Images in Clinical Oncology

OSTEOPETROSIS (ALBERS-SCHONBERG DISEASE)

Fig-1 Chest X-ray showing densely sclerotic bones with poor differentiation between cortical and cancellous bone.

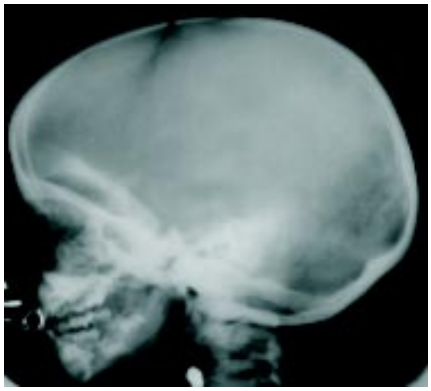

Fig-2 Skull radiograph showing absence of mastoid air cells and densely sclerotic bones of the skull base.

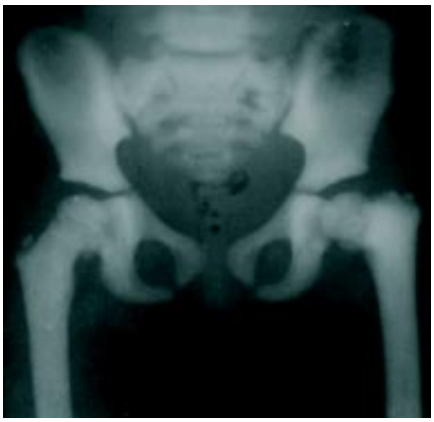

Fig-3 AP view of pelvis showing abundant callus formation and coxa vara deformity of the hip.

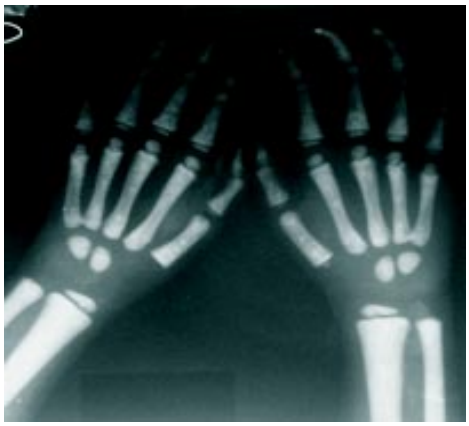

Fig-4 Radiograph of hand demonstrates bone within bone appearance.

A six year old female child presented to paediatrics department with high grade fever and ecchymotic patches and was diagnosed with haemorrhagic dengue fever. Clinical examination revealed severe pallor, short stature, left eye squint and hepatosplenomegaly Blood tests showed features of myelosuppression. X-rays (fig1-4) were suggestive of densely sclerotic bones and obliteration of medullary cavity. Past history revealed history of fracture of left femoral neck with trivial trauma. She received immunization as per schedule and had normal development milestones. The patient has been found to be HLA-identical with her brother and is cuurently undergoing evaluation for allogeneic transplantation.

Osteopetrosis is a clinical syndrome characterized by the failure of osteoclasts to resorb the bone which results in skeletal fragility inspite of increased bone mass (marble/ chalk bones), hematopoietic insufficiency, disturbed tooth eruption with maxillary/mandible osteomyelitis, nerve entrapment syndromes and growth impairment. Allogeneic bone marrow transplant is the only curative treatment for pediatric osteopetrosis.

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