Cerebral Aneurysmal Arteriopathy in HIV Infected Children

We read with interest the article on cerebral aneurysmal arteriopathy in Human Immuno-deficiency virus infected children(1). Both the text and in the legend for figure 1 it has been mentioned as showing "T2W coronal section of Magnetic Resonance Angiography (MRA)…" We wish to point out that the said image is not an angiographic sequence i.e. it is not an MRA. It is just a coronal T2W section showing a large flow void in a portion of left internal carotid artery. The assertion about it being ectatic is also not clear in the given figure.

This is a very obvious and glaring error which does not befit an esteemed journal like *Indian Pediatrics.*

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Pamidronate for Fibrous Dysplasia due to McCune Albright Syndrome

Fibrous dysplasia (FD) is a rare disorder wherein scar tissue replaces normal bone-tissue, weakens the bone, causing deformity and intense pain. We present 2 cases of FD due to McCune Albright Syndrome (MAS) showing remarkable clinical improvement with pamidronate.

An 8 year-old girl presented with excessive weight gain since 3 years after fracture of right tibia/fibula following a trivial trauma; early fatigue, generalized bone pains and inability to bear weight due to extreme right leg pain. She weighed 45 kg with body mass index of 26.6 kg/m^2. Her height was 118 cm. She had multiple café-au-lait spots and bilateral genu valgum. Her pubertal status was B2P1A1M0, indicating early puberty. Serum calcium (9.6 mg/dL), phosphorus (4.8 mg/dL), parathyroid hormone (42 pg/mL, normal range: 9-65 pg/mL) and 25-hydroxyvitamin D (25OHD3) (21 ng/mL, normal range: 12-40 ng/mL) were normal, while alkaline phosphatase (ALP) (609 IU/L) (range: 40-240 IU/L) was high. Skeletal survey showed healing fracture of tibia with distal tibial cystic lesion, generalized osteopenia of foot bones with cortical thinning of leg bones (***Fig.*1**). Radiograph of shoulders revealed patchy sclerotic areas in proximal ends of both humerii with 'cotton wool' appearance. The diagnosis of FD was confirmed by MRI of right ankle and Dexa and bone scan (***Fig.*1**).

The second case was a 2 year-old female child with complaints of limping, bony pains, flat foot, and deformity of right ankle since 1 and a half years of age, which gradually progressed to right leg lengthening at presentation to our institute. She weighed 16.6 kg while her length was 82.3 cm. She had multiple café-au-lait spots (>6 mm with irregular borders). Parathyroid hormone levels were 19.6 pg/mL (normal range: 9-65 pg/mL). ALP was 585 IU/L (normal range: 40-240 IU/L) while calcium and

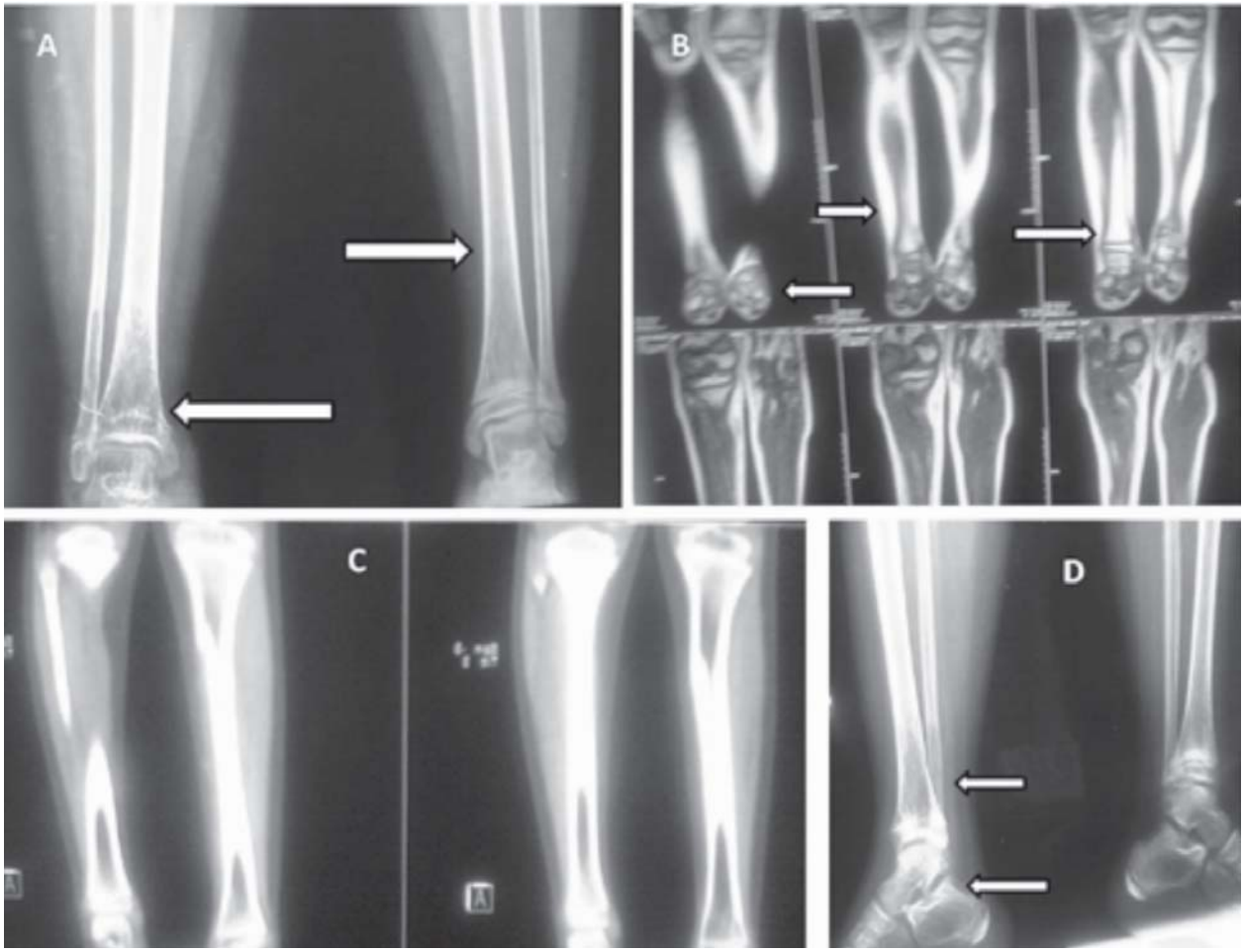

FIG.1 *A: X ray of bilateral tibia and fibula with ankles depicting healing fracture of tibia with distal-tibial cystic lesion with hohmogeneous ground glass matrix and cloud of smoke calcification, generalized osteopenia of foot bones with cortical thinning of leg bones. B: T2 weighted images of MRI depicting heterogenous hyperintensity in distal third of right tibia and small heterogeous hyperintensities in ankle bones. C: A section from computed tomography scan showing ground glass appearance of lower end of tibia. D: Lateral X ray depicting distal tibial cystic lesion, healing fracture and osteopenia of foot bones especially talus and calcaneus.*

phosphorus were 8.5 mg/dL and 4.1 mg/dL, respectively. Nuclear scan showed mildly increased tracer distribution in right tibia and left fibula, suspicious of FD. MRI right leg showed remodeling of proximal third of right tibia with cortical/ periosteal thickening and depressed postero-medial bone with focal cortical break in proximal third with surrounding tissue edema. The left fibula also showed remodeling along with periosteal thickening in the middle third.

Both patients were diagnosed as MAS and treated with pamidronate (in view of pain and reduced mobility) 1 mg/kg/day for 3 days given 3 monthly for 6 cycles along with metformin, calcium and vitamin D supplementation with appropriate diet, whereafter they improved. During the administration of pamidronate patients were under continuous electrocardiogram monitoring while vital signs were frequently recorded. Hypocalcemia (biochemical and manifest) was specifically observed. Calcium, phosphorus, ALP, 25OHD3 levels (27.5 ng/mL in case 1 and 32.4 ng/mL in case; normal range: 12-40 ng/mL) were normal at follow-up.

At 2 years follow-up, they showed improvement in pain (assessed by visual analogue scale), mobility,

deformity, general well-being and quality of life (ALP improved after 1 year). These 2 patients were able to perform all the activities of daily living appropriate to their age whilst the parents were satisfied with their overall progress. To the best of our knowledge, this is the first report from India describing role of pamidronate in FD due to MAS.

FD is characterized by replacement of normal bone tissue by fibrous connective tissue with a characteristic whorled pattern containing trabeculae of immature non-lamellar bone. Histopathologically, FD shows fibrous stroma with spicules of disconnected woven bone with a few mature osteoblasts and osteoclasts. Biphosphonates inhibit osteoclasts, reduce bone resorption and can lead to refilling of dysplastic lesions.

As observed by most other investigators, our observations highlight that good results can be obtained with pamidronate in FD, which should be administered early to halt disease progression, preserve bone mass, reduce fracture rates, avoid deformities, alleviate symptoms and delay/avoid surgery(1,2). Since standard guidelines for its use are unavailable, therapeutic response to pamidronate is noteworthy while longterm follow-up is awaited(3). Pamidronate therapy appears to be useful in children and adolescents with FD with a good short term safety profile. Potential multisystem (renal, hepatic, cardiovascular, gastrointestinal and skeletal) and oncological adverse effects of long term use are open to observation and speculation(4,5).

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Episodic Cluster Headache: A Rare Diagnosis in Children

Cluster headache is a rare disorder in childhood. We report an 8 years old girl who was referred by her family doctor at seven years of age with complaint of episodic headache for 7-8 weeks. Symptoms resolved and the child was symptom free for about nine months. She presented again with similar type of headache for 2 weeks. The headache was on forehead and vortex mainly on left side, was throbbing in nature and she described it "as if somebody is pushing on her head". The headache was worse in the evening and sometimes used to wake her from sleep. The headache occurred three to four times a day lasting for one to two hours each time. She did need regular analgesia (paracetamol and ibuprofen) to relieve the headache. During the episode of headache, she used to be in tears, restless and agitated because of severe pain and agony. She also had occasional eye pain on the left side along with rhinorrhea. There was no associated nausea but she vomited on a couple of occasions. She had good appetite without any weight loss. There was history of migraine in mother and maternal uncle, and cluster headache in paternal uncle. The positive findings on the general physical examination were