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An unusual cause of advanced chronic kidney disease in a middle-aged female

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Case Study: Background: Kidney involvement in IgG4-related disease (IgG4-RD) usually manifests as acute progressive renal dysfunction, mostly in the elderly. Breast involvement in IgG4-RD is rare and may clinically resemble malignancy. We report a 42-year-old female who presented with advanced kidney disease and breast mass mimicking malignancy who was diagnosed as IgG4-RD. We describe the protean clinical and pathological manifestations of IgG4-RD.

Case Report: A 42-year-old female presented with complaints of generalized weakness and low-grade fever occasionally for a period of 3 months. She had loss of appetite and weight loss of 12 kgs over 3 months. She had pallor, left breast mass with left cervical and left axillary lymphadenopathy. Serum creatinine was 6.3 mg/dl; ultrasonography was suggestive of shrunken kidneys with focal lesions bilaterally. The Mantoux test was strongly positive. Imaging revealed focal left breast mass with nipple retraction and left axillary and cervical lymphadenopathy. Cervical lymph node biopsy revealed granulomatous inflammation, however, GeneXpert was negative for tuberculosis. Breast biopsy was suggestive of granulomatous mastitis with no features of malignancy. Kidney biopsy from the focal lesions was suggestive of interstitial lymphoplasma cell infiltration and advanced interstitial fibrosis with tubular atrophy. IgG4 immunohistochemistry, done in view of plasma cell infiltration, showed 30 IgG4-positive plasma cells/high power field on kidney biopsy. IgG4 immunostaining was then done on the breast biopsy which showed plenty of plasma cells with >40% being IgG4-positive. Serum IgG4 level was 516 mg/dl. IgG4-RD was diagnosed involving kidneys, breast and lymph nodes. Patient was started on oral prednisolone. At a follow-up of 1 month, her left breast mass was the same and lymph nodes were smaller.

Conclusion: We report an atypical cause of advanced chronic kidney disease in a middle-aged female. The presence of granulomas, although odd, can be a feature of IgG4-RD.