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CASE REPORT

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Asymptomatic appearance of splenic infarction in Wegener's granulomatosis

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Summary

Splenic involvements in Wegener's granulomatosis (WG) are rarely diagnosed ante-mortem, while an autopsy is able to reveal a high rate of spleen lesions (78–100%). To date, there have been a few reported cases of splenic abnormalities in WG, including: splenomegaly, capsular adhesion, dysfunction and infarction. We reported a case of biopsy-verified WG with radiological evidence of diffuse spleen infarction despite the lack of any clinical symptoms. We concluded that due to a potential risk of severe hemorrhagic complications when anticoagulant therapy is necessary, radiological assessment of spleen should be performed regularly in this group of patients, particularly because spleen involvement can be asymptomatic.

Key words: Wegener's granulomatosis • spleen • splenic infarction

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Background

Wegener's granulomatosis (WG) is a systemic necrotizing vasculitis which affects small and medium-sized vessels and is associated with granuloma formation. Typically, the upper and lower respiratory tract and kidneys are involved. Clinical manifestations of WG are related to the location of lesions, their extensiveness and severity of the disease [1].

Splenic involvements are rarely diagnosed ante-mortem. To date, there have been a few reported cases of splenic abnormalities in WG, including: splenomegaly, capsular adhesion, dysfunction and infarction [2,3]. However, an autopsy revealed spleen lesions in high rate of patients (78–100%), even the asymptomatic ones, ante-mortem [4,5].

Case Report

A 34-year-old, non-smoking male patient treated with cyclophosphamid and prednisone for Wegener's granulomatosis (WG) was admitted to our hospital for a routine follow-up, at 1 year after starting the treatment.

The diagnosis of the disease was based on clinical manifestation (predominantly a massive lung involvement accompanied by relatively minor changes within the upper respiratory tract), mild erytrocyturia, elevated positive c-antineutrophil cytoplasmic antibodies (c-ANCAs – 194 RU/ml) and a histopathologically confirmed presence of vasculitis in the tissue obtained from bronchial infiltration.

During treatment, a clinical and radiological improvement was observed.

During hospitalization, the patient was in a rather good general condition, he had no fever, and had an episodic mucopurulent productive cough. He denied abdominal complaints on admission or in the past. Laboratory tests revealed an elevated white blood count (14.3×10^9/l, granulocytes – 74%, lymphocytes – 16.5%, monocytes – 8%, eosinophils – 1%) and no anemia. The platelet count was – 469×10^9/l, and the erythrocyte sedimentation rate (ESR) was 51 mm/h. C-reactive protein (CRP) was elevated to 34.9 U/l. Blood urea, creatinine and electrolytes fell within the normal range. Urinalysis revealed: 15–20 erythrocytes and 10–15 leucocytes per high-power field, with a negative culture for possible pathogens.

The level of c-ANCAs was elevated to 67.3 RU/ml. Anticardiolipin antibodies (ACA) were negative. Fibrinogen

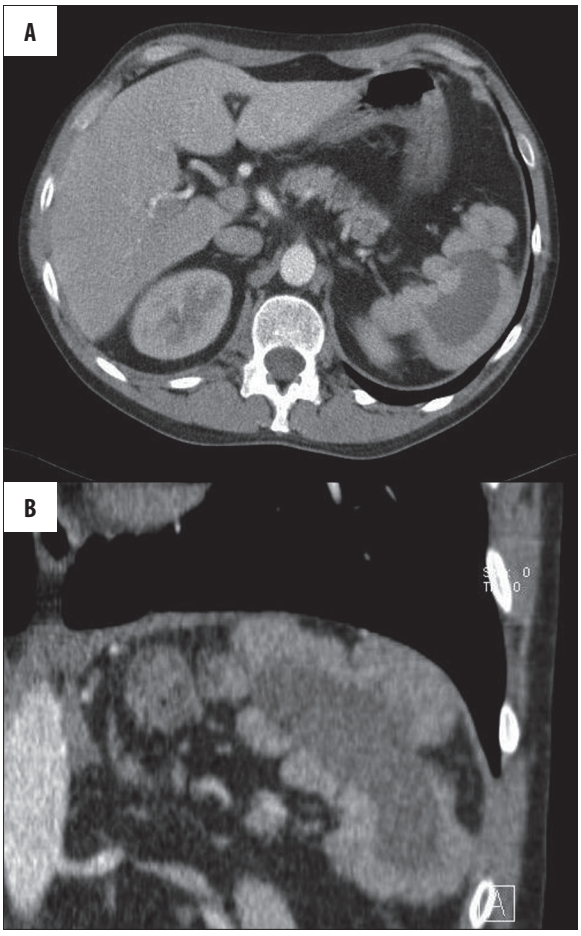

Figure 1A, B. Chest CECT unexpectedly revealed a slightly enlarged spleen with a central well-defined hypodense area involving almost 80% of the spleen.

degradation product – D-dimer – was evaluated at 834 μg/l (normal range of 68–494), fibrinogen – 6.34 g/l (normal range of 1.8–3.5).

The chest X-ray and contrast-enhanced computed tomography (CECT) of the thorax revealed no signs of WG progression. A coincidentally included fragment of the upper abdomen showed a slightly enlarged spleen (about 120×52 mm) with polycyclic margins and a central hypodense irregular area occupying almost 80% of the spleen (Figure 1A, B). A comparison with the previous chest CT (performed 10 months ago) showed enlarged spleen, with no enhancement except for the subcapsular area (Figure 2). An ultrasound (US) examination of the abdomen showed spleen with lobular margins, but there was no well-defined focal area, only heterogeneous echotexture of spleen parenchyma (Figure 3). The above described lesions suggest spleen infarction ensuing from WG and healing process with secondary overgrowth of splenic parenchyma.

Discussion

Reports of splenic involvements, particularly infarctions in WG patients, are rare. There are only a few cases published to date [3].

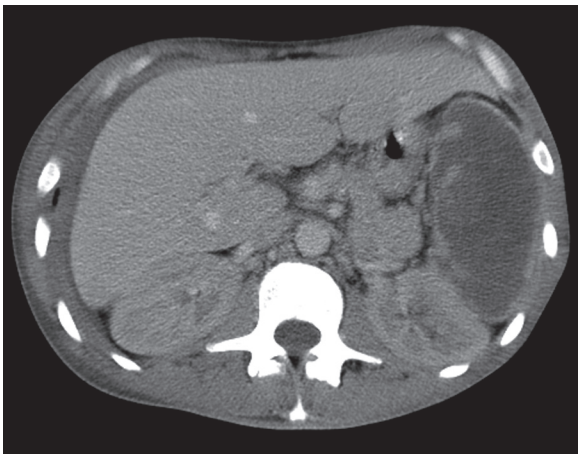

Figure 2. Chest CECT performed 10 months earlier showed an enlarged spleen with no enhancement except for the subcapsular area.

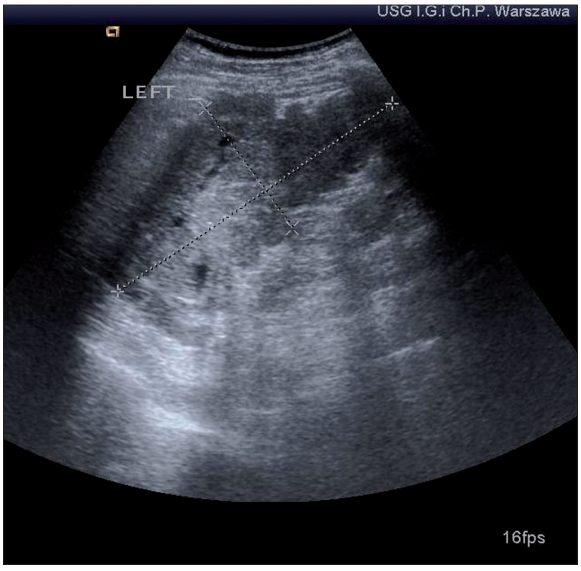

Figure 3. US of the spleen showed a heterogeneous echotexture of the spleen parenchyma (see irregular margins of the spleen).

Splenic infarction resulting from an occlusion of the distal parenchymal splenic arteries may be focal or diffuse. A diffuse infarction appears on CT scan as a massive hypodense lesion, as it was in the presented case. Splenic infarction shown on US examination may present a wide range of nonspecific manifestations including wedge-shaped or round, hypoechogenic to echo-free areas. It is difficult to determine the age of an infarction because of possible coexisting edema, necrosis, bleeding and inflammation, especially in clinically silent events [6]. MRI with gadolinium has also been successfully employed to show diffuse infarctions in WG patients [7].

Splenic infarction in WG usually remains asymptomatic. In some cases, a nonspecific abdominal pain was described [3,8]. In some WG patients, splenic infarction may lead to complications like: abscess, bleeding, or even rupture [9–11]. The risk of developing severe infections increases with the use of immunosuppressive treatment [12].

Some data suggest that antithrombotic treatment entails an elevated risk of bleeding in patients with WG. When anticoagulation therapy is necessary in this group of patients, CT of the abdomen should be systematically repeated and if splenic infarction is discovered, splenectomy should be considered [9].

Splenic infarction in patients under 40 years of age is often associated with hematological disorders. A case of splenic infarction in vasculitis such as polyarteritis nodosa coexisting with anticardiolipin antibodies (ACAs) was reported [13]. In our case, we did not observe any hematological disturbances, and ACAs were negative.

Conclusions

Splenic infarction should be considered in WG patients, even if they are asymptomatic. We concluded that radiological assessment of the spleen should be performed regularly in this group of patients, especially in the presence of an increased risk of hemorrhagic complications.

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