CASE REPORT

Extreme bony pelvic deformity in a renal transplant patient

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SUMMARY

A 35-year-old female renal transplant recipient was referred to the metabolic bone clinic because of a 15 cm loss of height. She was noted to have thoracic kyphosis and vertebral X-ray confirmed a wedge fracture of the eighth thoracic vertebra. She was managed accordingly but on subsequent follow-up mentioned in passing that her gynaecologist had been unable to obtain a cervical smear for routine screening. This led to further questioning, and the patient reported intermittent urinary stress incontinence, feelings of vaginal fullness and severe dyspareunia. It became apparent that these symptoms had been ongoing for 4 years, but the patient had not brought them to the attention of a healthcare practitioner due to feelings of embarrassment and her religious beliefs. These complaints prompted radiological investigation, which revealed extensive bony pelvic deformity, thought to be an extreme manifestation of chronic kidney disease mineral and bone disorder.

encountering the skeletal and extra-skeletal consequences of CKD-MBD. This is achieved primarily through prevention and early treatment of SHPT by ensuring normal levels of calcium, phosphate and vitamin D. Available therapy for SHPT includes phosphate binders, vitamin D receptor activators, calcimimetics and parathyroidectomy.⁴ Initiated promptly, when indicated, these therapies can keep mineral imbalance to a minimum and reduce the risk of the complications described. Having said this, however, crucial to the success of such treatments is a high level of patient concordance.

CASE PRESENTATION

A 35-year-old female renal transplant recipient was referred to the metabolic bone clinic because of a 15 cm loss of height. She was noted to have thoracic kyphosis and vertebral X-ray confirmed a wedge fracture of the eighth thoracic vertebra. Her bone mineral density scan showed a T-score of -1.1 in the lumbar spine and -1.3 in the hip. In addition, her bone resorption and formation markers were suppressed, indicative of an adynamic bone disease.

She was managed conservatively but on subsequent follow-up, as a passing comment, mentioned that her gynaecologist had been unable to obtain a cervical smear for routine screening. This led to further questioning, and the patient reporting a history of intermittent urinary stress incontinence, feelings of vaginal fullness and an inability to participate in penetrative vaginal intercourse due to discomfort. It became apparent that these symptoms had been ongoing for 4 years, but the patient had not brought them to the attention of a healthcare professional due to feelings of embarrassment and her strong religious beliefs. This prompted radiological investigation, which revealed the extensive pelvic bony deformity seen in the accompanying images (figures 1 and 2).

There was no history of congenital disorder or pelvic trauma to explain these findings. We could find no previous pelvic imaging prior to those presented here, and the patient did not recall ever previously experiencing any problems that may have been attributable to pelvic deformity before those mentioned above. A mother to three children, our patient has never had a vaginal delivery, having had a Caesarean section for the birth of her first child due to its breech presentation and subsequently opting for Caesareans for both future deliveries.

BACKGROUND

Chronic kidney disease (CKD) is an important clinical entity, which, along with its vast array of complications, presents an ever-increasing burden on healthcare systems, with an adult prevalence of 8%–16% worldwide.¹ As renal function deteriorates and patients progress through the stages of CKD, problems with bone mineral homeostasis often occur; these can be grouped together under the umbrella term chronic kidney disease mineral and bone disorder (CKD-MBD), formerly known as renal osteodystrophy.

The characteristic biochemical profile of CKD-MBD involves fibroblast growth factor 23 and phosphate elevation, a reduction in serum calcitriol and calcium levels and an increase in parathyroid hormone (PTH) concentrations, resulting in altered bone turnover, vascular calcification and left ventricular hypertrophy.^{2,3} From a skeletal perspective, high-turnover and low-turnover forms of renal bone disease are typically distinguished,² with the former, involving the effects of chronic secondary hyperparathyroidism (SHPT), most likely to have been responsible for the clinical picture described in this case. Furthermore, when compared with the general population, patients with end-stage renal disease (ESRD) have a twofold to fourfold increase in fracture risk.³

Through several effective medical interventions, it is now possible to significantly reduce the risk of

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Figure 1 Anteroposterior radiograph of the pelvis showing a chronically deformed and remodelled anterior pelvis and pelvic outlet (arrows), due to serial insufficiency fractures and remodelling over time secondary to chronic kidney disease mineral and bone disorder.

The patient's background included IgA nephropathy diagnosed in 2005, while she was carrying her third child. This rapidly progressed to ESRD, which was initially managed with thrice-weekly haemodialysis for 7 years prior to cadaveric renal transplant in February 2013. Our patient's long-standing time in ESRD, on dialysis, was marked by persistent non-concordance with agreed treatment regimens and chronically poor attendance at clinic appointments and dialysis sessions. As a direct result of this, the patient developed complications of chronic renal failure including SHPT, with protracted periods of markedly deranged serum calcium, phosphate and 25-hydroxy vitamin D levels.

Our patient was managed for many years by an expert team in a tertiary care setting, in accordance with clinical guidelines and accepted standard practice. In light of her worsening parathyroid status despite medical therapies and with the patient initially refusing surgery, she started on a trial of the calcimimetic agent Cinacalcet (30mg once daily) in January 2011. However, this drug still failed to alleviate her symptoms (diffuse bone pains) and normalise PTH and calcium/phosphate status,

and as a result, despite early reluctance, the patient eventually underwent parathyroidectomy to definitively treat her SHPT in October 2012.

INVESTIGATIONS

Biochemical evidence of CKD-MBD was present over the 7-year period leading up to parathyroidectomy and renal transplant. The patient was persistently hypocalcaemic from the outset (serum total calcium value ranging from 1.36 to 2.40 mmol/L (normal range 2.20–2.60 mmol/L)). Serum PTH level (normal range 10–65 ng/L) rose from the lowest value available (197 ng/L, September 2005) to 1807 ng/L (September 2009) before all readings after December 2010 prior to parathyroidectomy came back >2500 ng/L. Alkaline phosphatase (ALP, normal range 30–130 U/L) was also raised over this period (range 380–793 U/L), in keeping with a high bone turnover state. All serum biochemistry including calcium, phosphate, vitamin D and ALP was seen to normalise after renal transplant.

As regards the pelvic deformity, workup included flexible cystoscopy, which revealed introital obstruction. After this, plain film X-ray and CT scans (figures 1 and 2) were performed to enable characterisation of the bony deformity. Dual-energy X-ray absorptiometry scanning was also performed in light of the vertebral insufficiency fracture seen.

DIFFERENTIAL DIAGNOSIS

1. CKD-MBD including osteomalacia.
2. Bone disorders after renal transplantation—bone and mineral disorders occur frequently in renal transplant recipients, partly due to the effects of immunosuppressive medications such as corticosteroids.
3. Traumatic pelvic bony injury—can be excluded from the history.

OUTCOME AND FOLLOW-UP

Despite undergoing a renal transplant to treat her ESRD and the apparent normalisation of bone metabolism as a result of this, the severity of the effects of CKD-MBD on the bony architecture of our patient's pelvis has caused significant distress and had a detrimental impact on her overall quality of life. Orthopaedic opinion was sought both at the local orthopaedic hospital and at a specialised pelvic deformity and reconstruction unit in London. While the patient was assessed with a view to possible corrective surgery, she has, however, been advised against any intervention. This is due to the extent and complexity of the deformity and the feeling among specialists that any intervention would be inappropriate, with the risks outweighing the benefits.

DISCUSSION

Our case provides a unique example of an extreme manifestation of what is quite a common entity, CKD-MBD, involving severe deformity of the female bony pelvis. To the best of our knowledge and despite an extensive literature search, no other cases of such a substantial skeletal deformity in a CKD or transplant patient have been reported. The extent of the deformity described here is rare owing to the success of modern medicine, presence of good practice guidelines to inform management and generally good, if not excellent, patient adherence to treatment plans.

Pre-existing CKD-MBD can increase the risk of future bone and mineral disorders as well as fractures post-transplant. Subsequent softening of bone due to the effects of a potent immunosuppressant regimen after transplant (in this case prednisolone

Figure 2 Three-dimensional reconstruction of a pelvic CT examination showing bony remodelling of the inferior pubic rami causing pelvic outlet stenosis (A, arrows) and deformity of the pelvic inlet (B, arrows).

(maximum dose: 10mg once daily), tacrolimus and mycophenolate mofetil)) may have also contributed to the reduced bone density and vertebral insufficiency fracture seen and may well have also exacerbated the existing pelvic deformity. However, our patient's symptoms and loss of height were found to predate her renal transplant by quite some time, and as such it is likely that the deformity seen is a consequence of CKD-MBD from her lengthy time with ESRD. Additionally, her bone densitometry scans done 2 years post-transplant have shown an increase in density of 11.6% and 16% in the lumbar spine and hip, respectively. Hence, it is clear that the overall effect of the

immunosuppressant regimen on our patient's pelvic geometry and architecture is likely to be very minimal.

Despite undergoing a renal transplant to treat her ESRD, the severity of the associated CKD-MBD complications has caused significant distress to the patient and has had a detrimental impact on her overall quality of life. The patient was assessed for corrective pelvic surgery, both at the local orthopaedic hospital and a more specialist pelvic reconstruction unit in London. She has, however, been advised against having any corrective intervention due to the complexity of the deformity and the inherent risks of surgery.

Learning points

- Despite definitive treatment of end-stage renal disease with kidney transplant and normalisation of renal function thereafter, patients may still continue to suffer over the long-term from the effects of chronic kidney disease (CKD) complications such as CKD mineral and bone disorder.
- Patient concordance with treatment regimens in CKD is paramount, to minimise the risk of encountering permanent manifestations of disease, which will persist and continue to cause distress even after the CKD is treated.
- The case highlights the significance of clinicians taking seriously all concerns raised by their patients. A remark that a patient makes about a symptom should not simply be dismissed if it is mentioned in passing or if it is unfamiliar and not directly related to the clinician's immediate field of practice.

Contributors AHM and PC saw the patient in clinic on multiple occasions and drafted the manuscript. SC interpreted the imaging findings. TG was responsible for the overall care of the patient. All authors reviewed and finalised the manuscript prior to submission.

Competing interests None declared.

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