Trauma and reconstruction

“Noodle-like” deformity and spontaneous fracture of the lower limbs in uremia-a case report

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ABSTRACT

Renal osteopathy is common in patients with end-stage renal disease. For certain patients, treatment opportunities are lost due to extremely superstition and poor compliance. This article reports a rare case in which the patient developed “noodle-like” deformity of the bilateral lower limbs and spontaneous fracture of bilateral distal end of femur. The purpose of this article is to offer a warning to future patients.

Introduction

Chronic kidney disease (CKD) is complicated by abnormalities in calcium and phosphorus metabolism, which worsen as the disease stage advances. The risk of fractures is higher in CKD patients than in the general population. Doctor-patient cooperation and early intervention can minimize the risk. Even if the disease progresses to secondary hyperparathyroidism (SHPT) that is difficult to control with drugs, prognosis can still be improved via parathyroidectomy or radiofrequency ablation. This report describes an extreme patient who developed a rare “noodle-like” deformity of the bilateral lower limbs and distal femoral fracture due to superstition and poor compliance.

Case presentation

The patient (male, 53 years old, stage 5 CKD) suffered from chronic glomerulonephritis and had undergone hemodialysis for 23 years. He was admitted to the hospital for 4 days because of spontaneous left femoral fracture. At the time of hospital admission, the patient was 130 cm tall weighed 34 kg, cachectic state with normal vital signs and a clear mind. He had to in decubitus position, with pectus, left scoliosis, and “noodle-like” curly bilateral lower limbs. At 3 cm above the right knee joint, the right femur had deformed healed under the skin and protruded approximately 3 cm above the main trunk. Corresponding position of left lower limb had an newly protruded left femur under bruised skin (Fig. 1A). An X-ray showed fracture-dislocation of the left femur (Fig. 1B). The patient was diagnosed with hemodialysis-dependent chronic kidney disease (CKD5HD), SHPT, and left lower femoral fracture.

The patient had only a junior middle school education. He and his family members were extremely superstitious. He often relied on the Yellow Calendar and divination to determine treatment and examination options. An elevated serum level of intact parathyroid hormone (PTH) had been detected for him 18 years ago. Fluctuations ranges of the patient’s calcium and phosphorus metabolic indicators were as follows: serum phosphorus 1.4–2.2 mmol/L; alkaline phosphatase 104–180 U/L; serum calcium 2.44–2.7 mmol/L; iPTH 1200–2900 pg/dl; and serum 25(OH)VitD3, 12–55 ng/dl. The patient received irregular calcitriol pulse therapy (1.5–2 μg). However, he refused to take calcium due to vomiting and bone pain. In addition, he was given calcium carbonate to reduce his phosphorus level and a subcutaneous injection of salmon calcitonin to relieve bone pain. He had begun to develop generalized bone pain and experience progressive aggravation of thoracic and bilateral lower extremity deformities 12 years ago. Eleven years ago, parathyroidectomy was recommended to him but never done because he was afraid of surgery related complication and asked the surgeon complication free guarantee. Eight years ago, he developed a flexion deformity of the bilateral lower limbs, unable to walk and relied on wheelchairs for mobility, gradually became fully bedridden. Three years ago, he experienced spontaneous fracture of the lower femur at 3 cm above the right knee joint (Fig. 1C). He claimed that X-ray examination would causing generalized bone pain and refused relevant assessments. After home care, the right femoral fracture healed with malunion. During the past 12 years, existed X-rays have shown extensive calcification of soft tissues (such as eyeball, cardiovascular, lung, trachea, and brain tissues), severe osteoporosis, pectus carinatum, and left scoliosis. The patient had gradually developed curly noodle-like bilateral lower limbs. Moreover, these conditions had progressively

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worsened (Fig. 1 D and E). His height had gradually decreased from an original value of 170 cm–130 cm. In addition, he had developed complete calcification of his left carpal autologous radial artery-cephalic vein fistula, which nonetheless continued to exhibit good function (Figure F).

Treatments and results

After admission, the patient received multidisciplinary consultation. A joint orthopedic fixator was customized for him. However, the patient abandoned it due to leg pain and discomfort. He was then given only maintenance dialysis, nutritional support, and analgesic treatment. Nursing care was extremely difficult. The patient developed pressure sores on his back. Any slight passive or active activity had the potential to cause secondary injury. On numerous occasions, the soft tissue at the site of left femoral fracture was punctured by the broken bone, a phenomenon accompanied by repeated local hematomas and skin damage. The patient was severely depressed and refused to eat and treatment. He died of extreme exhaustion at home approximately 3 months after the fracture.

Discussion

The ultimate manifestation of calcium and phosphorus metabolism disorder in CKD patients is spontaneous fracture, which occurs frequently at the hip joint but rarely at the distal end of the femur. The Dialysis Outcomes and Practice Patterns Study (DOPPS) showed that dialytic patients with an iPTH level higher than 900 pg/L experienced a significantly elevated risk of fracture compared with patients with an iPTH level of 100–300 pg/L. In addition, the mortality rate was 3.7 times higher among patients who suffered a fracture than among the remaining patients, with a particularly notable difference in the first month after fracture. The patient in this report suffered from progressive aggravation of SHPT for 12 years. If he had followed medical advice to control his calcium, phosphorus, and iPTH levels at an early stage, his subsequent tragedy could have been prevented. Therefore, CKD patients need to use multiple methods to continuously and effectively maintain their blood calcium and phosphorus levels in the normal range and control their iPTH levels at 2–9 times normal, starting as early as the CKD3a stage. Good patient compliance is an important condition for reaching this goal. For the patient described in this report, effective opportunities to control various key indicators were lost due to extreme superstition, resulting in metastatic calcification of multiple organs, vascular calcification, rare “noodle-like” deformity of the bilateral lower extremities, bilateral distal femoral fracture, and eventually death by cachectic. This case provides an important warning, has educational significance, and showed natural development of renal osteopathy.

Conclusion

The patient’s compliance is basic condition to CKD-MBD management. Follow the right medication for calcium-phosphate-iPTH control at early stage could avoid severe consequences like cardiology calcification and bone fracture and finally benefit for survival.

Declaration of competing interest

No conflict.

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Fig. 1. The visual appearance and an X-ray image of the left lower extremity at hospital admission (A, B); the appearance of the spontaneous fracture of the right femur in March 2017(C); thorax deformity, osteoporosis, bone cyst formation, scoliosis deformation, and massive calcification of the lung, trachea, brain tissue calcification (D); thickening and bone density enhancement of the skull, and eyeball and lens calcification (E); full calcification of vascular fistula and reduced ulna/radius cortices (F). (All picture and data used in this paper had permission from the patient’s family).