Chronic renal failure is characterized by phosphate retention, thereby producing hyperphosphatemia. There is a decreased ability of the diseased kidney to synthesize 1,25 dihydroxy vitamin D, the active metabolite of vitamin D. Calcium reabsorption in the intestines is impaired when circulating levels of this active metabolite are low. This leads to a profound hypocalcemia. This hypocalcemia stimulates overproduction of the parathyroid hormone, which develops into secondary hyperparathyroidism with clear cell hyperplasia within all four glands. The increased amount of parathyroid hormone does not effectively promote intestinal or renal tubule reabsorption of calcium because of the absence of vitamin D and the increased phosphate levels. This secondary hyperparathyroidism accounts for the skeletal changes seen in osteitis fibrosa cystica.

Clinical symptoms of bone disease are uncommon, occurring in less than 10% of predialysis patients with advanced renal failure. Radiologic and histologic abnormalities are observed in 85% and 90%, respectively. Symptoms of bone disease are a major cause of morbidity in patients on long-term dialysis. These symptoms consist primarily of bone pain in the extremities. Fractures rarely occur. Joint pain occurs secondary to calcium deposition in the bursa and other periarticular structures.

In chronic renal failure, there is a tendency for extraosseous or metastatic calcifications. Medium-size blood vessels, subcutaneous articular and periarticular tissues, the myocardium, eyes, and lungs are common sites of metastatic calcifications.

Fully developed osteitis fibrosa cystica is uncommon today since hyperparathyroidism is usually treated at a much earlier stage. However, inadequate management of the electrolyte and mineral imbalances that occur in chronic renal failure can result in marked bony abnormalities. Aluminum toxicity in patients with renal failure who are maintained on hemodialysis for a long period often contributes to the skeletal changes in renal osteodystrophy. The sources of aluminum are dialysis solutions prepared from water with an aluminum content and the use of oral aluminum phosphate binders.

Histology

The earliest bony alterations observed consist of demineralization producing an excess of osteoid. Osteoclasts are then activated, increasing bone resorption. This resorption occurs initially at the subperiosteal and endosteal surfaces and is later generalized to the trabeculae. An increased number of odd-shaped osteoclasts are found within scalloped resorption cavities. In this manner, the cortex and trabeculae are weakened. This bone loss is particularly evident as subperiosteal resorption within the phalanges and the distal aspect of the clavicle creating “moth-eaten” surfaces.

As bone resorption increases, microfractures and microhemorrhages occur, yielding hemosiderin-laden macrophages dispersed throughout a delicate fibrous tissue that fills the marrow spaces (osteitis fibrosa). With progression, the bone resorption, microhemorrhages, and fibrosis become more marked and are often accompanied by the formation of macroscopically visible cysts within the fibrous tissue: osteitis fibrosa cystica.

Treatment

The management of renal osteodystrophy involves not only the treatment of the primary disorder of renal failure through dialysis (or renal transplantation) but also by phosphate-level control. This is accomplished with a phosphate-restricted diet and phosphate-binding agents. Calcium carbonate is the...
preferred phosphate-binding agent. Intravenous calcitriol appears to suppress hyperplastic parathyroid cell secretion in chronic uremia. Occasionally, parathyroidectomy is required to control hyperparathyroidism. Vitamin D administration may also be useful.

Case Presentation

A 35-year-old male presented with the chief complaint of a painful, swollen left fifth toe of 2 weeks' duration. The patient did not recall any previous injury to the area. The pain had increased in severity to the point where light touch was intolerable. The patient had a 15-year history of chronic renal failure secondary to Alport’s syndrome. Other significant history included unilateral hearing loss, surgical removal of calcification deposits from the shoulder, and hemodialysis three times weekly.

Radiographs of the left foot revealed multiple radiodense locular formations along the distal interphalangeal joint of the fifth digit (Fig. 1). The patient was scheduled for surgical excision of the calcifications.

During surgery, a copious amount of brown mucous material extruded from the wound. The material was sent for culture and pathologic examination. thinning and discoloration of the subcutaneous tissue was also noted. The bone of the proximal and middle phalanges appeared hemorrhagic and was very friable in nature. The distal two thirds of the proximal phalanx and base of the middle phalanx were resected and sent to pathology. The possibility of subsequent amputation pending pathology results was discussed with the patient.

Culture and sensitivity results on the mucous exudate showed no bacterial growth. Microscopic examination of the bone fragments revealed prominent, reactive, new bone formation and multiple cystic spaces within the bone marrow. The center of the cystic spaces consisted of calcified debris surrounded by scar tissue and prominent, multinucleated giant-cell reaction. The morphologic changes suggested a combination of metastatic calcification and osteitis fibrosa cystica (Fig. 2). There was no evidence of malignancy.

The patient was treated postoperatively for wound care and referred to his primary physician for evaluation and treatment of the underlying secondary hyperparathyroidism. Further surgical intervention was deemed unnecessary.

Conclusion

Osteitis fibrosa cystica is a rare bony complication observed in patients with chronic renal failure. Infection is generally considered on presentation of an acutely swollen joint in a young patient. It may prove beneficial to include the possibility of renal osteodystrophy as a differential diagnosis when facing a renal patient on long-term hemodialysis.
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References