PHOTO QUIZ

Necrosis of small intestine

Y. Erzin[1*], A.S. Yalın[2], A.S. Erdamar[3], A.F. Celik[1]

Departments of [1]Gastroenterology, [2]Internal Medicine and [3]Pathology, Istanbul University, Cerrahpasa Medical Faculty, Istanbul, Turkey, *corresponding author: tel.: +90 (0)532-265 50 08, fax: +90 (0)216-467 88 70, e-mail: dryusuf@doruk.net.tr/dryusuferzin@yahoo.com

CASE REPORT

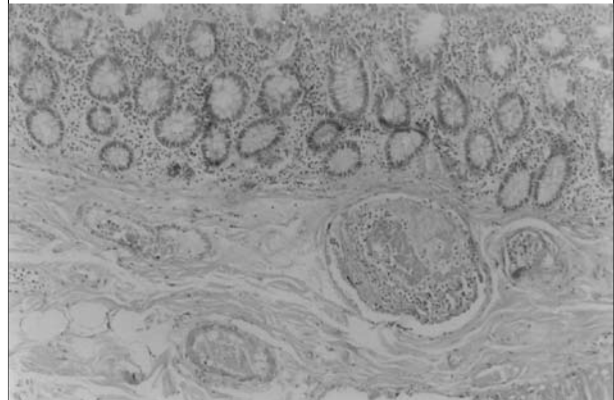
A 52-year-old man with no prior symptoms was admitted to the emergency unit with troublesome abdominal pain due to ileus and underwent an extensive surgical resection, as most of his small intestine and ascending colon had become necrotic due to occlusion of the superior mesenteric artery (SMA). The macroscopic examination of the resected material showed transmural haemorrhagic necrosis of bowel loops and multiple fresh thrombi in multiple small branches of SMA. The histopathological examination revealed the total absence of any atherosclerosis or vasculitis (*figure 1*).

In his past medical history, he had no oral – genital ulcers, rash, photosensitivity, arthralgias – arthritis, medications and no prior abdominal surgery, but he had been smoking one pack of cigarettes daily for 20 years. His physical examination was unremarkable but he was pale. Looking for the aetiology of the mesenteric thrombus, hepatitis B surface antigen, antihepatitis C antibody, Venereal Disease Research Laboratory test, cryoglobulin, p-ANCA, c-ANCA, anti-PR3, anti-MPO, FANA, anti-double stranded DNA, RF, C_3, C_4 levels were checked and found to be within normal limits. Ham's test and sucrose lysis test were negative and molecular analysis showed no factor V Leiden, or prothrombin G20210A mutations. Protein C, S, antithrombin III levels were also normal. A skin pathergy test, ophthalmological examination and an electromyography of the extremities revealed no pathology either. Transthoracic and subsequent transoesophageal echocardiography showed no valvular abnormalities or intracardiac thrombi. The lupus anticoagulant was found negative but the level of anticardiolipin antibodies (aCL) of class G was 20.3 GPL (normal 0-8) and 13.5 MPL of class M (normal 0-8). Eight weeks later, before the patient's discharge, the aCL levels were checked and found to be above the normal level again.

WHAT IS YOUR DIAGNOSIS?

See page 401 for the answer to this photo quiz.

Figure 1. *Recent thrombi formation is seen in submucosal vessel lumen (HE x200)*



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ANSWER TO PHOTO QUIZ (ON PAGE 400)

(ON PAGE 400)

NECROSIS OF SMALL INTESTINE

DIAGNOSIS

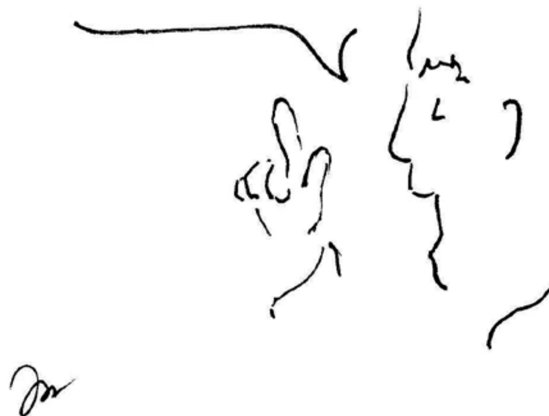
The diagnosis of primary antiphospholipid syndrome (APS) was made and the patient was put on warfarin treatment. APS is defined by the presence of antiphospholipid antibodies with vascular thrombosis and/or pregnancy morbidity.[1] When not associated with systemic lupus erythematosus (SLE), other collagen diseases or ingestion of medications, the condition is called primary APS.[2] APS, showing a female gender predominance and mostly accompanied by SLE, may affect any organ in the body and display a broad spectrum of manifestations such as livedo reticularis, thrombocytopenia, leucopenia, stroke, epilepsy, chorea, myocardial infarction and arterial and venous thrombosis, with particularly cerebral ischaemic attacks and deep venous thrombosis as the most frequent thrombotic events.[3] Unfortunately, most of the patients with APS are prone to recurrent thrombosis, generally occurring at the same site as the previous one, and treatment with high-intensity warfarin (producing an INR of >3) with or without low-dose aspirin has been significantly found to be the most effective prophylactic regimen in preventing recurrences.[4]

All patients, especially the young ones with thrombotic events and no identifiable risk factors, should be tested for the presence of aPL antibodies. After identifying the syndrome, the second step should be to determine whether it is primary or secondary as an underlying true vasculitis sometimes needs to be treated more aggressively with immunosuppressive drugs.

REFERENCES

1. Wilson WA, Gharavi AE, Koike T, et al. International Consensus Statement on Preliminary Classification Criteria for Definite Antiphospholipid Syndrome: report of an international workshop. Arthritis Rheum. 1999;42:1309-11.
2. Asherson RA, Khamashta MA, Ordi-Ros J, et al. The 'primary' antiphospholipid syndrome: major clinical and serological features. Medicine (Baltimore). 1989;68:366-74.
3. Marai I, Levi Y, Godard G, et al. Following 90 patients with antiphospholipid syndrome with antibody titers and correlations with clinical manifestations: symptoms of the disease, a new antibody and correlations with clinical manifestations in the Israeli population. Harefuah. 2001;140:495-500.
4. Munther A, Khamashta MA, Cuadrado MJ, et al. The management of thrombosis in the anticardiolipin-antibody syndrome. N Engl J Med. 1995;332:993-7.

If you don't think of it,
you cannot diagnose it!*



*adapted from Johan Cruyff
(contemporary philosopher
and former soccer player)

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