ANSWER TO PHOTO QUIZ (PAGE 173)

A HAEMODIALYSIS PATIENT WITH PROGRESSIVE LEG PAIN

DIAGNOSIS

The history of chronic kidney failure and secondary hyperparathyroidism together with these radiological findings are typical for secondary tumoral calcinosis. A biopsy, performed to exclude a malignancy, revealed non-specific calcified material without malignant-appearing cells. Tumoral calcinosis is a rare benign condition characterised by lobular calcified soft tissue deposits that can manifest around any joint. The hip, elbow and shoulder are the most common locations.[1] It is associated with hyperphosphataemia but the exact aetiology is still being unravelled. Tumoral calcinosis may be classified as a) primary normo-phosphataemic tumoral calcinosis; b) primary hyperphosphataemic tumoral calcinosis, where gene mutations lead to reduced urinary phosphate excretion causing hyperphosphataemia; c) secondary tumoral calcinosis mostly seen in chronic renal failure associated with secondary or tertiary hyperparathyreodism.[2,3]

The diagnosis of tumoral calcinosis in patients with chronic renal failure is mainly made by radiological evaluation.[3] Imaging studies show distinct amorphous, multilobulated and cystic calcifications with fluid-fluid levels ('sedimentation sign') in periarticular locations.[2] Serum biochemical tests (calcium, phosphorus, renal function tests, parathormone level and 1,25-dihydroxy-vitamin D) can help to distinguish between the different forms of tumoral calcinosis and from diseases that can mimic it. A characteristic presentation does not usually require pathological evaluation. When malignancy is suspected, however, a biopsy is needed. Conditions that can mimic tumoral calcinosis are osteochondromatosis, synovial sarcoma and osteosarcoma.[3]

Treatment of tumoral calcinosis in renal failure is often challenging and includes calcium and phosphorus restricted diets, intensive dialysis treatment, phosphate binders and adequate management of secondary or tertiary hyperparathyroidism, tailored to individual cases. When these medical interventions remain unsuccessful, parathyroidectomy may be necessary. Kidney transplantation may be another treatment option, after correction of the hyperparathyroidism. Surgical treatment of the calcifications is generally not recommended in the secondary form while this remains the first choice of treatment for the two primary forms of tumoral calcinosis.[3] The patient was treated with a phosphate and calcium restricted diet and phosphate binders but was incompliant and he rejected parathyroidectomy. The condition of the patient deteriorated and he passed away. Autopsy was refused.

REFERENCES

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