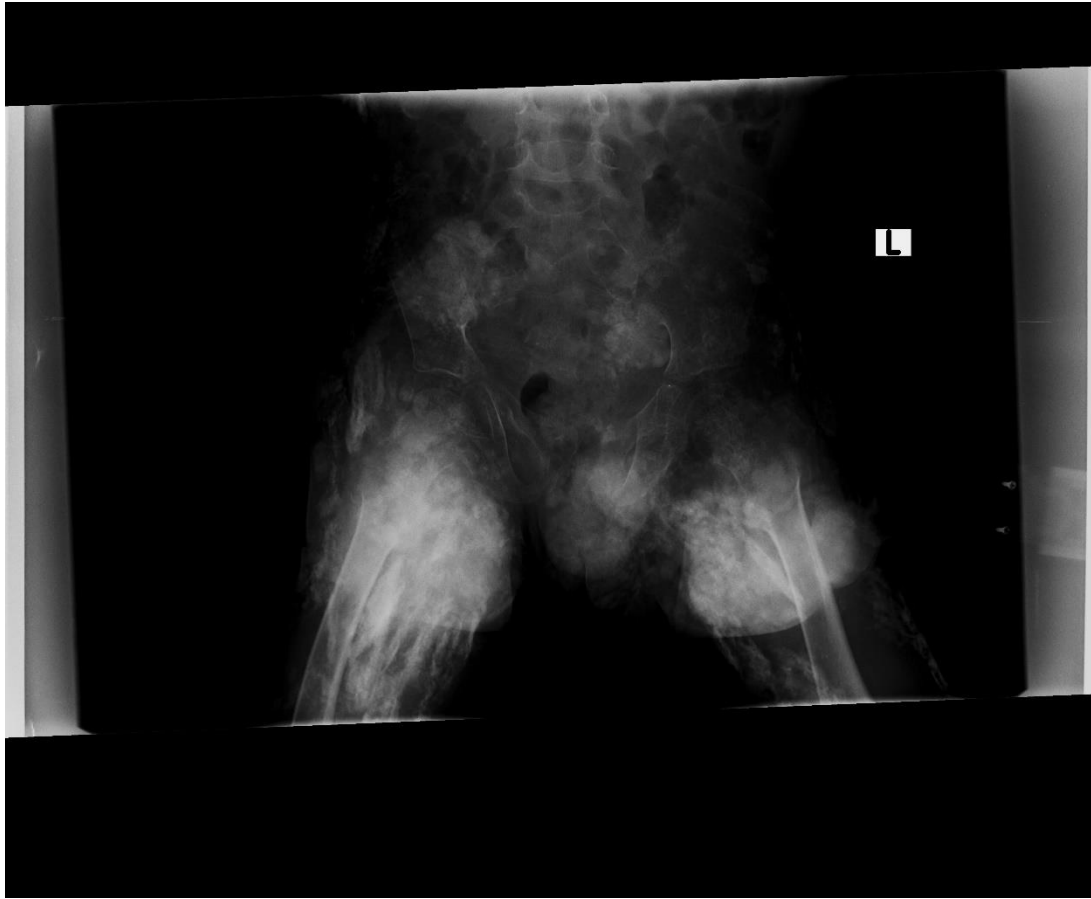
- Febrile, in fair general condition
- No pallor, enlarged lymph nodes, edema or jaundice
- Multiple discrete subcutaneous hard nodular swellings all over the body except the head. White gritty exudate in some lesions
- Contractures lower limbs with tender ulcerated swellings around the knees
- Lower limb movement limited by pain and contractures at the knees
- Upper limbs: normal muscle bulk, tone and deep tendon reflexes

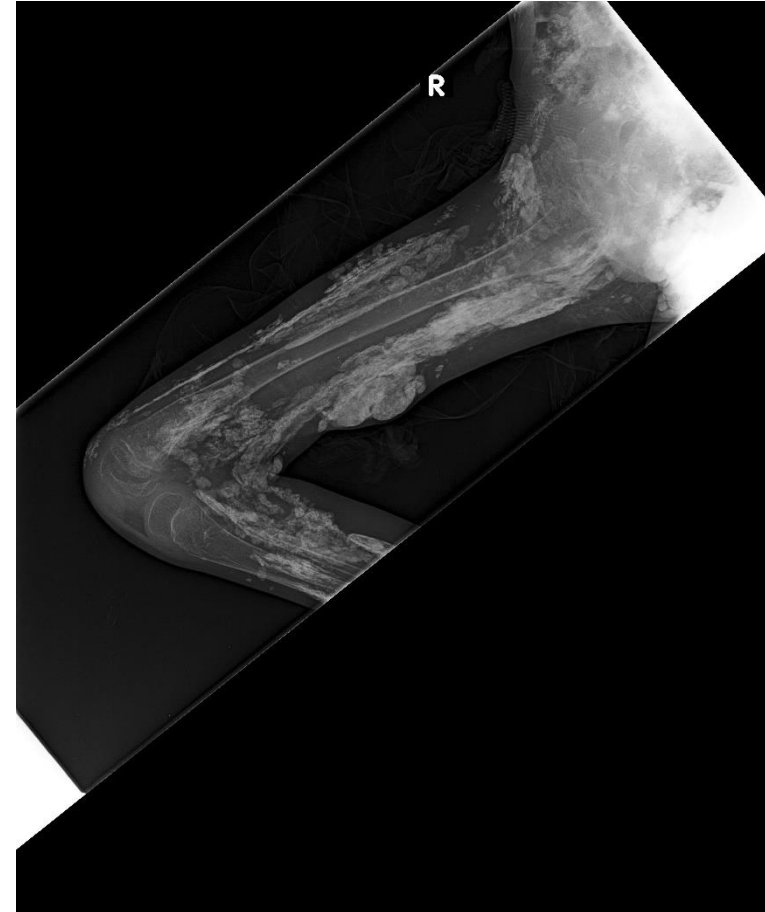
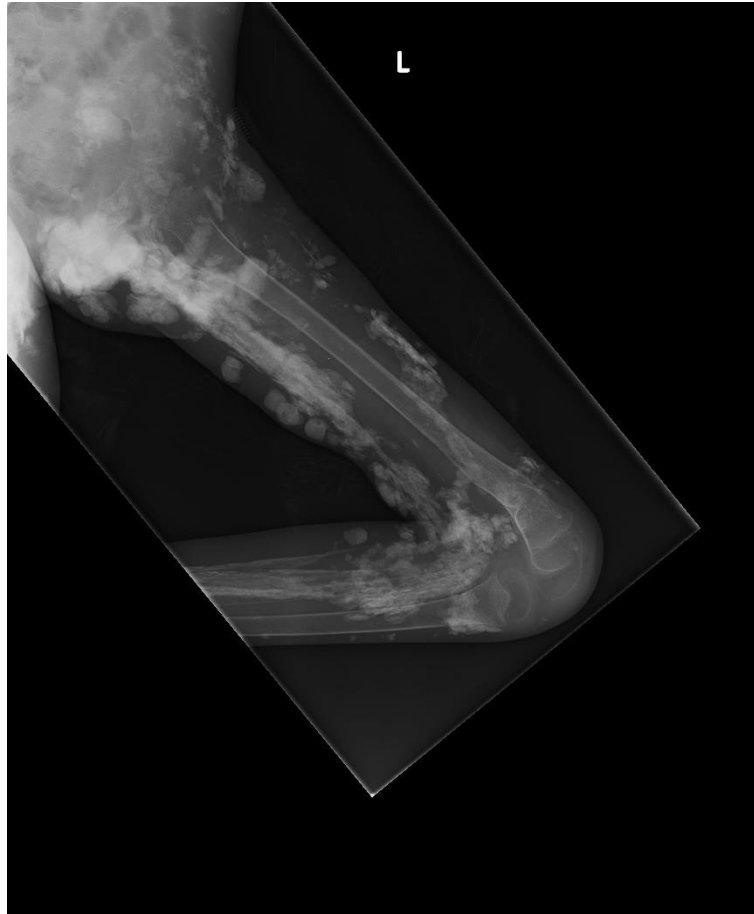
# Laboratory Investigations

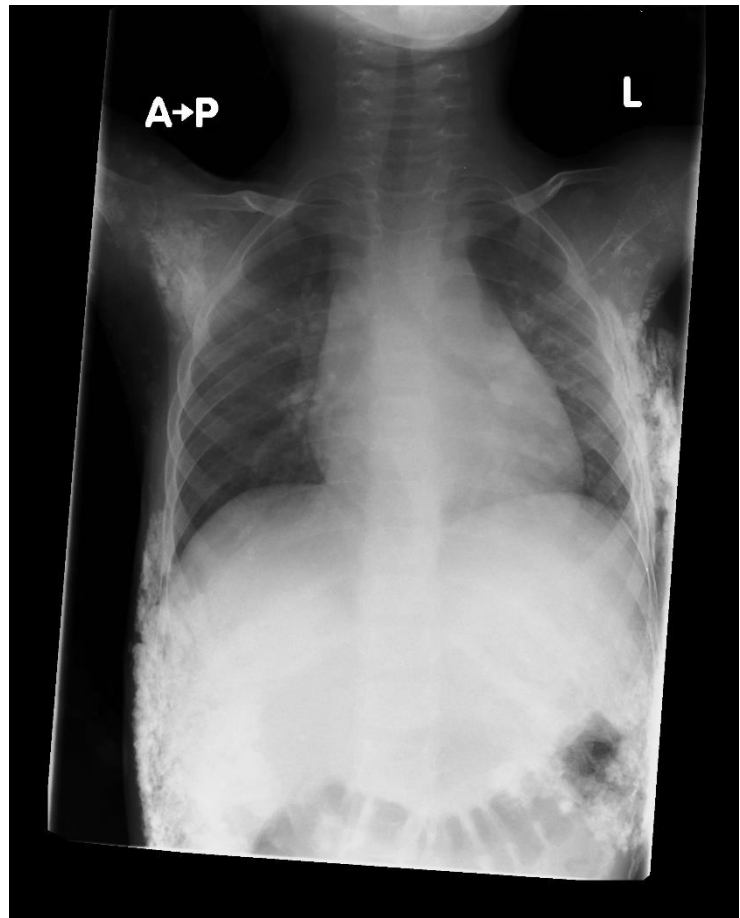
- ESR 35 mm/hr
- WBC 10.3
- Hb 11.9g/dl
- LDH 1086 (313-618)
- CPK 46 (55-170)
- LFTs, UECs normal.
- HIV negative
- Normal serum calcium, phosphorus, alkaline phosphatase

# Imaging

- CXR, X-ray limbs: extensive calcifications in muscle and under the skin.
- Osteopenia with fractures distal ends femora







J.K. MBUTHIA&L. OWINO



# Muscle Biopsy Histology

- Features suggestive of connective tissue disorder associated with subcutaneous calcinosis: Differential diagnosis include pathologies of thyroid and parathyroid, Ehlers Danlos syndrome

- Skeletal muscle within which are connective tissue septae exhibiting chronic inflammation with lymphocytes and histiocytes. Within the connective tissue are large foci of dystrophic calcification forming spherules and psammomatoid bodies. The skeletal muscle fibers are predominantly normal except in areas adjacent to inflammation where reactive nuclei are observed. No larvae, parasites, fungal or other infectious agents noted.