P Baburaj, VS Nandkumar, Laxmi Khanna
Department of Medicine, Medical College Hospital,
Vellapaya, Trichur 680 596, Kerala.
Received: 24.7.2003; Revised: 30.11.2005; Accepted: 10.3.2006

REFERENCES


Pachydermoperiostitis and Bisphosphonates

Sir,

Pachydermoperiostitis is a rare disorder and is characterized by clubbing and subperiosteal new bone formation.1 Typically, this constellation of findings occurs secondary to involvement of the respiratory, gastrointestinal or hematological systems with a neoplastic or infectious process. This is termed as “hypertrophic pulmonary osteoarthropathy (HPOA)” as it is most commonly associated with lung cancers. The primary/idiopathic form usually initiating around the peripubertal period, is known as pachydermoperiostitis. It is characterized by cylindrical thickening of the legs and forearms, “spade like” enlargement of the hands and feet and clubbing of the terminal phalanges involving the bones and the soft tissue. There is also progressive coarsening of facial features and greasiness of skin of the face. X-ray examination of the affected bones reveal irregular periosteal ossification. Diagnosis of idiopathic hypertrophic osteoarthropathy is made after excluding the causes of secondary hypertrophic osteoarthropathy. In the classic case, the condition is self-limited and progression stops at the end of adolescence.

A 14 years boy presented with gradually progressive acral enlargement and pain in both hands and feet of 2 years duration. The pain was moderate to severe and mainly present during limb movements and was partially relieved with rest. He had neither history of rapid height gain, coarsening of facial features, headache or visual disturbance suggestive of pituitary pathology nor any suggestion of pulmonary, cardiovascular or gastrointestinal disease. No other sibling or family members were afflicted by similar complaints. On general examination, the lower part of both upper and lower limbs was thickened, and hands and feet enlarged with palmarplantar hyperhydrosis. There was gross clubbing of all the digits without cyanosis. Systemic examination was unremarkable. Serum biochemistry including calcium and thyroid profile were normal. X-ray of the legs showed marked thickening of the periosteum and non-visualization of the marrow spaces and that of the skull was unremarkable with normal appearing sella. The 99mTc MDP bone scan showed markedly increased uptake in the distal ends of forearms and legs. The overall clinical profile was characteristic of pachydermoperiostitis with no discernible secondary causes. He was managed with intravenous pamidronate (60 mg IV) with which the pain subsided remarkably and is planned for repeated administration. One of the main concerns in the management of this condition is the pain associated with the bone resorption and periosteal thickening of various bones. In the secondary form, treatment of the underlying condition ameliorates the symptoms in at least a minority of the patients. Usual line of treatment includes use of non-steroidal anti-inflammatory drugs and corticosteroids for pain relief. Bisphosphonates, especially pamidronate has been used extensively in various other conditions like lytic lesions,2 secondaries, with varying success in HPOA associated with cystic fibrosis. In pachydermoperiostitis, bisphosphonates have been used in meliorating pain.3 In our case the patient reported significant improvement in pain after administration of a single dose of intravenous pamidronate (60mg) and no untoward effects (hypocalcemia) were observed. The dose may be repeated on follow-up depending on the clinical course. The rationale of bisphosphonate use in pachydermoperiostitis is an extension of their pharmacological action - osteoclast inhibition and anti-resorptive effect. Also, bisphosphonates are now being increasingly used in the pediatric age group for various other metabolic bone disorders viz fibrous dysplasia, osteogenesis imperfecta. Patients with osteogenesis imperfecta improved (reduction in fractures) without comprising linear growth.

A Bhansali, R Singh, M Srinraam, S Bhadada
Department of Endocrinology, Postgraduate of Medical Education and Research, Chandigarh - 160 012, India.
Received: 28.2.2005; Revised: 9.1.2006; Accepted: 10.3.2006

REFERENCES

