'Rachitic rosary sign' and 'tie sign' of the sternum in tumour-induced osteomalacia

Partha Pratim Chakraborty,¹ Rana Bhattacharjee,² Satinath Mukhopadhyay,² Subhankar Chowdhury²

¹Department of Medicine, Midnapore Medical College & Hospital, Midnapore, West Bengal, India

²Department of Endocrinology & Metabolism, Institute of Postgraduate Medical Education & Research/SSKM Hospital, Kolkata, West Bengal, India

Correspondence to

Dr Partha Pratim Chakraborty, docparthapc@yahoo.co.in

Accepted 8 February 2016

DESCRIPTION

A 40-year-old woman presented after 3 years of progressively increasing proximal muscle weakness, severe myalgia and generalised bone pain. Clinical examination revealed severe proximal muscle weakness (power grade 3/5), diffuse bony tenderness and kyphoscoliosis. Baseline biochemistry revealed low serum phosphate (1.5 mg/dL) and elevated alkaline phosphatase (552 IU/L (reference: 98–251 IU/L)) with normal serum calcium (corrected calcium 9.2 mg/dL), creatinine (0.5 mg/dL), 25-hydroxy-vitamin D (34 ng/mL), normal calcitriol (32 pg/mL (reference: 19.6–54.3 pg/mL)) and parathyroid hormone (55 pg/mL (reference: 19.6–54.3 pg/mL)). Urinary concentrations of creatinine and phosphate in a timed sample were 27.7 mg/dL, 10.4 mg/dL, respectively. Tubular maximum for phosphate (TmP), corrected for glomerular filtration rate (GFR) (TmP/GFR) was 1.3 mg/dL (normal 2.8–4.4 mg/dL). Twenty-four hour urinary calcium excretion was 152 mg (2.9 mg/kg/day). Plasma fibroblast growth factor 23 (FGF23), measured using a C-terminal ELISA, was 603 relative units/mL (reference: <150 RU/mL). A working diagnosis of oncogenic osteomalacia was considered but whole body fluorine-18 fluorodeoxyglucose positron emission tomography (PET)/CT scan and whole body MRI failed to localise any suggestive lesion. Ga-68Dotatate PET/CT was planned and the patient was put on a gradually increasing dose of oral sodium acid phosphate (in four divided dosages) and calcitriol (in two divided dosages). With a total daily dose of 3 g of elemental phosphate (57 mg/kg), which she could tolerate and 2 µg of calcitriol (38 ng/kg), she achieved significant symptomatic improvement with a serum phosphate level of 2.2 mg/dL (measured before the second dose of the day) and parathyroid hormone level of 60 pg/mL. Because of generalised bone pain, she had been subjected to a technetium 99m methylene diphosphonate whole body bone scan by her primary care physician prior to her presentation to us. The bone scan demonstrated increased radiotracer uptake over the maxillae, sternum (figure 1; white arrow), vertebrae, pelvic bones and both humeri. Interestingly, there were prominent costochondral beadings (figure 2; black arrow) and increased tracer uptake over growth plates around the knee joints (figure 1; solid black arrow).

Tumour-induced osteomalacia (TIO), also known as oncogenic osteomalacia, is a less frequently encountered form of metabolic bone disease, in which small tumours of mesenchymal origin secrete one or more phosphaturic substances or phosphatonins that inhibit reabsorption of

Figure 1 Tie sign of the sternum (white arrow) and accumulation of tracer over growth plates around the knee joints (black solid arrow).

phosphate from the proximal renal tubule and result in abnormal phosphate homeostasis. Patients usually present with muscle weakness, myalgia and severe generalised bone pain of long duration, which at times render them bed bound.

A number of imaging modalities, functional and structural, have been used to identify the culprit lesion in oncogenic osteomalacia, with varying sensitivity and specificity. Bone scan, though highly sensitive for detecting skeletal metastases, is of limited use in localising TIO-related tumours. A whole body bone scan is advised at times to establish the cause of bone pain if a diagnosis of TIO is not considered by the treating physician. It frequently shows uptakes over areas of fractures and thus may suggest multiple metastatic bone lesions.

CrossMark

To cite: Chakraborty PP, Bhattacharjee R, Mukhopadhyay S, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2016-214766

Figure 2 Uptake of tracer over costochondral junctions creating the 'rachitic rosary sign' (black arrow).

However, bone scans in TIO at times may demonstrate some typical findings such as the 'rachitic rosary sign', 'tie sign' of the sternum and prominent epiphyses of the large joints particularly the knees.¹ The uptakes at the costochondral junctions and areas of the bone in skeletally matured adults where the growth plates were previously located may represent a form of 'pseudo-reactivation' of growth plates in the adult skeleton.² Though the pathophysiological mechanism of this interesting

radiological observation is yet to be identified, it perhaps suggests a non-specific phenomenon of metabolic bone disorders.³

Learning points

- ▶ The 'rachitic rosary sign', 'tie sign' of the sternum and tracer uptake over areas of previous growth plates are typical findings on bone scan in patients of tumour-induced osteomalacia (TIO).
- ▶ Bone scan, if advised inadvertently in unsuspected patients of TIO, can be helpful to diagnose underlying metabolic bone disease.
- ▶ Tracer uptake over multiple areas should not be misinterpreted as metastatic tumour if a bone scan is advised in TIO.

Contributors PPC and RB were involved in diagnosis, literature search and preparing the manuscript. SM and SC were involved in diagnosis, providing intellectual inputs and editing the manuscript.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Rai GS, Webster SG, Wright EP. Isotopic scanning of bone in the diagnosis of osteomalacia. *J Am Geriatr Soc* 1981;29:45–8.
- 2 Chong WH, Molinolo AA, Chen CC, *et al.* Tumor-induced osteomalacia. *Endocr Relat Cancer* 2011;18:R53–77.
- 3 Sood A, Agarwal K, Shukla J, *et al.* Bone scintigraphic patterns in patients of tumor induced osteomalacia. *Indian J Nucl Med* 2013;28:173–5.

Copyright 2016 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow