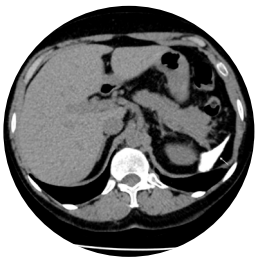

Stellate spleen. A case report

Bazo estrellado. A propósito de un caso

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Summary

Splenic lesions may go unnoticed in imaging studies requested for other purposes. Splenic calcifications represent a wide spectrum of lesions that can arise secondary to various underlying pathological processes. Autosplenectomy is an extremely rare entity with some cases described in the literature associated to autoimmune diseases. The case of a 46-year-old patient with Systemic Lupus Erythematosus (SLE) is presented, in whom the calcified and atrophic spleen is found as an incidental finding in a Chest tomography.

Key words (MeSH)

Spleen
Calcification
Computed tomographic

Palabras clave (DeCS)

Bazo
Calcificación
Tomografía computarizada

Resumen

Las lesiones esplénicas pueden pasar desapercibidas en los estudios imagenológicos solicitados con otros fines. Las calcificaciones esplénicas representan un amplio espectro de lesiones que pueden originarse secundarias a diversos procesos patológicos subyacentes. La autoesplenectomía es una entidad extremadamente rara con algunos casos descritos en la literatura asociados a enfermedades autoinmunes. Se presenta el caso de una paciente de 46 años de edad con lupus eritematoso sistémico (LES) en quien se encuentra el bazo calcificado y atrófico como hallazgo incidental en una tomografía axial computarizada (TAC) de tórax.

Introduction

The spleen is an organ that is often neglected despite the fact that it is involved in numerous pathological processes. In a great variety of cases, splenic calcifications have been described secondary to different entities, such as granulomatous diseases associated with infectious processes, hematological and autoimmune diseases (1). Autosplenectomy is a rare entity in which numerous infarctions occur causing hyposplenia and complete calcification of the spleen, leading to a decrease in the function of this organ. While true congenital asplenia occurs sporadically with systemic lupus erythematosus (SLE), acquired functional asplenia has been described in only 3-7% of cases and is often associated with thrombocytosis (2). Since the first case, described by Dillon et al. in 1980 (3), less than 20 cases of acquired functional asplenia in SLE patients have been published to date (2). We present the case of a patient with a history of SLE in whom splenic calcification is found as an incidental finding in computed axial tomography (CT).

Case report

A 46-year-old female patient diagnosed with SLE at 19 years of age, which debuted with Staphylococcus aureus pneumonia, who was under radiological follow-up for a left pulmonary subsolid nodule. As an incidental finding, the chest X-ray showed a dense triangular calcified image, compatible with the splenic silhouette (Figure 1). Chest CT showed a highly calcified spleen, with a density of 481 Hounsfield Units, atrophic appearance, with dimensions of 21 × 51 × 18 mm and splenic index of 19, with a volume of 41 cm3 and an estimated weight of 43 grams (Figure 2).

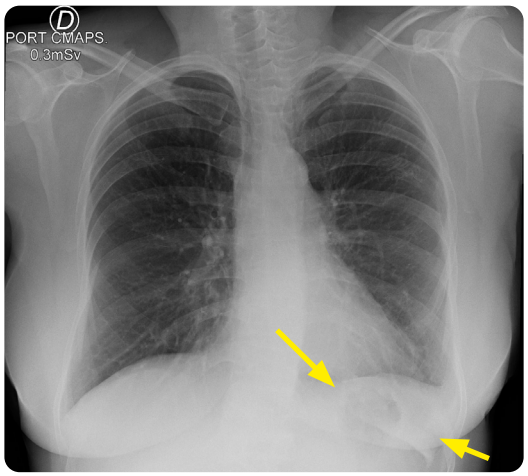

Figure 1. Chest X-ray, PA projection. As an incidental finding, there is a dense calcified image of triangular appearance in the region of the left hypochondrium, compatible with the splenic silhouette.

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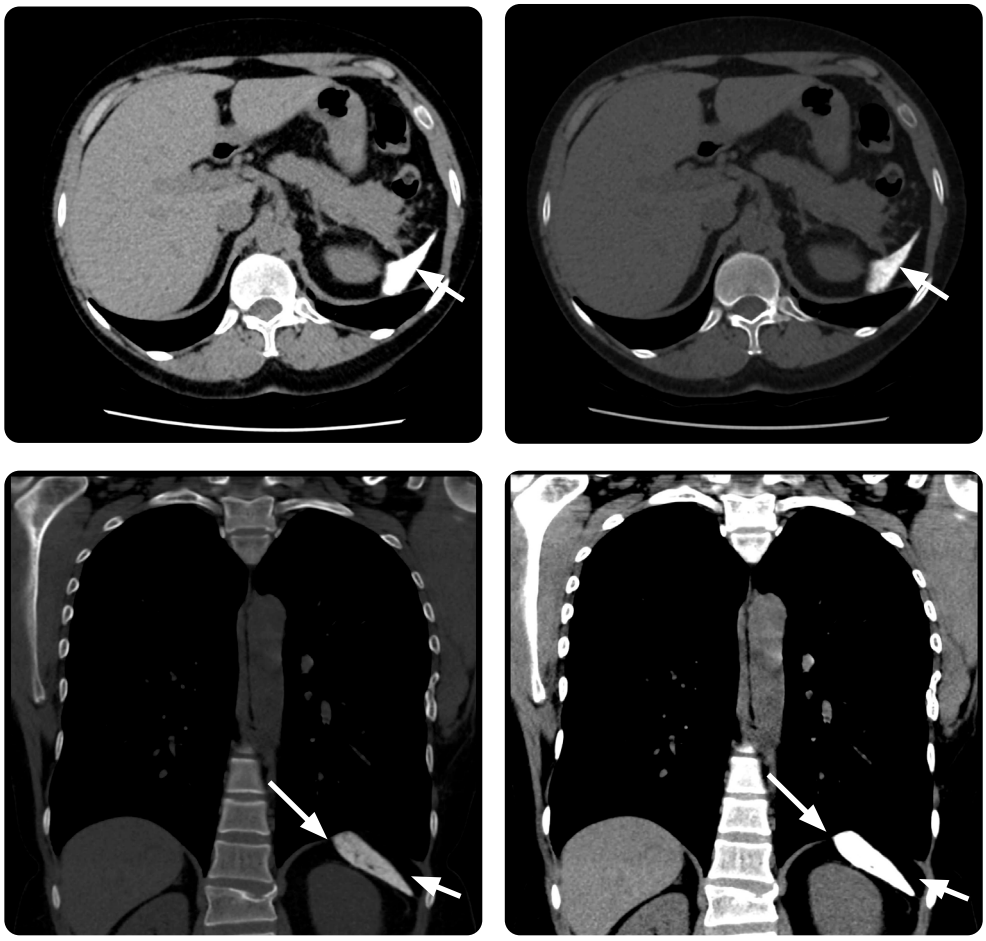

Figure 2. Chest CT, images a) coronal in bone window, b) axial in bone window, c) axial in soft tissues window and d) coronal in soft tissues window. There is an atrophic aspect spleen (white arrows), densely calcified (481 HU), with an appearance frequently known as "stellate spleen".

Discussion

SLE is a multisystem disease involving several organs, including the spleen (4-6). Splenomegaly, traumatic rupture, infarction and hyposplenism are among the most common conditions (4, 5); however, calcifications have not been frequently described in SLE.

There are multiple causes of splenic calcifications, including infectious causes such as histoplasmosis, tuberculosis and brucellosis, which manifest as diffuse dotted or rounded calcifications in a normal sized spleen. Another cause of calcifications is autosplenectomy, which is seen in patients with sickle cell anemia, as a result of an alteration in spleen perfusion due to the presence of abnormal polymerized hemoglobin. Such alteration slows circulation and leads to congestion, splenomegaly and eventually multiple microinfarcts that will eventually lead to diffuse ferrocalcinosis with little functional residual splenic tissue and hyposplenia (1), which manifests as diffuse, amorphous calcification in an atrophic splenic parenchyma, findings that have also been documented in patients with silicosis and coal workers' pneumoconiosis (1).

In the case of SLE, autosplenectomy has been proposed as a pathophysiological mechanism the development of splenic vasculitis with silent infarcts or a blockage of the splenic reticuloendothelial system by elevated levels of circulating immune complexes, resulting in perivascular fibrosis and retraction of the spleen (7). Recently, the possible association of thrombocytosis states in association with antiphospholipid antibodies and the development of autosplenectomy in patients with SLE has been studied (7). Radiologically, patients may present with small rounded or rod-shaped calcifications. This tubular morphology in some of them suggests the possibility that the calcifications are located in blood vessels (Figure 2); in contrast to the calcifications typically observed in other granulomatous processes, calcifications in SLE appear to be found in greater numbers and more profusely distributed in the splenic parenchyma (4, 5), acquiring an "onion skin" pattern appearance; it is believed that this may correspond to a late manifestation of an autoimmune inflammatory process of the blood vessels (4).

Finally, it is important to highlight that patients with autosplenectomy may be asymptomatic; however, they usually debut with multiple repetitive episodes of pain in the left hypochondrium secondary to venoocclusive phenomena. In the face of an alteration in spleen function, it is important to consider that these patients are at increased risk for bacterial infections, so it is ideal that they be immunized against encapsulated bacteria. Cases of sepsis have been described in these patients, so it would be worthwhile to recognize this finding in diagnostic imaging (2).

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