CASE REPORT

A rare case describing catastrophic destruction of the whole skeleton caused by severe hyperparathyroidism

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SUMMARY

Osteitis fibrosa cystica is a skeletal disorder seen in advanced stages of persistent hyperparathyroidism. Although the measurement of serum Ca and intact-parathormone levels provides early diagnosis and decreases the incidence of radiographic bone involvement, progressive major bone lesions may still be seen in developing countries even in the modern era. We aimed to share, by writing up this report, our astonishment after observing how the skeletal system can be ruined by persistent hyperparathyroidism.

BACKGROUND

Brown tumours (BT) are rare non-neoplastic lesions resulting from abnormal bone metabolism in advanced stages of hyperparathyroidism and represent the end-stage of bone-remodelling. Although the measurement of serum Ca and intact-parathormone (PTH) levels provides early diagnosis and decreases the incidence of radiographic bone involvement, progressive major bone lesions may still be seen in developing countries even in the modern era. In this report, we present interesting scintigraphic and radiological findings of a patient with multiple expansile bone lesions causing serious deformities throughout the skeleton, due to hyperparathyroidism, which was previously misdiagnosed as giant cell tumour (GCT).

CASE PRESENTATION

A 48-year-old woman was diagnosed as having a GCT on her mandible and was referred for bone scintigraphy to evaluate the extent of her disease. On bone scan, increased and patchy Tc99m-methylene diphosphonate (MDP) uptake suggestive of expansile, mixed lytic-osteoblastic bone lesions was seen throughout the skeleton, most prominently in the appendicular skeleton (figure 1). Serious deformities in bilateral humerus and bilateral femurs were noted. Focal intense MDP uptake was clearly seen in the mandible from where the histological specimen was obtained. In the axial skeleton, multiple sites of heterogeneously increased MDP uptake were seen in the ribs, costochondral junctions and in the pelvis. On X-rays of the long and flat bones, multiple expansile and multiloculated lytic lesions covering the entire bone were noted (figure 2).

We considered the bone scan and radiological findings as inconsistent with GCT and its

metastasis. On the differential diagnosis, we considered that these widespread expansile bone lesions may have been related to a systemic and/or a metabolic bone disorder. Questioning the patient revealed a medical history of a mediastinal parathyroid adenoma, 2.5 cm in size, which was treated with surgical excision 4 years prior. Serum calcium, alkaline phosphatase (ALP) and PTH levels were measured and found to be significantly elevated (12.5 mg/dL, 987 U/L and 276 pg/mL, respectively). The serum vitamin D level was within normal range and serum phosphate value was slightly decreased (2.1 mg/dL). The patient had no biochemical or clinical signs of renal failure; serum creatinine and urea levels were completely normal. To the best of our knowledge, the radiological appearance and the histopathological findings of GCT may both closely mimic BT.¹ Therefore, given the radiological appearance of the lesions, along with the bone scan findings and laboratory results,

Figure 1 Anterior and posterior whole body scan of Tc99m methylene diphosphonate (MDP) bone scintigraphy demonstrating intense and heterogeneous MDP uptake throughout the whole skeleton, well matched with the expansile bone lesions. The scintigraphic findings are interesting and exaggerated.

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Figure 2 (A) Anteroposterior X-ray of the pelvis and proximal femurs showing multiple expansile and multiloculated lytic lesions of the entire proximal femurs and pelvic bones. Loss of normal anatomical structure of both femurs and deformities at the middle femoral shafts are demonstrable. Lesions are expansile but not destructive. (B) Anteroposterior X-ray of the right humerus and shoulder showing expansile and multiloculated lytic lesions of the entire humerus, proximal radius and ulna, similar to those of the femoral and pelvic lesions. Glenoid bone is also affected. Deformities at the proximal ulna and radius are prominent.

we considered the condition to be related to an advanced stage of hyperparathyroidism with osteitis fibrosa cystica (OFC) and multiple BTs.

OUTCOME AND FOLLOW-UP

The parathyroid adenoma could not be identified in this patient and she is being continued on medical treatment.

DISCUSSION

OFC is a skeletal disorder seen in the advanced stage of persistent hyperparathyroidism.² BT is an extremely rare osseous lesion that constitutes a focal manifestation of OFC. These tumours are made up of mononuclear stromal cells mixed with osteoclasts, multinucleated giant cells and fibrous tissue collection, and represent the end stage of a bone-remodelling condition.³ As radiological and histopathological appearances are very similar to GCTs, differential diagnosis of BT from GCT generally depends on clinical and laboratory findings.⁴ The osteoclastic activity and bone breakdown is increased by elevated secretion of PTH. Therefore, destructive bone lesions and BTs occur only in the more advanced stages of hyperparathyroid bone disease.⁵

Progression of focal bone resorption and fibrosis can result in macroscopically visible cysts, osseous expansion and pathological fractures. However, due to their ample facilities for measuring serum Ca and intact PTH levels, gross bone lesions have become rare in industrialised countries. Therefore, radiographic bone involvement of patients with hyperparathyroidism significantly decreased in the 1990s.⁶ However, in developing countries, even today, some patients may not be referred to hospitals until significant bone destruction occurs, as seen in our case.

OFC may also be seen in patients with parathyroid carcinoma having a physiopathological mechanism similar to that seen in primary hyperparathyroidism. However, the histopathology result that was obtained from the mediastinal mass excluded parathyroid cancer and was positive for parathyroid adenoma. It must be remembered that end-stage renal disease may also lead to severe bony destruction, nevertheless, renal function was normal in this case, which excluded the diagnosis of renal osteodystrophy and Sagliker syndrome. In the clinical differential diagnosis of patients with bone dysplasias, other common diseases such as Paget's disease, McCune

Albright syndrome and osteomalacia must also be kept in mind by clinicians. In our patient, the findings of bone scintigraphy were helpful to exclude the diagnosis of Paget's disease and osteomalacia. In addition, the patient's medical history of parathyroid adenoma excision, high serum calcium, ALP and PTH levels, normal vitamin D levels and normal renal functions, supported the diagnosis of OFC in our patient.

In the present case, we aimed to share our astonishment on interpreting the exaggerated findings of the X-rays and bone scan. We consider that this case provides a unique opportunity to observe how the skeletal system can be ruined by persistent hyperparathyroidism.

Learning points

- ▶ Osteitis fibrosa cystica is a skeletal disorder seen in the advanced stage of persistent hyperparathyroidism.
- ▶ Brown tumours are rare non-neoplastic lesions resulting from abnormal bone metabolism in advanced stages of hyperparathyroidism.
- ▶ The radiological appearance and the histopathological findings of giant cell tumour may both closely mimic brown tumour.

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Competing interests None declared.

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