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POLYARTERITIS NODOSA OF THE SPLEEN

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Key-word: Arteritis

Background: A 64-year-old male presented with pain in his right lower leg and progressive swelling of the right ankle, without history of previous trauma. Further anamnesis revealed fever, anorexia and weight loss over the past few weeks. Laboratory results showed increased inflammatory parameters (CRP 203 mg/l).
A lower extremity ultrasonographic examination ruled out deep venous thrombosis.
A contrast-enhanced CT scan of the abdomen revealed multiple splenic infarctions.

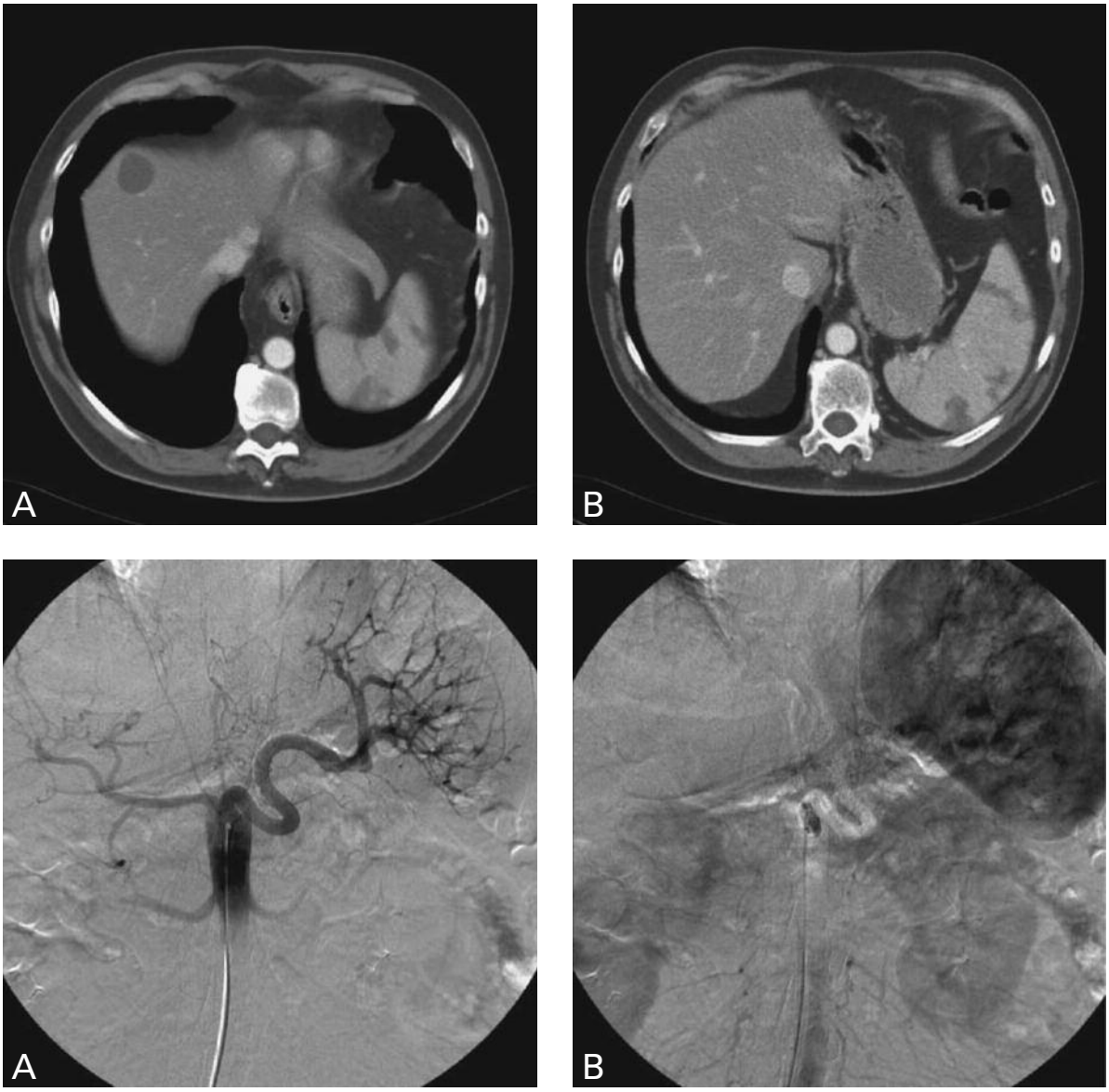

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Fig. 1A | 1B / 2A | 2B

Work-up

Contrast-enhanced CT scan of the abdomen (Fig. 1) shows splenomegaly with multiple hypodense peripheric lesions with triangular morphology dispersed throughout the spleen, suggestive of splenic infarctions and presence of a cyst in the right liver lobe.

Arteriography of abdominal aorta and splenic artery (Fig. 2). Shows on A (early phase) irregular aspect of the intraparenchymal small arteries of the spleen with multiple areas of stenosis and dilatation, as well as multiple intraparenchymal microaneuryms. Late phase (B) demonstrates heterogenous contrast-enhancement with radiolucent cortical areas, diagnostic for multiple areas of splenic infarction.

Arteriography of the renal and hepatic arteries showed no abnormalities.

Radiological diagnosis

Based on the arteriographic finding of multiple intraparenchymal splenic microaneurysms, combined with the clinical presentation of fever and weight loss, the patient was diagnosed with *polyarteritis nodosa*. Immunosuppressive therapy was started with administration of prednisone.

Discussion

Polyarteritis nodosa (PAN) is a progressive inflammatory vasculitis of small- and medium-sized arteries that leads to necrosis and destruction of the wall of vessels. Thus, PAN belongs to the pathologic group of necrotizing vasculitis which also includes other diseases such as Churg-Strauss syndrome, Kawasaki disease and Wegener granulomatosis.

PAN is a rare condition and predominantly affects people in their fourth to sixth decade of life, with the male-to-female ratio being 2-3:1.The exact cause is unknown, but studies suggest that PAN is probably mediated by deposition of immune complexes. Viruses play a role in the pathogenesis in some cases, most notably hepatitis B virus.

Many of the clinical symptoms are related to organ ischemia secondary to arterial branch occlusions.

The kidneys are most frequently affected (in 70-80% of cases). Renal failure can occur because of multiple infarcts; hypertension can develop from renal artery involvement. Cutaneous manifestations occur in approximately 40% of affected patients and include palpable purpura, livedo reticularis and ischemic ulcers. Peripheral neuropathies (mononeuritis multiplex) are noted in 50% of patients and are often symptomatic early. One half of patients complain of arthralgia or myalgia. Blind muscle biopsies reveal 30-50% arteritis.

Gastro-intestinal involvement is noted in 15% of individuals with PAN, with ischemia occurring most often in the small intestine. The heart, testicles, lung and spleen are rarely involved.

Early diagnosis and treatment of PAN are necessary to prevent serious organ damage. Ten diagnostic criteria of PAN have been classified by the American College of Rheumatology and include weight loss, diffuse myalgias, mononeuropathy and hypertension. A positive angiogram with typical findings is one of the ten criteria. PAN is diagnosed if at least three of the ten criteria are present.

A definitive diagnosis may be made by performing tissue biopsy from a symptomatic organ site. Arteriography is the preferential imaging technique for diagnosis with the most common findings being multiple saccular intraparenchymal microaneurysms, areas with intermittent smooth tapering and dilatation, and occlusions. The latter can result in organ infarcts which can be detected easily by CT. The main target organs are the kidneys and the liver. The arteriographic findings are not pathognomonic and can be found in any necrotizing vasculitis as well as in some cases of drug abuse.

Without treatment almost all patients die within 2 to 5 years. Immunosuppressive therapy is effective and has been reported to increase 5-year survival rates to 48%.

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