Nephrology Dialysis Transplantation

Nephroquiz For The Beginner
(Section Editor: T. J. Rabelink)

‘The vanished occiput’

A 45-year-old patient suffering from malignant hypertension and who had been on maintenance hemodialysis for 3 years, was admitted to an outside institution for bone scintigraphy. This investigation was asked for because of several abnormalities of calcium metabolism including elevated intact PTH (160 pmol/l; normal <6). The bone scan showed a ‘black’ occiput, but was otherwise completely normal.

The radiologists considered the following possibilities:

— Brown tumour in a patient with hyperparathyroidism
— Paget’s disease
— Vanishing bone disease (Gorham’s disease)
— Metastasis of an unknown primary

What is your diagnosis? (Answer on the next page)

Fig. 1. 99m-Techmeticum–MDP—bone szintigraphy.

Fig. 2. CT of the skull.
Answer to quiz on preceding page

A brown tumour as a result of secondary hyperparathyroidism is excluded because the CT scan did not show expansion of the lesion as a mass. The shape of the strongly demineralized occiput is completely preserved as shown in Figure 2.

Paget’s disease with localized bone loss could also be excluded because bone resorption was not ‘deforming’ (ostitis deformans Paget) and the bone volume was preserved. There was also only borderline elevation of alkaline phosphatase.

Vanishing bone disease (Gorham’s disease) is a rare form of angiomatosis which destroys the affected bone more or less completely. This diagnosis is not consistent with the appearance of the demineralized bone, the shape of which is perfectly preserved.

Metastasis of an unknown primary is extremely unlikely, because metastases would destroy (and not only demineralize) the bone structure.

So a more intelligent approach was required. The simplest approach is the most productive one, i.e. to talk to the patient. The patient had worked in a coal power plant from 1977 to 1982. The degree of filling of the coal containers was monitored using a radioactive cobalt source. It was the task of the patient to monitor this device. Because this was convenient for him, although it was forbidden, he often exposed his occiput to irradiation by moving through the cobalt-gamma-beam. We calculated a cumulative dose of 0.2–2 Gy to the occiput. This dose of radiation is sufficient to increase vascularization and perfusion, but not sufficient to cause necrosis, of the bone. The patient had no external sign of radiation damage, e.g. hair loss, trophic changes of the skin etc. Increased perfusion of the occiput induced massive bone resorption, presumably aggravating resorptive bone loss via parathyroid hormone. This hypothesis would explain the circumscribed bone loss in the exposed compared to the non-irradiated adjacent bone of the skull (Figure 3). The rest of the skull showed low grade, but unequivocal, osteitis fibrosa with circumscribed osteolytic lesions (‘pepper pot skull’).

After parathyroidectomy, a follow-up X-ray of the skull (Figure 4) showed complete normalization of the mineralization of the skull with the exception of the occiput which was hypermineralized.

So in summary, the occiput, having been exposed to radiation injury in the past, underwent excessive resorption and excessive remineralization in response to secondary renal hyperparathyroidism and parathyroidectomy, respectively.

Suggested reading


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