

Erdheim-Chester Disease with Perirenal Masses Containing Macroscopic Fat Tissue

지방을 함유한 신장 주변 종괴의 형태로 발현한 Erdheim-Chester Disease

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Erdheim-Chester disease is a rare non-Langerhans-cell histiocytosis involving multiple organs. On histological evaluation, lipid-laden histiocyte aggregates in Erdheim-Chester disease is detected, but fat tissue in affected organs is not noted grossly on computed tomography. A 40-year-old man presented with bilateral perirenal masses containing fat tissue. He was diagnosed as perirenal involvement of Erdheim-Chester disease. This report describes a case of Erdheim-Chester disease with perirenal involvement that demonstrates unusual features.

Index terms

Erdheim-Chester Disease
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Kidney
Retroperitoneal Fibrosis

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INTRODUCTION

Erdheim-Chester disease (ECD) is a rare non-Langerhans-cell histiocytosis involving multiple organs. Most previous reports of this disease have described musculoskeletal or central nervous system involvement. Abdominal involvement has been reported in the perirenal space, ureter, or adrenal gland (1, 2). Here we report a case of ECD with perirenal involvement that demonstrates unusual features with fat tissue on abdominal CT image.

CASE REPORT

A 40-year-old man presented with dyspnea for one week. He had mild mental retardation and diabetes insipidus. Physical examination and laboratory results were unremarkable. He underwent abdominal CT, chest CT, and positron emission tomography (PET)/CT. The abdominal CT revealed bilateral perirenal masses with bilateral hydronephrosis (Fig. 1A, B). The perirenal

masses were first discovered five years earlier on chest CT due to pulmonary tuberculosis. There had been no interval change in the masses since that time. The perirenal masses surrounded both kidneys and even invaded the lower pole of the left kidney. The masses did not compress the kidneys. Fat planes between the kidney and the mass were partially preserved except at the lower pole of the left kidney. Bilateral renal hila were spared. There was no displacement of either kidney or mass effect on adjacent organs. The perirenal masses showed lobulated contours with small amounts of multifocal fat tissue. Intralesional fat tissue demonstrated a comb-like appearance with linear shape perpendicular to the kidney surface. After contrast injection, the masses were poorly enhanced with only subtle enhancement in the periphery. The differential diagnosis for these perirenal masses included retroperitoneal fibrosis, lymphoma, liposarcoma, metastasis, and ECD. However, the perirenal masses contained fat tissue, which restricted the differential diagnosis. Fused PET/CT showed diffuse and moderate fluorodeoxyglucose (FDG) activity (maxi-

maximum standardized uptake value range, 2.2–3.8) in bilateral perirenal masses (Fig. 1C). In addition, some parts of the lung and the pleura on fused PET/CT showed mild patchy FDG activity (Fig. 1D). On chest CT, multifocal ground glass opacities with interlobular and intralobular septal thickening and multifocal thickening of the right pleura were noted in both lungs, suggestive of interstitial fibrosis (Fig. 1E).

CT-guided gun biopsy with an 18-G core needle was performed to further evaluate the perirenal masses. On histological evaluation with hematoxylin and eosin staining, fibrous tissue

proliferation with lipid-laden histiocyte aggregates was detected (Fig. 1F). On immunohistochemical staining, CD68 was positive but CD1a and S-100 (markers of dendritic cells) were negative (Fig. 1G-I). These results were consistent with ECD. Wedge resection of the right upper and lower lung lobes was performed. The lung specimen showed subpleural and interstitial fibrosis with mild chronic inflammation and proliferation of smooth muscle and bronchiolar epithelium, consistent with interstitial pneumonia. On immunohistochemistry of the lung specimen, CD68 was positive.

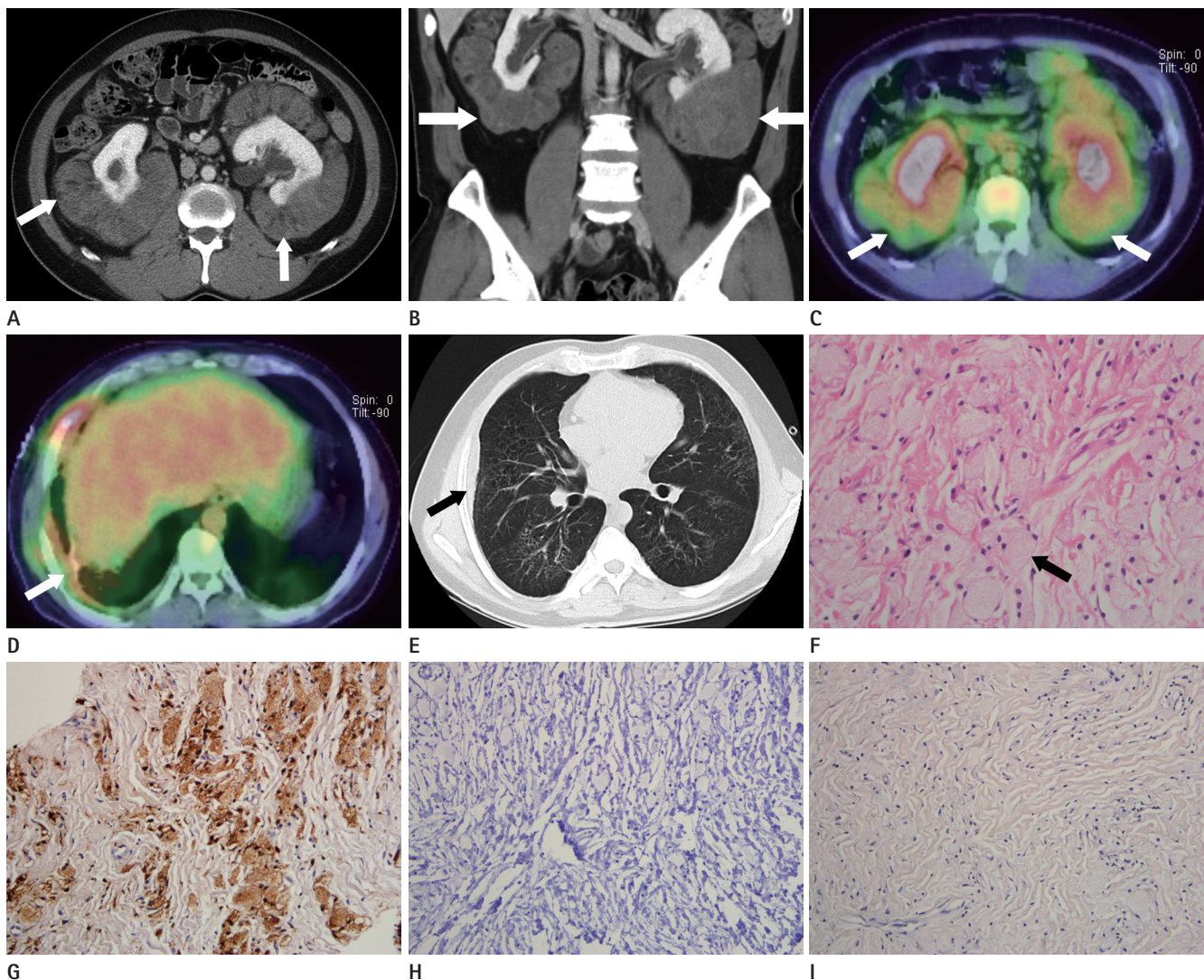


Fig. 1. A 40-year-old man with Erdheim-Chester disease. Bilateral perirenal masses with faint fat tissue (white arrows) are revealed on axial images (A) and coronal image of abdominal CT (B). Fused positron emission tomography (PET)/CT shows diffuse and moderate fluorodeoxyglucose (FDG) activity (maximum standardized uptake value range, 2.2–3.8) in bilateral perirenal masses (white arrows) (C). Some parts of lung and pleura (white arrow) also show mild patchy FDG activity on fused PET/CT (D). Multifocal ground glass opacities with interlobular and intralobular septal thickening and thickening of the right pleura (black arrow) are noted on chest CT (E). H&E stain ($\times 400$) indicates lipid-laden histiocyte aggregation (black arrow for one among many lipid-laden histiocyte) (F). Immunohistochemical stains show that CD68 ($\times 200$) (G) is positive and CD1a ($\times 200$) (H) and S-100 ($\times 200$) (I) are negative.

After glucocorticoids administration for symptom control, dyspnea was relieved. During follow-up, he underwent recurrent urinary tract infection due to vesicoureteral reflux. Foley catheter has indwelled due to neuromuscular dysfunction of urinary bladder. At serial follow-up CT for chest and abdomen, the perirenal masses and abnormal findings of lung and pleura showed no significant change.

DISCUSSION

ECD was first described by William Chester in 1930 (3). Erdheim-Chester disease is a rare systemic xanthogranulomatous infiltrative disease in which lipid-laden histiocytes deposit in various organs, including long tubular bones, lung, heart, kidney, retroperitoneum, breast, brain, skin, and orbit (4, 5). It usually affects adults over 40 years with a slight male predominance (3, 5, 6). Radiologic examinations in previous reports have described bilateral and symmetric involvement (1, 2, 6, 7). Patients with ECD frequently have diabetes insipidus and central nervous system involvement (8, 9). The cause of ECD is unknown (1). However, its monoclonal proliferation of histiocytes suggests neoplastic nature (9). Diagnosis of ECD can be established by histological findings of tissue infiltration by foamy histiocytes without cytoplasmic Birbeck granules and positive immunohistochemical staining for CD68 but negative staining for CD1a and S-100 (1). The clinical course of ECD is variable depending on the involved organ and the extent of the disease (3). Treatments for ECD include steroids, chemotherapy, radiation therapy, immunotherapy, and surgery (9).

In this case, our patient with ECD had diabetes insipidus, lung involvement with interstitial lung disease, and perirenal involvement. Long tubular bone involvement occurs frequently in patients with ECD. However, there was no bone involvement in this case. Patients with ECD often have perirenal involvement. Previously, one report described this perirenal involvement as 'hairy kidneys' (1). A review of diseases in the perirenal space classified ECD as one of the rind-like soft-tissue lesions based on the distribution and imaging features (6). In this case, the patient had perirenal involvement with imaging features slightly different from previous reports. First, the perirenal masses contained macroscopic fat tissue. To the best of our knowledge, there have been no other case of ECD with this finding. Histo-

logically, sheets and cords of foamy histiocytes diffusely infiltrate the tissues and viscera with ECD (3). In ECD, perirenal mass formed by lipid-laden macrophages' aggregation proven on histologic examination failed to show fat attenuation at CT. The macroscopic fat tissue within the bilateral perirenal masses may be preexisting perirenal fat engulfed by aggregation of foamy histiocytes rather than aggregated lipid-laden macrophage itself. Second, perirenal masses surrounded both kidneys. However, there was no severe compression of renal parenchyma or ureters. In addition, the fat plane between kidney and perirenal mass was partially preserved. Pelvocalyceal dilatation was thought to be caused by neurogenic bladder and vesicoureteral reflux due to the patient's physical disability inflicted by idiopathic brain atrophic changes rather than ureteral obstruction causing the pelvocalyceal dilatation. In previous reports of ECD, no space was found between the kidneys and the rind-like soft tissue lesion surrounding them (6). Third, perirenal masses in this case had a lobulated contour with clear margins without perirenal fat infiltration. Typically, fat infiltration in the perirenal space coexists with ECD.

In conclusion, we described a patient with Erdheim-Chester disease with perirenal masses containing macroscopic fat tissue. Perirenal involvement of ECD may present as bilateral perirenal masses with macroscopic fat tissue.

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지방을 함유한 신장 주변 종괴의 형태로 발현한 Erdheim-Chester Disease

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Erdheim-Chester disease는 비랑게르한스세포 조직구증으로 신체의 다양한 부위를 침범하는 희귀질환이다. 조직검사에서는 지질을 함유한 대식세포가 모여 침윤되어 있는 것을 관찰할 수 있지만 영상검사에서는 지방 조직의 침착은 관찰되지 않는다. 40세 남성이 신장 주변에 지방을 함유한 종괴에 대한 검사를 위해 내원하였고, 이 종괴는 Erdheim-Chester disease에 의한 신장 주변 침범으로 진단되었다. 이에 신장 주변에 지방을 함유한 종괴 형태로 발현한 Erdheim-Chester disease를 경험하였기에 보고하는 바이다.

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