MULTIPLE SPLENIC INFARCTIONS COMPLICATING GRANULOMATOSIS WITH POLYANGITIS

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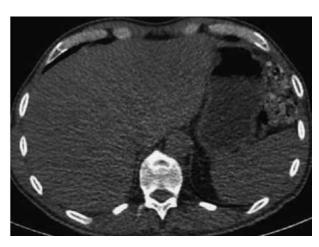
Key-word: Wegener granulomatosis

Background: A 57-year-old patient was admitted for high-grade fever, asthenia, sweating, dry cough and diffuse arthro-myalgias. Two years earlier, elevated titers of anticytoplasmic antibodies (ANCA) of anti-proteinase 3 specificity and renal biopsy led to a diagnosis of granulomatosis with polyangiitis (GPA) with lung and renal involvement. GPA was treated by steroids, cyclophosphamide and rituximab with subsequent clinical and biological remission. The current chest CT scan was performed for a lung opacity that eventually was proved to be an organising pneumonia. CT also showed an unsuspected pattern of the spleen that was compared with a previous chest CT.









Work-up

Contrast-enhanced CT scan of chest and upper abdomen (Fig. 1) consisted of axial CT slices obtained at the level of upper abdomen (A,B) and reformatted images in the coronal plane (C).

Multinodular delineation of the spleen with slightly heterogeneous enhancement is observed. The size of the spleen has decreased in comparison to a CT scan performed 2 years previously (see Fig. 2).

On unenhanced CT scan of the upper abdomen, performed two years earlier (Fig. 2), a slightly enlarged spleen with normal contour is seen.

Radiological diagnosis

Based on the radiological findings and the clinical history, the diagnosis of *multiple splenic infarctions complicating granulomatosis with polyangiitis* was made.

Discussion

Granulomatosis with polyangiitis (GPA, previously named Wegener granulomatosis) is a systemic necrotizing vasculitis affecting small and medium-sized vessels with granuloma formation. GPA is an uncommon autoimmune disease occurring between 40 and 50 years of age with a slight male predominance. Ear-Nose and Throat (ENT), upper and lower respiratory tract, kidneys and, less frequently, other organs such as the skin, joints, central nervous system, heart, and eyes are mainly affected. Although splenic involvement has been rarely reported, some necropsy series have demonstrated a high incidence of splenic involvement in GPA (splenomegaly, vasculitis, granuloma, necrosis, capsulitis), suggesting that it remains mostly

asymptomatic. Splenic infarctions seem to result from distal occlusion of intrasplenic arteries.

Cross sectional imaging including CT scan or MRI is sensitive for the detection of splenic infarctions that may present with various patterns. In contrast to the classic wedge-shaped peripheral hypodense lesions as seen in embolic disease, in GPA CT scan shows a more widespread involvement with a heterogeneous splenic enhancement or a large low attenuation central region. In our case, splenic enlargement was observed in the acute phase whereas a multilobulated pattern with shrinkage and return to normal splenic enhancement was demonstrated at distance of the acute episode. Rare complications can arise as a consequence of the splenic infarction, including secondary infections with sometimes abscess formation, bleeding or spontaneous rupture. A conservative approach is mostly justified except in acute complications such as spleen hemorrhage or rupture. In the long term, these patients may be more susceptible to pneumococcal infection because of the resulting hyposplenism.

Bibliography

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