JBR–BTR, 2012, 95: 80-82.

PRIMARY HYPERPARATHYROIDISM PRESENTING WITH BILATERAL SLIPPED CAPITAL FEMUR EPIPHYSIOLYSIS. A PICTORIAL ESSAY

T. De Beule[1], L. Ardies[1], Ph. Simons[1], P. Gillardin[2], P. Vanhoenacker[1]

It has become rare to find the typical radiological manifestations of primary hyperparathyroidism like generalized osteoporosis, brown tumors and advanced bone resorption because of the generalized usage of biochemical screening techniques. We present a 17-year-old patient with a parathyroid adenoma resulting in these typical skeletal manifestation throughout the skeleton combined with secondary bilateral slipped capital femur epiphysiolysis.

Key-word: Parathyroid, hyperparathyroidism.

Primary hyperparathyroidism is a rare condition caused by parathyroid adenoma, carcinoma or parathyroid gland hyperplasia. This condition is characterised by the elevated serum concentration of parathyroid hormone resulting in bone demineralisation throughout the whole skeleton and secondary hypercalcaemia. A generalized osteoporosis occurs if the disease is not treated in an early stage. These advanced signs of bone resorption are only seldom seen these days because of the widespread availability of diagnostic tests. We present a case of a young child with a delayed diagnosis of the disease, showing almost all the bony changes. A bilateral slipped capital femur epiphysiolysis (SCFE) was diagnosed as a complication of the bone resorption. Only five cases of SCFE in combination with primary hyperparathyroidism are reported in the literature to our knowledge.

Case report

We present a case of a 17-year-old patient with a long history of gait problems and a postural tension problem combined with symptoms of hyperflexia. Standard conventional radiographs were taken during a long diagnostic quest. These showed irregular translucent areas in the metaphysic bone adjacent to the growth plate (Fig. 1), an erosive delineation of the sacroiliac joints and the symphisis pubis (Fig. 2, 3), trabecular bone resorption throughout the skeleton (Fig. 4) and subligamentous bone resorption at tendon insertions (Fig. 5). A non-sclerotic lesion in the proximal tibia was found without any periosteal reaction (Fig. 6), this lesion fitted the description of a brown tumour. CT scan revealed multiple other brown tumours (Fig. 7). Due to the secondary effects of this global bone resorption a rather rare secondary bilateral SCFE had developed (Fig. 8). These combined skeletal manifestations of global bone resorption and brown tumours lead to the assumption of hyperparathyroidism. The culprit lesion consisted of a parathyroid adenoma, which was surgically removed.

Discussion

Primary hyperparathyroidism is a common endocrine disease with a prevalence of 1 in 500 women a year and 1 in 2000 men. The incidence increases with age affecting about 1% of the population. A sporadic form (80%-85%) and a familial form exist. The sporadic form is in general caused by a solitary parathyroid adenoma creating an overproduction of parathyroid hormone (PTH) (1).

Hyperparathyroidism is defined by the risen concentration of PTH, which affects the calcium metabolism. A risen PTH results in a higher bone mineralization turnover caused by an increased activation of osteoclastic cells. This demineralization creates an excess of calcium in the serum. PTH increases the calcium reabsorption at the level of the kidney and promotes the excretion of phosphates. PTH creates an intestinal absorption of calcium due to an overproduction of 1,25-dihydroxycalciferol. Overall, a risen concentration of PTH results in a higher serum calcium level which promotes the formation of kidney stones, osteoporosis with secondary fractures and neuromuscular weakness. These are all symptoms of a long-lasting disease. Due to the non-specificity of the symptoms, diagnostic delay was a common problem. In the early 1970s, biochemical screening became available creating the possibility for an early detection of the disorder without overt clinical manifestations. A marked decrease of symptomatic hyperparathyroid diseases is noted these days (2, 3).

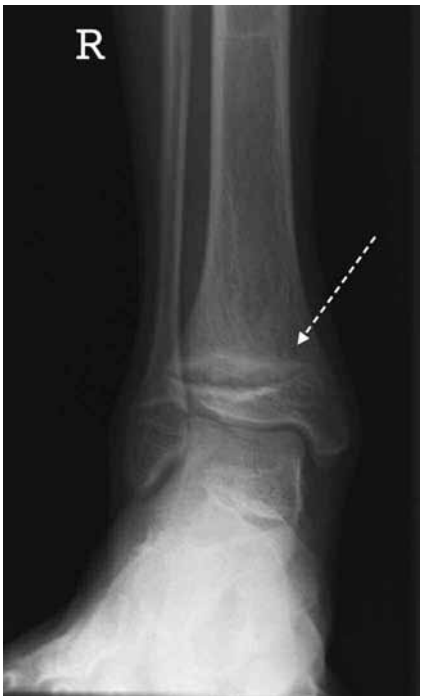

Fig. 1. — Bilateral widening of the growth plate with an irregular sclerotic delineation.

Because of the demineralization effects of PTH on the bone, radiographical changes at multiple locations throughout the skeleton can be noticed in late stage disease.

One of the first possible presentations at imaging is the subphyseal, subchondral, subligamentous and trabecular bone resorption (4).

Irregular translucent areas appear in the metaphysic bone adjacent to the growth plate as a result of subphyseal bone resorption caused by the higher level of serum PTH. This results in a widening of the growth plate with irregular sclerotic mar-

From: 1. Department of Radiology and Imaging, 2. Department of Pediatry, OLV Ziekenhuis, Aalst, Belgium.
Address for correspondence: Dr T. De Beule, M.D., Dept of Radiology, Olv Ziekenhuis, Moorselbaan 164, 9300 Aalst, Belgium. E-mail: Tom.debeule@gmail.com

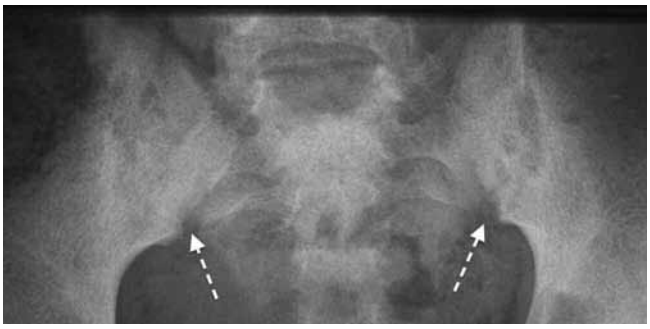

Fig. 2. — Erosive delineation of the sacro-iliacal joints

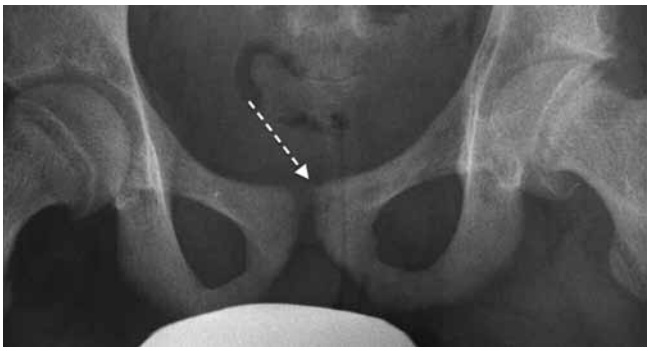

Fig. 3. — Erosive delineation of the symphisis pubis

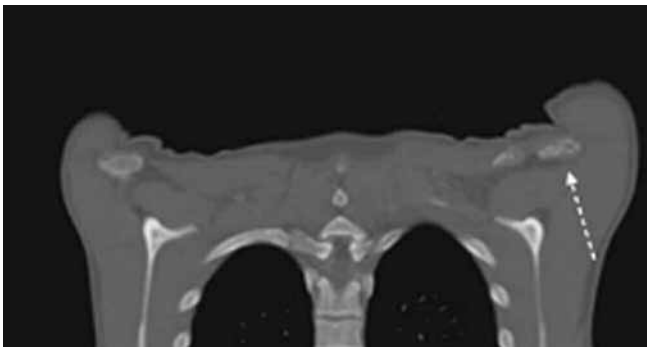

Fig. 5. — Osseous resorption occurs at the sites of tendon and ligament attachment to bone and is mostly seen at the distal ends of the clavicula, trochanters, ischial tuberosties and humeral tuberosities.

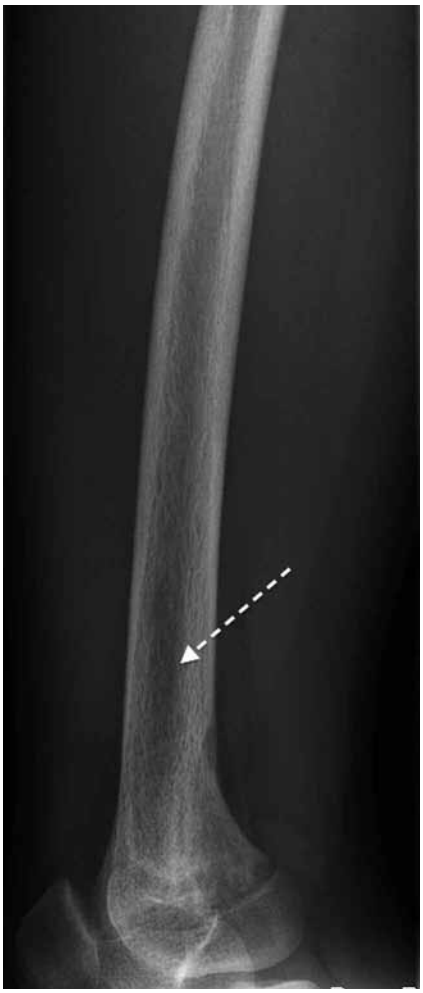

Fig. 4. — Trabecular bone resorption resulting in diffuse trabecular bone pattern. This resorption can occur throughout the skeleton.

gins. Subchondral bone resorption is most frequently seen in the sacro-iliac, sternoclavicular and acromio-clavicular joints, symphisis pubis, and the discovertebral junctions. This bone resorption is characterised by an erosive delineation located next to the articular surfaces. A third sign is the trabecular bone resorption throughout the skeleton.

In the cranium, this resorption results in an osteopenic and speckled appearance represented by the multiple tiny hyperlucent areas throughout the skull vault. This sign is also known as the salt and pepper appearance or pepper pot skull (5). Subligamentous bone resorption can also be depicted at the sites of tendon and ligament attachment to bone (6). This phenomenon can be seen at the distal ends of the clavicula, trochanters, ischial tuberosties and humeral tuberosities.

Another finding due to this excess of osteoclast activity are is a brown tumor. In locations with particularly rapid octeoclastic activity, hemorrhage, reparative granulation and fibrous tissue can replace the normal bone. These localized accumulations of fibrous tissue and giant cells may produce osseous expansion. Brown tumors appear as single or multiple well-defined lesions of the axial or appendicular skeleton, without sclerosis on conventional radiography (7). The lesions may be in an eccentric or cortical location. Computed tomography discloses a tissue mass that enhances after contrast injection but does not invade the soft tissues; in addition, there is no periosteal reaction.

Brown tumours have a higher prevalence in association with secondary hyperparathyroidism and are more rarely associated with primary hyperparathyroidism (8).

The effect of parathyroidectomy on brown tumors depends on their composition (9). Ossification can be expected if the cortex is blown out by a tissue mass that is reddish-brown as a result of interstitial bleeding. Fluid-filled masses, in contrast, tend to persist as cysts filled with hemorrhagic fluid. The main differential diagnosis is a giant-cell tumour, which is a highly vascular lesion located in the metaphysis or epiphysis of a limb bone or in the pelvis, sacrum, or spine.

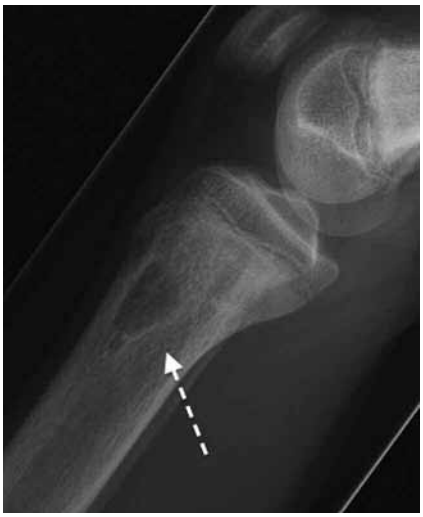

Fig. 6. — Non-sclerotic, lytic lesion anteriorly localized in the proximal right tibia without a periosteal reaction.

Slipped capital femur epiphysis is another secondary finding due to the bone resorption around the growth plate of the femur capital. SCFE caused by hyperparathyroidism or other endocrine imbalances is well known and correlated (10). Trethowen's sign can be used to detect slippage of the femur capital, which is positve when Klein's line no longer transects the lateral capital epiphysis. SCFE in case of primary hyperparathyroidism is a rare condition and to our knowledge only described in five case reports (11).

Conclusion

Excessive production of parathyroid hormone, termed hyperparathyroidism, is classified as primary, secondary, or tertiary in form. Primary hyperparathyroidism, due to autonomous hypersecretion of PTH, usually occurs in the setting of a parathyroid adenoma (80%), which is rather rare in children.

Bone demineralisation throughout the skeleton can only be detected with conventional radiographs after a prolonged risen concentration of PTH. The excessive osteoclastic activity results in subperiosteal, endosteal, subchondral, trabecular, subphyseal and subligamentous and subtendinous bone resorption. These changes can be helpful in detecting asymptomatic lesions although biochemical tests are more sensitive.

Brown tumours can occur because of a local higher osteoclastic activity and has to be differentiated from giant cell tumour, which are highly vascular compared to brown tumours.

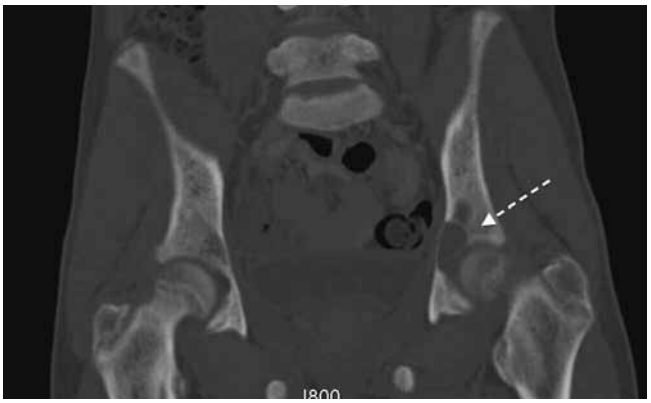

Fig. 7. — Brown tumor in the acetabular roof presented as a sharp demarcated lytic lesion with strong thinning of the surrounding cortex around the lytic lesion.

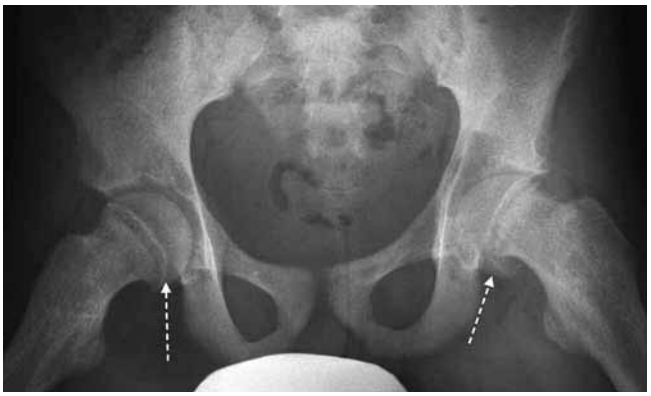

Fig. 8. — Bilateral slipped capital femur epiphysis

Because primary hyperparathyroidism is regularly diagnosed earlier in the course of the disease, nowadays it is only rarely seen that the bony resorption occurs so manifest throughout the skeleton. It is important to keep the possibility of a SCFE in mind when young patients present with gait problems combined with hyperparathyroidism.

Acknowledgements

Special thanks to Dr. L. Lateur Dept. Radiology UZ Gasthuisberg who provided the case material.

References

1. Elaraj D., Clark O.: Current status and treatment of primary hyperparathyroidism. *The Permanent Journal*, 2008, 12: 32-37.
2. Bilezikian J.P., Silverberg S.J.: Asymptomatic Primary Hyperparathyroidism. *N Engl J Med*, 2004, 350: 1746-1751.
3. McDonald D.K., Parman L., Speights V.O.: Primary Hyperparathyroidism Due to Parathyroid Adenoma. *Radiographics*, 2005, 25: 829-834.
4. Resnick D.: Resnick, Bone and joint imaging second edition Ch 52: p 552-561.
5. Roche C.J., et al.: Selections from the Buffet of Food Signs in Radiology. *Radiographics*, 2002, 22: 1369-1384.
6. Teplick J.G., Eftekhari F., Haskin M.E.: Erosion of the sternal ends of the clavicles. A new sign of primary and secondary hyperparathyroidism. *Radiology*, 1974, 113: 323.
7. Merz M.N., Massich D.D., Marsh W., Schuller D.E.: Hyperparathyroidism Presenting as Brown Tumor of the Maxilla. *Am J Otolaryngol*, 2002, 23: 173-176.
8. Jouan A., Zabraniecki L., Vincent V., Poix E., Fournie B.: An unusual presentation of primary hyperparathyroidism: Severe hypercalcemia and multiple brown tumors. *Joint Bone Spine*, 2008, 75: 209-211.
9. Franco M., Bendini J.C., Albano L., et al.: Radiographic follow-up of a phalangeal brown tumor. *Joint Bone Spine*, 2002, 69: 506-510.
10. Qada L., et al.: Slipped Capital Femoral Epiphysis Associated with Primary Hyperparathyroidism and Severe Hypercalcemia. *Clin Pediatr*, 2003, 42: 43.
11. Loder R.T., Hensinger R.N.: Slipped capital femoral epiphysis associated with renal failure osteodystrophy. *J Pediatr Orthop*, 1997, 17: 205-211.