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The Collapsing Variant of Focal Segmental Glomerulosclerosis (FSGS) Secondary to Sarcoidosis, A Very Rare Case

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Sarcoidosis is a multisystemic granulomatous disease of unknown etiology. Renal involvement in sarcoidosis patients is occurred, but the incidence and prevalence is uncertain. The most common renal involvement of systemic sarcoidosis is nephrocalcinosis and interstitial nephritis. After sarcoidosis was diagnosed in a 31-year-old male patient, we performed a renal biopsy because of nephrotic range proteinuria and renal dysfunction. The collapsing variant of focal segmental glomerulosclerosis (FSGS) secondary to sarcoidosis was diagnosed by kidney biopsy.

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Case Report

INTRODUCTION

Sarcoidosis is a multisystemic granulomatous disease of unknown etiology. The incidence of renal involvement ranges from 3-23% with a wide spectrum of abnormalities.[1] Because of the inadequate and late diaognosis of renal disease, it can't be obtained exactly the frequency of kidney disease in sarcoidosis patients.[1,2] The most common findings in renal involvement are hypercalcemia and hypercalciuria.[3] Spectrum of renal lesions characteristics are changed for tubulointerstitial granulomatosis to glomerulonephritis.[1,4,5] Although interstitial granulomatous nephritis and nephrocalcinosis is a common pathology in the renal involvement of sarcoidosis, collapsing variant of the focal segmental glomerulosclerosis (FSGS) secondary to sarcoidosis is a very rare condition.[6]

CASE REPORT

A 31-year-old male patient applied to the nephrology outpatient clinic with complaints of shortness of breath, cough, sputum, and weight loss. Patient had medical history including hypertension, but he didn't use any medication. The patient's biochemical examination was as following: creatinine, 4.77 mg/dL; urea, 99 mg/dL; corrected calcium, 12.9 mg/dL; uric acid, 8.6 mg/dL; 25-hydroxyvitamin D, 14.1 ng/mL; parathormon, 11.5 pg/mL; magnesium, 2.1 mg/dL; and phosphorus, 3.6 mg/dL.

In radiological imagings, bilateral hilar and mediastinal multipl lymph nodes and bilateral pleura based wide consolidation sites and reticulonodular densities were observed (Figure 1). The patient's sputum acid-fast bacilli (AFB) and culture were negative. The serologic parameters were negative. 24-hour urine calcium was 0.4 g/d and 24-hour urine protein was measured as 2.49 g/d. Serum level of angiotensin converting enzyme (ACE) was seen in the reference range. Endobronchial lesion and mucosal pathology were not observed in bronchoscopy. Broncholaveolaer

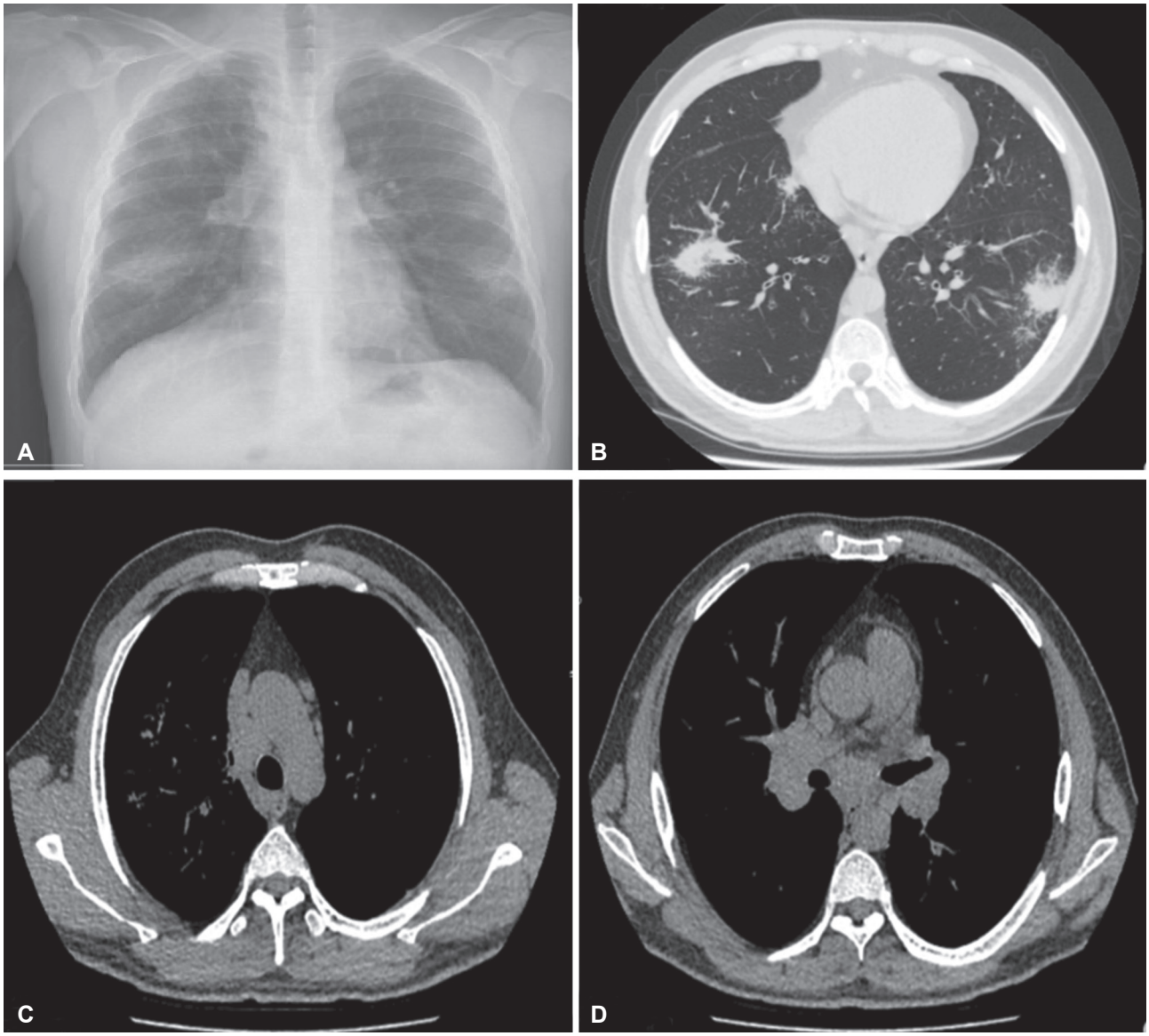

Figure 1. A) Posteroanterior Chest X-ray. Peripheral reticulonodular densitic areas were observed in bilateral lower zones, B-C-D) Non-contrast Thorax Computed Tomography (B: peripheral consolidation areas in bilateral lower lobes, C: right inferior paratracheal lymph nodes, and D: subcarinal lymph nodes).

lavage (BAL) was performed from the superior lobe of right lung. In BAL culture, AFB negative, CD4 / CD8 ratio was measured as 1.32. No growth was observed in the nonspecific culture of BAL, and no malignant cells were observed in cytology. The pathology of the patient, whose sampling of upper paratracheal, right paratracheal and subcarinal lymph nodes were performed by mediastinoscopy, was reported as noncaseating granulomatous lymphadenitis.With the diagnosis of pulmonary sarcoidosis, methylprednisolone treatment at a dose of 1 mg/kg/d was started.

One month after starting treatment, renal biopsy was performed because of 24-hour urine protein level increasing to 7.5 g/d. No deposition was seen in immunofluorescence microscopy. In immunohistochemical examination, positive staining in favor of podocyte was observed with vimentin in the proliferation observed in 2 glomerules. Global sclerosis (12 / 17), focal segmental sclerosis (4 / 5), tubular atrophy and injury, interstitial inflammation and fibrosis were observed in the glomerules (Figure 2). Focal segmental glomerulosclerosis (collapsing variant) was diagnosed. After the reduction and discontinuation of steroid therapy in the 10th month, renal function tests progressed

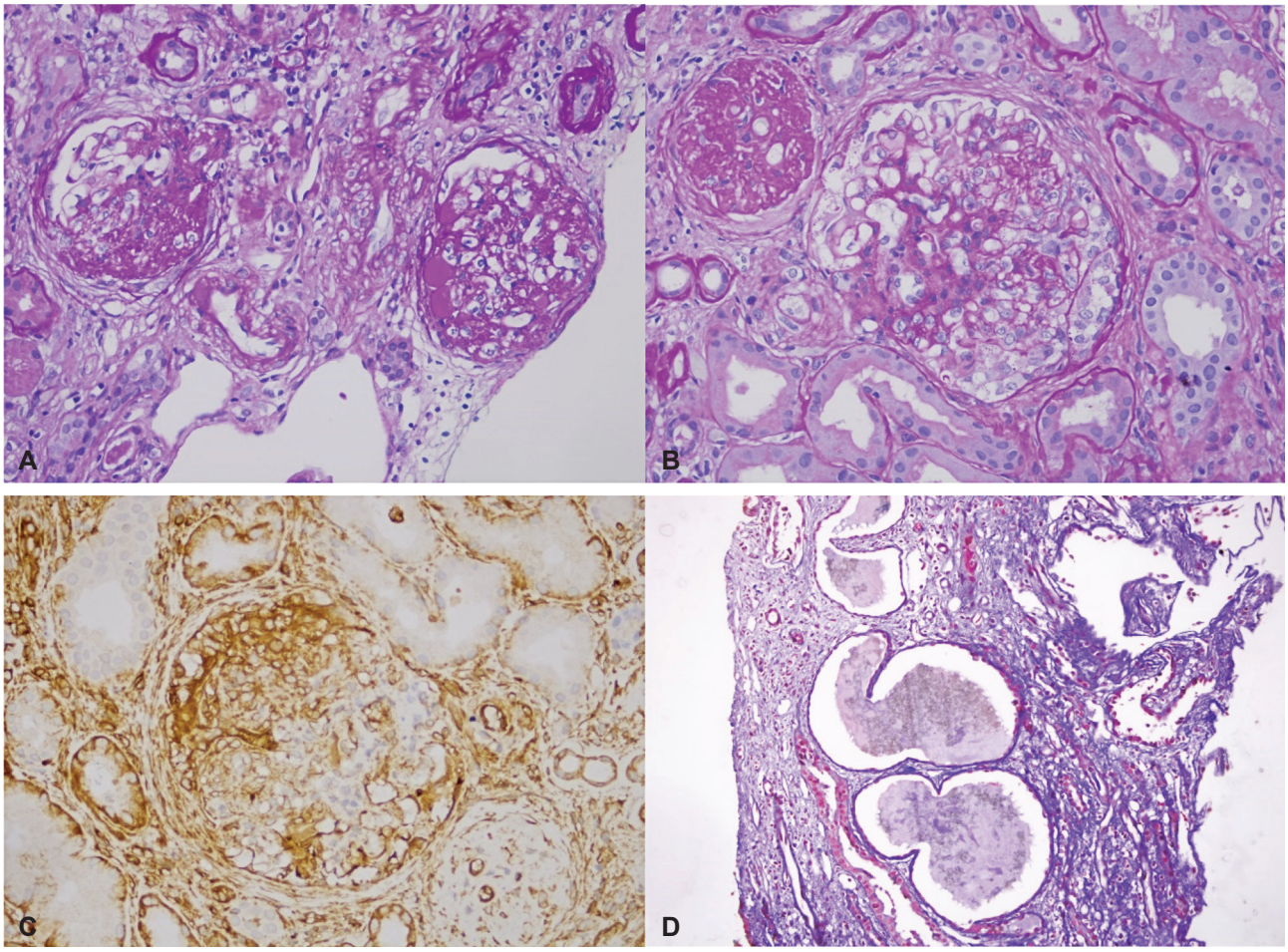

Figure 2. A) focal segmental mesengial matrix increase and segemental hyalinosis in two glomerules, Periodic acid shift (PAS)×200, B) glomerular capillary tuft collaps and podocytes proliferation, global sclerosis in one glomerule, Periodic acid shift (PAS);×200, C) staining in proliferated podocytes with Vimentin. IHK;×400, and D) cystic dilatation, tubular atrophy and interstitial fibrosis in tubules. Masson-trichrome; ×200.

to end-stage renal failure, although lung findings regressed as seen in Figure 3.

DISCUSSION

Sarcoidosis related renal diseases often show a good response to initial steroid theraphy in most patients after the first month.[7] In our case, we didn't have a good response to this theraphy, so that we must perform the diagnosis of the renal disease. In this situation, we don't have a chance to distinguish nephrotic syndrome etiology without renal biopsy like measurement of urine CD80 levels in Ahmed *et al.* study.[8] We performed renal biopsy and the pathology result comes collapsing variant of FSGS. In consecutive studies, in renal involvement sarcoidosis patients, performed renal biopsy: 11 membranous nephropathy in 26 patients [9] and 26% Ig A nephropathy in 27 patients.[10] In the literature, there is no case of the collapsing variant of FSGS secondary to sarcoidosis.

In development of collapsing variant FSGS, the main cause is HIV infection or idiopathic. Sarcoidosis is not included in the other rare etiology.[11] Collapsing FSGS often progresses rapidly to the end-stage renal failure among patients who are resistant to initial immunosuppressive therapy like our case, and it predicts poor prognosis.

In Yassari *et al.* study, serum uric acid levels were found above 7 mg/dL in patients with sarcoidosis who had renal disorders, similar to our case.[12] Although a significant association between proteinuria and kidney failure has not been shown in this study, in our case, it can be considered that uric acid and proteinuria both may be responsible for the progression of kidney failure.

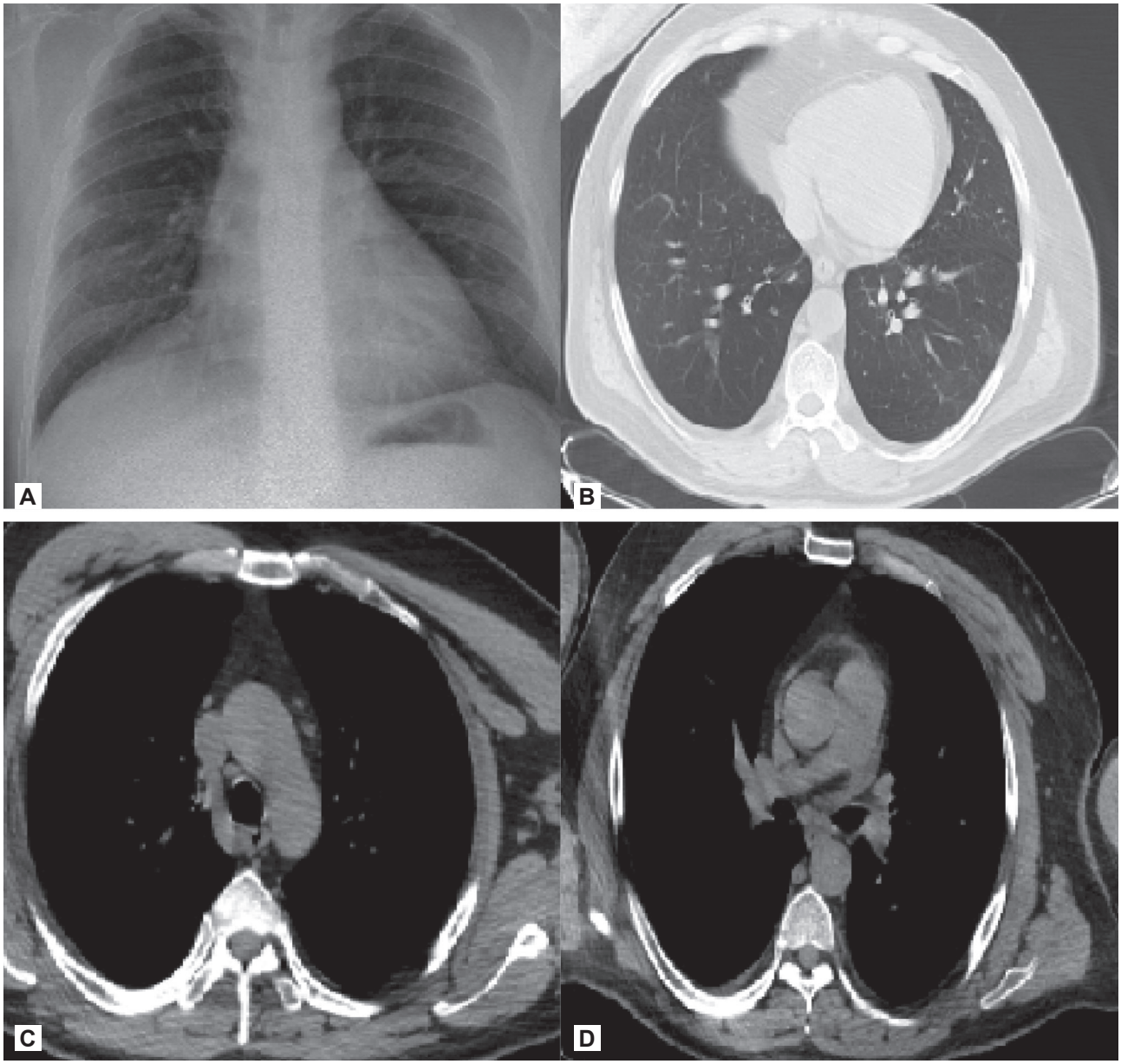

Figure 3. A) posteroanterior chest x-ray, B-C-D) non-contrast thorax computed tomography (nearly normal radiological imaging were found at the tenth month after diagnosis**).**

CONCLUSION

Collapsing FSGS should be brought to mind in case of nephrotic syndrome and impaired renal function that do not respond to steroid treatment in patients with sarcoidosis.

CONFLICT OF INTEREST

There is no conflict of interest and financial disclosure.

INFORMED CONSENT

Informed consent was obtained from the patient.

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