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# Systemic Lupus Erythematosus: An Exceptional Cause of Diffuse Splenic Calcifications. A Case Report and Review of the Literature

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**Abstract** **Case Report**

Splenic calcifications have been reported in various diseases other than SLE, such as rheumatoid arthritis, systemic sclerosis, amyloidosis, sickle cell anemia, lymphoma, and post-infectious or post-traumatic sequelae. However, they are extremely rare in SLE, and only a few cases have been reported in the literature. In this article we discuss the case of a 32-year-old woman with systemic lupus erythematosus (SLE) who was found to have diffuse splenic calcifications, discovered incidentally during a CT scan performed to assess her pancreatic involvement due to SLE.

**Keywords:** Lupus – Splenic calcifications – rare condition.



## INTRODUCTION

Lupus Erythematosus Disseminatus (LED) is characterized by multisystem involvement; it affects various organs, including the spleen. Splenic involvement is mainly manifested by splenomegaly, hypersplenism, infarction, spontaneous rupture, functional asplenia and peri-arterial thickening of the spleen (Tieng *et al.,* 2011a) (Krauser, 1976). Splenic calcifications have been reported mainly in rheumatoid arthritis, systemic scleroderma, infections, sickle cell disease, splenic hemangiomas, as well as B-cell lymphoma (A. J. Fyfe *et al.,* 2009.). LED is not recognized as a cause of splenic calcifications, and few cases have been reported in the literature. We report the case of a woman followed for LED in whom diffuse splenic calcifications were discovered incidentally.

## OBSERVATION

This is a 32-year-old unmarried woman, an instructor by profession, who has been followed for 15 years for acute systemic lupus erythematosus, with joint, skin, and renal involvement consisting of Type III segmental and focal glomerulonephritis. The patient presented with dry eye syndrome alone without oral dryness, treated symptomatically. The patient was hospitalized in October for management of her first renal flare-up. The laboratory tests showed elevated levels with a creatinine level of 16.1 with an estimated glomerular filtration rate (GFR) of 39ml/min/1.73m² and proteinuria of 1.17g/24h. During her hospitalization, the patient had transfixing epigastric pain, with a slightly elevated lipase level of 186, and a diagnosis of pancreatitis was made.

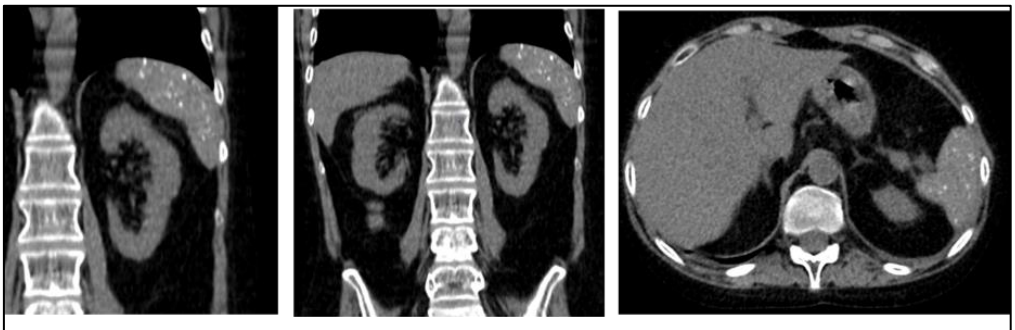

**Figure 1:** CT sections showing a normal sized spleen with diffuse calcifications

An abdominal CT scan was performed to stage the pancreatitis, the abdominal scan was performed without contrast injection as the patient had severe renal insufficiency. The abdominal CT scan showed a normal-sized, well-defined, homogenous, lobulated pancreas with mild infiltration of peri-pancreatic fat. The gallbladder was of normal size, with spontaneously hyperdense material of calcic density (microcalculi). The spleen was of normal size, with regular contours, showing multiple diffuse punctuate calcifications. **(Figure 1)**

A complementary ultrasound was performed and showed a spleen of normal size, with regular contours, showing diffuse calcifications of variable size, hyper-echoic generating a posterior shadow cone, with respect to the splenic capsule and subcapsular parenchyma. **(Figure 2)**

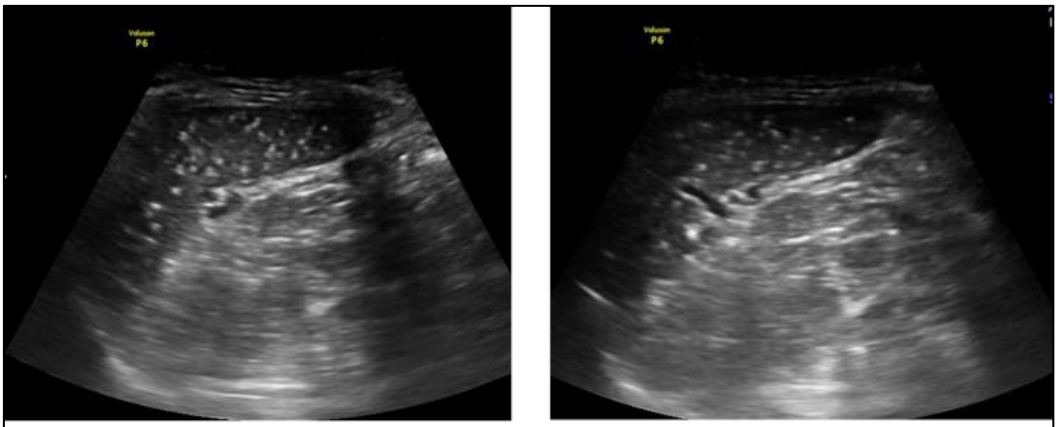

**Figure 2:** Ultrasound showing a normal sized spleen with diffuse calcifications

In summary, it was concluded that the patient had pancreatitis that could be classified as Balthazar stage B with gallbladder lithiasis. Given the absence of collection or infectious history, it was concluded that the diffuse splenic calcifications were of lupus origin. Unfortunately, the patient subsequently developed multi-visceral failure due to renal insufficiency, and she passed away within two weeks of her acute pancreatitis.

## DISCUSSION

Systemic lupus erythematosus (SLE) is a multisystem autoimmune disease that affects various organs, including the spleen. Splenic involvement is mainly characterized by splenomegaly, hypersplenism, infarction, spontaneous rupture, functional asplenia, and periarterial splenic thickening (Tieng *et al.*, 2011a). Splenic calcifications have been generally reported in rheumatoid arthritis, systemic sclerosis, infections, sickle cell disease, splenic hemangiomas, as well as B-cell lymphoma (A. J. Fyfe *et al.*, 2012.). SLE is not recognized as a common cause of splenic calcifications, and few cases have been reported in the literature. Since their first description in 1957, only about ten cases of splenic calcifications related to SLE have been reported (Tieng *et al.*, 2011a).

These calcifications can be detected incidentally during a radiographic examination (chest or abdomen without preparation), an ultrasound, or an abdominal CT scan. In the case of our patient, the discovery was also incidental, and we ruled out trauma through questioning and tuberculosis through questioning and negative biological testing. Other etiologies were eliminated during her hospitalization based on precise biological and immunological assessments.

Radiologically, splenic calcifications have a generally characteristic appearance, presenting as small round calcifications, distributed diffusely and homogeneously in the splenic parenchyma, sparing the capsule and subcapsular tissues (Enfrein *et al.*, 2020) (Kwee & Kwee, 2015). They are considered the result of the long-term evolution of peri-arteriolar fibrosis lesions, which creates an aspect of "onion skin" lesions that are considered characteristic of this condition (Tieng *et al.*, 2011a) (Kwee & Kwee, 2015).

Tan Tieng *et al.*, (Tieng *et al.*, 2011b) (Farras *et al.*, 2012) and Ferras *et al.*, (Farras *et al.*, 2012) reported on a series of 4 patients that their patients mainly presented discreet, small, round calcifications, some of which are cylindrical and larger than the punctate calcifications observed in granulomatous infection. Additionally, they observed that the more linear, tubular, or ovoid morphology of many calcifications observed suggests that calcifications could be in blood vessels.

They also reported that the spleens studied were of normal size or slightly enlarged, and that splenic calcifications associated with lupus generally spare the external parenchyma and capsule, which is

consistent with cases reported in the literature and with our patient's case.

It should also be noted that abdominal computed tomography reports in lupus patients may not mention splenic calcifications as one of the imaging findings (Phongkitkarun *et al.*, 2007), and may consider them as calcification consequences, which could explain the number of cases reported in the literature (Phongkitkarun S *et al.*, 2007;90:2112-20, s. d.).

Another hypothesis suggests that diffuse splenic calcifications may be related to past infections due to the increased use of immunosuppressants and corticosteroids in lupus treatment protocols (Chiu & Lee, 2021). However, this hypothesis is not well-supported since lupus patients undergo close monitoring and rigorous clinical and biological control, and all reported cases in the literature have ruled out the possibility of primarily tuberculosis infection. Additionally, the morphological description of lupus-related calcifications is different from the granulomas described in infections affecting the spleen.

## CONCLUSION

In conclusion, splenic calcifications are rarely observed in patients with chronic systemic lupus erythematosus and are often discovered incidentally. Their detection and description can lead to further investigation of possible mechanisms. Additional studies are now needed to better understand the association between SLE and diffuse splenic calcifications as well as their pathological significance.

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