Severe bone and mineral disease in an adolescent with chronic kidney disease: a case from the 70s?

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DESCRIPTION

There is currently great focus on prevention of mineral and bone disorder from early stages of chronic kidney disease (CKD) in children.¹ Nowadays, the extreme complications of bone disorder in end-stage CKD are rarely seen in developed countries. However, access to medical resources in developing countries is difficult, and severe CKD bone and mineral disorder is still common.

A 13-year-old girl was transferred from Angola due to end-stage CKD. She was undergoing haemodialysis from the age of 9 years, 3 years after the diagnosis of CKD of undetermined aetiology. Her mother reported progressive bone deformities in the child and an increase in head circumference during the previous year. At physical observation, a head/body disproportion with macrocrania was noted (figure 1); the patient weighed 22 kg (SD=5.33), had a height of 130 cm (SD=3.9) and head circumference of 59.5 cm (>p98). Ancillary testing revealed serum calcium 2.65 mmol/L, ionised calcium 1.24 mmol/L, phosphorus 1.52 mmol/L, parathyroid hormone 323.3 pmol/L (3047 pg/mL) and 25-hydroxy vitamin D 49.7 nmol/L. Skull radiography and head CT scan revealed a diploic enlargement of skull and face bones (figure 2), and the patient was diagnosed with uraemic leontiasis ossea. During follow-up, she had continuing bone and mineral disease that was difficult to control, with persistent high levels of parathormone (72.3–1006.9 pmol/L), despite therapeutic adjustments with increasing doses of vitamin D analogues.

Figure 2 Cranial tomography scan with diploic enlargement and mineralisation changes of cranial bones.

Uraemic leontiasis ossea is an inflammatory hyperostotic bone disease, localised to the cranial and maxillary bones.^{2 3} This entity is a rare manifestation of CKD and seldom seen in children, especially in developed countries. Management of refractory secondary hyperparathyroidism is not clear and could include a trial of off-label treatment with cinacalcet, before considering parathyroidectomy.

Figure 1 Macrocrania and enlarged facial bones—leontiasis ossea.

Learning points

- ▶ Uraemic leontiasis ossea is a rare entity but should be kept in mind in children with head enlargement and long-term chronic kidney disease (CKD).
- ▶ Phosphorus, calcium, parathyroid hormone and vitamin D control is essential from early stages of CKD in order to prevent bone deformities.

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