

中文題目：不明熱罕見病例個案：原發性腎臟淋巴瘤

英文題目：fever of unknown origin related to primary kidney lymphoma : A case report

作者：范聖暉¹，林敬業^{1,2}

服務單位：彰化基督教醫院¹內科部，²血液腫瘤科

Case presentation

A 43-year-old woman without any medical history was admitted to the hospital with intermittent fever, chills, myalgia, dry cough, and decreasing appetite for one month. She had already been hospitalized for one week prior to this admission; at the time, she was treated with empirical antibiotics for community-acquired pneumonia. She was transferred to our hospital owing to prolong fever despite having completed the prescribed course of antibiotics. She did not report dysuria, urinary frequency, flank pain, nausea, vomiting, abdominal pain, diarrhea, abnormal skin lesions, joint pain, body weight loss, or night sweating. Physical examination revealed an enlarged spleen, and there was no sign of lymphadenopathy or hepatomegaly. The white blood cell count was 7300/ μ L, and the platelet count was 78,000/ μ L; red blood cell morphology was normal, as assessed using a peripheral blood sample. The differential counts were all within normal ranges. The level of hemoglobin was low at 8.0 g/dL, and the levels of serum ferritin, vitamin B₁₂, and folic acid were normal. The serum albumin level was 1.4 mg/dL, with a reversed albumin-globulin ratio of 0.4 and high C-reactive protein level of 11.6 mg/dL. Renal, liver, and coagulation functions were normal. Results of urinalysis revealed no hematuria, pyuria, or proteinuria. Serological tests for *Mycoplasma pneumoniae*, *Legionella pneumophila*, and *Chlamydia pneumoniae*, were negative. Results of sputum culture; acid-fast staining; blood culture; and extensive viral serology screening for cytomegalovirus, Epstein–Barr virus, parvovirus B19, human immunodeficiency virus were negative. Results of interferon-gamma release assays and skin tuberculin test were negative.

There were no obvious signs of infection according to clinical manifestations and laboratory data; therefore, we arranged examinations for autoimmune and oncological diseases. The antinuclear antibody (ANA) titer was 1:320. Tests results for anti-ds-DNA antibody, anti-Sm antibody, anti-Ro antibody, and anti-La antibody were negative. The anti-CCP antibody level was 1.5 EliAU/mL (normal range, 7–10 EliAU/mL). The levels of complement 3 and complement 4 were 132 mg/dL (normal range, 90–180 mg/dL) and 19.2 mg/dL (normal range, 10–40 mg/dL), respectively. Anti-neutrophil cytoplasmic antibody (ANCA) was absent. The levels of tumor markers such as carcinoembryonic antigen (CEA), CA-125, CA 19-9, and

alpha-fetoprotein were within normal ranges. The level of lactate dehydrogenase (LDH) was 355 U/L (normal range, 98–192 U/L) and that of beta-2 microglobulin was 3.66 mg/L (normal range, 1.09–2.53 mg/L). The levels of serum immunoglobulin and free light chain were normal, and the serum protein electrophoresis report was also normal.

Results of abdominal sonography (Fig. 1) revealed presence of severe splenomegaly and absence of hepatomegaly or enlarged lymph nodes. Abdominal computed tomography (CT) scan (Fig. 2) and gallium scan (Fig. 3) obtained for evaluating fever of unknown origin revealed bilateral striated nephrograms and increased 2-deoxy-2-[18F] fluorogluucose (FDG) metabolism in both kidneys, respectively. Ultrasound-guided biopsy was arranged for the right kidney, and results of histological analysis revealed the presence of a large B-cell lymphoma. Whole-body positron emission tomography/computed tomography (PET/CT) (Fig. 4) performed for lymphoma staging revealed increased FDG metabolism in the bilateral kidneys and no other abnormal metabolic lesions. Bone marrow examination revealed hypercellular marrow, and adequate numbers of megakaryocytes. Immunohistochemical study results showed that the level of CD34 was less than 5% of blasts. The CD3 and CD20 stain highlights scattered reactive lymphocytes.

Based on these findings, she was diagnosed with stage I primary renal lymphoma (PRL). She was treated with a chemotherapy regimen comprising rituximab, vincristine (1.4 mg/m²), epirubicin (75 mg/m²), and cyclophosphamide (750 mg/m²). The fever remitted after chemotherapy, and there was no discomfort or complications after treatment.

Figure 1.

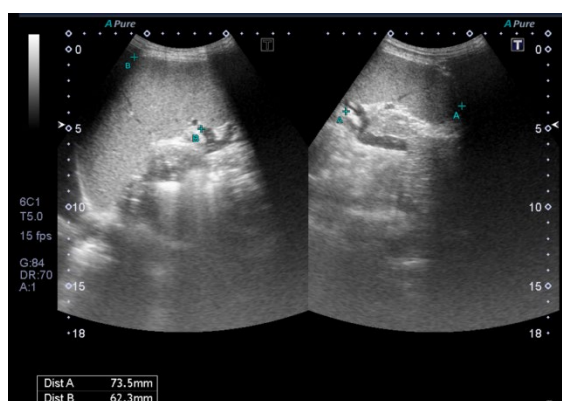


Figure 2.



Figure 3.

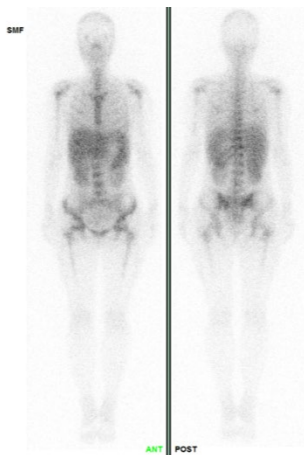
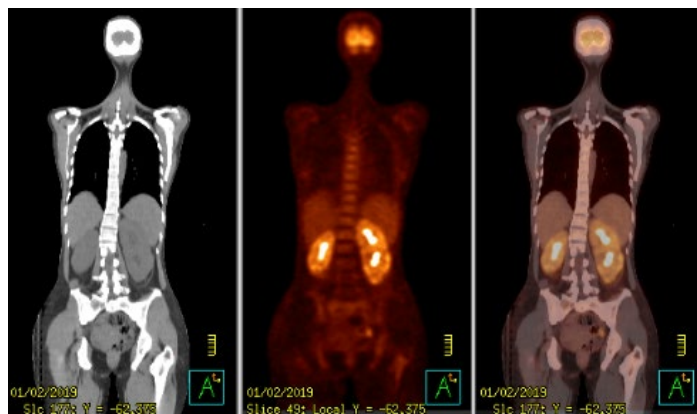


Figure 4.



Discussion

PRL is an extremely rare disease and is defined as non-Hodgkin's lymphoma involving the kidney in the absence of primarily extrarenal lymphatic disease. (1) The yearly incidence of PRL was 0.7 cases. (2) It accounts 0.1% of all malignant lymphomas in Japan. (3) The majority of PRL is large B-cell type and it is often presented on one side of the kidney in adults. (4) There were more male patients than female patients in previous reports and the ratio was $\sim 1.6:1$. (3) Patients with PRL may present with prolonged fever, gross hematuria, acute or chronic kidney injury, and flank pain or body weight loss. PRL is difficult to diagnose by imaging alone owing to its nonspecific manifestations. It can manifest as one or multiple renal masses, perirenal disease, or diffuse renal infiltration. (5,6) Results of a study showed that the levels of anti-dsDNA, c-ANCA, or ANA, may be high in patients with non-Hodgkin's lymphoma, especially in those with diffuse large B-cell lymphoma. (7) Chemotherapy remains the standard treatment for PRL.

References

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