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A rare case report : Erdheim chester disease(ECD) and langerhans cell histiocytosis(LCH) overlap syndrome presenting as Brown tumor in a patient with CKD-hemodialysis.

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Objectives : Secondary HPT (hyperparathyroidism) is usually due to end-stage renal disease (ESRD). hall marker of parathyroid hormone (PTH) excess is increased osteoclastic activity with bone resorption. Chronic kidney disease(CKD) may lead to secondary or tertiary hyperparathyroidism.
Brown tumor of bone, also called osteitis fibrosa cystica is a rare non-neoplastic lesion resulting from abnormal bone metabolism in CKD patients.
ECD and LCH overlap syndrome are extremely rare. ECD is a rare xanthogranulomatous histiocytic disorder, LCH is a proliferative disorder of histiocytes with phenotype similar to dendritic langerhans cells.
We report a rare case of Erdheim chester disease(ECD) and langerhans cell histiocytosis(LCH) overlap syndrome presenting as Brown tumor in a patient with CKD-hemodialysis.

Methods : Pathologic examination by bone biopsy, laboratory examinations, Magnetic resonance imaging(MRI) and WBBS (whole body bone scan) were used to this case.

Results : Fifty-three years old Korean male was admitted with a history of pelvic pain and left lower extremeties radiating pain and fever. He was diagnosed with idiopathic CKD 21 years ago and started hemodialysis. He was diagnosed with HNP three years ago. his pelvic and lower back pain became worse, and fever began to occur. So he had MRI and WBBS under infective spondylitis. Result of tests revealed suspicion of hematogenous intramedullary vasculitis or metastasis of multiple spine and pelvic bone, gluteal myositis. The bone biopsy of the L3 spine body for histologic confirmation was diagnosed as Brown tumor. the serum PTH level was slightly elevated to 111.51 pg/ml, It seemed to show hyperparathyroidism in CKD patients. The patient was discharged with conservative treatment.
Two weeks after discharge, he was hospitalized again for acute pain. Pain control was performed again. At that time, the PTH level was slightly decreased to 66.8 pg/ml.
However, after 2 weeks, the patient was admitted to the emergency room with

abdominal pain, watery diarrhea and was admitted to the hospital with antibiotic treatment under infectious colitis. After hospitalization, diarrhea improved, but the radiation therapy was initiated due to Brown tumor progression. However, there was a suspicion of hidden malignancy in a patient, which was different from progression of brown tumor. on past bone biopsy review, H & E stain (hematoxylin and eosin) showed favorable ECD and LCH overlap syndrome, but not on immunohistochemical stain (IHC). However, we could not completely exclude LCH, and diagnosed as ECD and LCH overlap syndrome.

Conclusions : In this case, a patient diagnosed with brown tumor, commonly seen in patients with chronic kidney disease receiving hemodialysis. However, since then he was diagnosed as very extreme disease - ECD and LCH overlap syndrome - by pathologic review.
This case shows that in CKD patients, there should be a wake-up call for another disease that may be considered simply a Brown tumor. Failure to do so may result in an inaccurate diagnosis, which may result in a delay in proper treatment.

Keywords : Erdheim chester disease, ECD, langerhans cell histiocytosis, LCH, brown tumor, hyperparathyroidism