中文題目: 紅斑性狼瘡病患合併瀰漫性脾臟鈣化

英文題目: Diffuse splenic calcifications in a patient with systemic lupus erythematosus

作　　者: 邱玲榕[1], 李禎祥[2]

服務單位: 高雄長庚紀念醫院內科部[1]，高雄長庚紀念醫院感染科[2]

Introduction

Splenic calcifications have been found in autosplenectomy, thorium dioxide exposure–related fibrosis, tuberculosis, Pneumocystis carinii infection, and Histoplasma capsulatum infection (1). Rarely, calcification can also be associated with systemic lupus erythematosus (SLE) of the spleen (2). Herein, we present a case with a history of SLE, who was subsequently infected with Mycobacterium kansasii and developed diffuse splenic calcifications.

Case Report

A 69-year-old female, with a history of SLE for 12 years, suffered from episodic epigastric pain and chronic cough for 7 years. She felt hunger pain, which radiated to the left shoulder, progressive abdominal fullness in the left hypochondrial region, and a weight loss of 10 kg in the past one year. A chest x-ray revealed the infiltration in the lower lobe of the right lung, while a plain abdominal x-ray showed diffuse calcification of the spleen (Figure 1A). Abdominal computed tomography (CT) showed splenomegaly with microlithiasis, without enlarged lymph nodes (Figure 1B). A bronchoscopy did not show any endobronchial lesion. The culture of the bronchial wash over the lower lobe of the right lung yielded no bacterial growth. Other test results, including those of acid fast staining, tuberculosis polymerase chain reaction test, cytology, Cryptococcus antigen, and aspergillus antigen were negative. However, a mycobacterial culture yielded M. kansasii one month later.

She received therapy with rifampicin, ethambutol, and azithromycin. The abdominal pain and cough gradually subsided. A chest x-ray showed the regression of the lesion in the lower lobe of the right lung, but the follow-up abdominal CT scan did not show the resolution of splenomegaly and microlithiasis.

Discussion

The exact pathological mechanisms of spleen calcification have not been identified. This patient received anti-mycobacterial therapy for 12 months, which had minimum effect on spleen calcification at the follow up abdominal CT scan. This suggests that spleen calcification might be due to SLE-related peri-vascular calcification and fibrosis rather than M. kansasii infection. Spleen calcification is a characteristic of connective tissue diseases, but the exact significance of diffuse splenic calcification is still unknown. This unique radiologic finding may be a result of the disease process itself (2). The association between high levels of anti-DNA antibodies and disease activity has been widely appreciated (3). In our case, SLE activity and splenic calcifications observed in the abdominal CT scan increased simultaneously during 2014-2016.

With increasing SLE activity, immunosuppressants are prescribed to control the disease activity. However, there is a higher incidence of multiple opportunistic infections in SLE patients with

increased disease activity controlled with the indicated doses of corticosteroids (4). Given that pulmonary calcifications were found to remain unchanged or increase in size after complete anti-tuberculosis medication in 17.8% cases (5), it is significant that spleen calcification did not diminish, although symptoms of cough and abdominal pain improved after anti-mycobacterial regimens. The fibrosis and scarring occurring in caseating granulomas have been found to be irreversible in a previous histopathologic analysis.

Conclusion

The significance of this case is the worsening of spleen calcifications in an SLE patient with increased disease activity after *M. kansasii* infection.

References

1.Consul N, Javed-Tayyab S, Lall C, Jensen CT, Menias CO, Pickhardt PJ, et al. Calcified splenic lesions: Pattern recognition approach on CT with pathologic correlation. AJR Am J Roentgenol. 2020;214(5):1083-1091.
2.Tieng AT, Sadow CA, Hochsztein JG, Putterman C. Diffuse calcifications of the spleen: a novel association with systemic lupus erythematosus. Semin Arthritis Rheum. 2011;41(2):187-193.
3.Wang X, Xia Y. Anti-double stranded DNA antibodies: Origin, pathogenicity, and targeted therapies. Front Immunol. 2019;10:1667.
4.Yang SC, Lai YY, Huang MC, Tsai CS, Wang JL. Corticosteroid dose and the risk of opportunistic infection in a national systemic lupus erythematosus cohort. Lupus. 2018;27(11):1819-1827.
5.Lee HS, Oh JY, Lee JH, Yoo CG, Lee CT, Kim YW, et al. Response of pulmonary tuberculomas to anti-tuberculous treatment. The European respiratory journal. 2004;23(3):452-455.

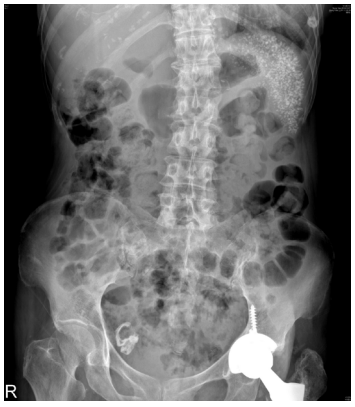

FIGURE 1A Abdominal plain film revealed diffuse spleen calcification.

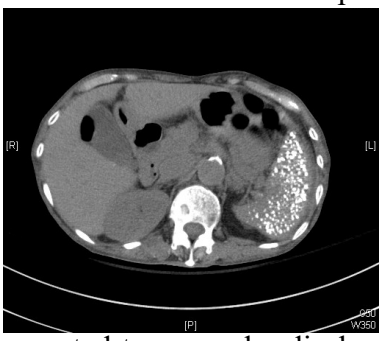

FIGURE 1B Abdominal computed tomography disclosed diffusely and densely distributed, coarse, round, andovoid splenic calcifications along with microlithiasis in the spleen.