

while others(4-7) reported incidence ranging from 33.6-38.1%. After lumping Wilm's tumors and neuroblastoma together, in a group of abdominal tumors it came at second place and recorded an incidence of 30.2% in our study. A similar observation was reported by Paul *et al.*(5) (35.6%), while Nair *et al.*(4) and Pathak *et al.*(7) reported a lower incidence of 20.4 and 13.5%, respectively. Retinoblastoma was present only in 1.3% in this study, while a high incidence of 17.2% reported by Thaper *et al.*(6). CNS tumors were present only in 1.7% of cases in the present study. Thaper *et al.*(6) reported similar observation (2.5%) while higher incidence was recorded by others 12.1-14.4%(4,5,7). The miscellaneous group had soft tissue tumors and epithelial carcinomas.

Our study reaffirms the view expressed by reports from other regions of India(4-7) that the incidence of reticuloendothelial malignancies is highest in distribution of childhood cancer, and in our country we have a pattern of increasing trend of leukemia, medium glioma and medium lymphoma(7).

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## Hypophosphatasia

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Hypophosphatasia is an autosomal recessive disease characterized by skeletal abnormalities, premature cranial synostosis, defective bone mineralization with low serum alkaline phosphatase and elevated excretion of phosphoethanolamine. There is defective regulation of alkaline phosphatase isoenzyme causing abnormal bone mineralization of growing bones. Severity of disease is variable. It may be congenital or develop later (hypophosphatasia tarda). The condition seems to be extremely rare in our subcontinent.

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We are presenting a case of hypophosphatasia (myopathic type) because of its rarity and interesting presentation.

### Case Report

A 4-year-old boy was admitted with history of growth failure and delayed milestones. The child had received treatment for rickets for last 2 years. The child was the product of non-consanguineous marriage and was 3rd in family of 4 children and other siblings were normal. The child's appearance at birth was normal and had delayed motor milestones, the child was not able to stand by 2 years and crawled after 4 years of age.

Physical examination revealed growth failure, the height and weight at 4 years were 80 cm and 14 kg respectively and, cranial sutures were closed. There was no clinical evidence of protein energy malnutrition. Patient had bowing of thigh, leg,

arm and forearm bones. There were non-tender broadening of metaphysis of long bones. There was history of delayed dentition and premature loss of deciduous teeth. He was not able to walk but could stand with support and move around by crawling because of muscle weakness of lower limbs (non-progressive myopathy). Clinically, the case appeared to be that of rickets with nonprogressive myopathy. Thyroid, parathyroid, pituitary, adrenals and genitalia were normal; liver, spleen and lymphnodes were not enlarged.

Skiagram of limb bones revealed a broadening of metaphysis, vertical multiple cystic areas of destruction giving appearance of ghost metaphysis characteristic of hypophosphatasia (*Fig.*). The epiphyses were of smaller size in relation to age and giving appearance of fine soap bubbles with irregular margin all around. Fibular epiphyses had not appeared even at age of



*Fig. Classical radiological features of hypophosphatasia in long bones.*

4 years. Similar changes were noticed in tarsal and metatarsal bones. Diaphysis appeared to be smaller with very thin cortex (Fig.).

Significant positive investigations were ESR-35 mm, alkaline phosphatase 2 KA units, serum calcium 14 mg/dl; serum phosphatase 1.8 mg/dl; and urinary phosphoethanolamine concentration 150 micro ml.

## Discussion

Hypophosphatasia is an extremely rare disease. This case appeared to be one of rickets with myopathy but radiological and biochemical findings were characteristic of hypophosphatasia. There are several case reports of hypophosphatasia in world literature(1-4). Seshia *et al.* observed association of nonprogressive myopathy in his two cases of hypophosphatasia as an important early sign(4). In our case there was weakness of muscles of lower limbs and the child was not able to stand by 2 years of age and started crawling and walking with support at age of 4 years (nonprogressive myopathy). They observed that this clinical feature resembles that of osteomalacia-myopathy described by Dastoor *et al.*(5) and concluded that there is no clear explanation for myopathy in few cases of hypophosphatasia. Wolfish *et al.*(2) also observed association of hyperparathyroidism and hypophosphatasia in a few cases. In our case there was no endocrinal disturbance.

We are presenting this case because of its association with nonprogressive myopathy and its extreme rarity in our subcontinent where rickets is common. This case was treated for rickets for 2 years without any response. Vitamin D therapy may have deleterious effects on hypophosphatasia(2).

Seshia *et al.*(4) concluded that non-progressive myopathy with osteomalacia syndromes in children may be an important early sign of hypophosphatasia.

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## Allergic Bronchopulmonary Aspergillosis

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*Aspergillus* species are ubiquitous in nature; with large numbers of conidia or spores dispersed in the air inhalation is inescapable. Allergic broncho-pulmonary aspergillosis (ABPA) is one of the hypersensitivity lung disorders caused by *Aspergillus fumigatus* (Af).

Following the first description of three

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