Case Report

Multiple Spontaneous Visceral Hematomas Revealing Polyarteritis Nodosa

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ABSTRACT. Polyarteritis nodosa is a rare life-threatening disease characterized by necrotizing vasculitis of small and median arteries. We describe the exceptional case of a 28-year-old man with successive spontaneous visceral hematomas of the kidney, bladder, and liver. Arteriography was performed for a recent spontaneous hepatic hematoma and a microaneurysm was detected, allowing the diagnosis of polyarteritis nodosa and prescription of appropriate treatment. (J Rheumatol 2004;31:1858–60)

Key Indexing Terms:
POLYARTERITIS NODOSA VISCERAL HEMATOMA

Polyarteritis nodosa (PAN) is a rare disease characterized by necrotizing vasculitis of small and medium size arteries[1]. The clinical symptoms that usually reveal PAN are neuritis, arthralgia, myalgia, cutaneous lesions, orchitis, and abdominal pain[2]. Prompt diagnosis is important because PAN can be life-threatening: severe organ manifestations include congestive heart failure, cerebrovascular events, gastrointestinal (GI) tract hemorrhage, and malignant hypertension. We describe a patient with an unusual presentation of PAN, revealed by successive spontaneous visceral hematomas involving the kidneys, bladder, and liver.

CASE REPORT

A 28-year-old Bulgarian man was admitted to our department for exploration of repeated spontaneous hematomas. Symptoms began in March 2000, with isolated, violent lumbar pain, and no decline in general health status. Ultrasound and computed tomography (CT) scans revealed a left perinephritic hematoma. Surgery was performed. Pathological analysis confirmed the diagnosis and identified no other abnormality. The pain disappeared with standard analgesic treatment, and he resumed all regular activities. One year later, he suffered the same symptoms, with right kidney involvement, and the same course. In June 2001, he suffered spontaneous bleeding of the bladder, as revealed by macroscopic hematuria, with no renal insufficiency. This bleeding stopped spontaneously within a few days. One year later, he reported spontaneous pain in the right upper abdominal quadrant, and CT scan revealed the presence of a hematoma in the liver. Investigations over this 2-year period revealed no coagulation or immunological abnormalities, but biological examinations showed repeated signs of transient inflammation with increased erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP). Histological analysis of a kidney sample removed during surgery revealed no inflammatory disease, specific infiltration, or vessel abnormalities other than hematoma.

By the time of admission to our hospital in March 2003, our patient had lost 20 kg over 2 years (weight 58 kg; height 1.75 m); he had had fever and abdominal pain for 4 days. On physical examination, he was pale and sweating. His blood pressure was 130/80 mm Hg, pulse 100/min, and temperature 38.8°C. Diffuse tenderness was noted on abdominal pressure, without palpable abnormality. Neurological and cardiopulmonary examinations were normal. Laboratory investigations gave the following results: white blood cell count 14,000/mm^3, ESR 74 mm/h, CRP 240 mg/l (normal < 5 mg/l), hemoglobin concentration 12.6 g/dl, aspartate aminotransferase 402 IU/l, alanine aminotransferase 648 IU/l, creatininemia 67 µmol/l, and absence of proteinuria. Hemostasis test results: prothrombin time 104%, activated partial thromboplastin time 40 s (normal = 40 ± 5), lupus anticoagulant absent; and factor VIII, IX, and von Willebrand levels were normal. Blood cultures, urinalysis, and tests for tuberculosis (skin test and gastric culture), human immunodeficiency virus, hepatitis C virus, and hepatitis B virus (last generation ELISA tests) were negative. No autoantibodies, including antinuclear, anti-dsDNA, antiphospholipid, antiextractible nuclear antigens, antineutrophil cytoplasmic antibodies, rheumatoid factor, or cryoglobulinemia were detected. Thoracic and abdominal radiographs, electrocardiogram, and echocardiography results were normal. Thoracoabdominal CT scan showed 2 recent intrahepatic hematomas, visible as spontaneous hyperdensity lesions (Figure 1). The clinical presentation and the CT results prompted us to perform arteriography, which revealed microaneurysm in the splenic and hepatic arteries (Figure 2), leading to a diagnosis of PAN. Treatment was begun immediately, with 3 pulses of 15 mg/kg methylprednisolone over 3 days, followed by 1 mg/kg prednisolone daily. He also received an infusion of cyclophosphamide (0.7 g/m^2).

Two weeks later, he was seen as an outpatient. He had no pain, normal ESR and CRP, and his hepatic variables had returned to normal. He then went back to Bulgaria to continue treatment with monthly cyclophosphamide infusions and oral prednisone; 6 months later he is still doing well with normal examination and biological tests.

DISCUSSION

PAN is a systemic panmural necrotizing vasculitis of unknown origin with manifestations in multiple organ systems. Renal and GI involvement, due to multiple infarc-

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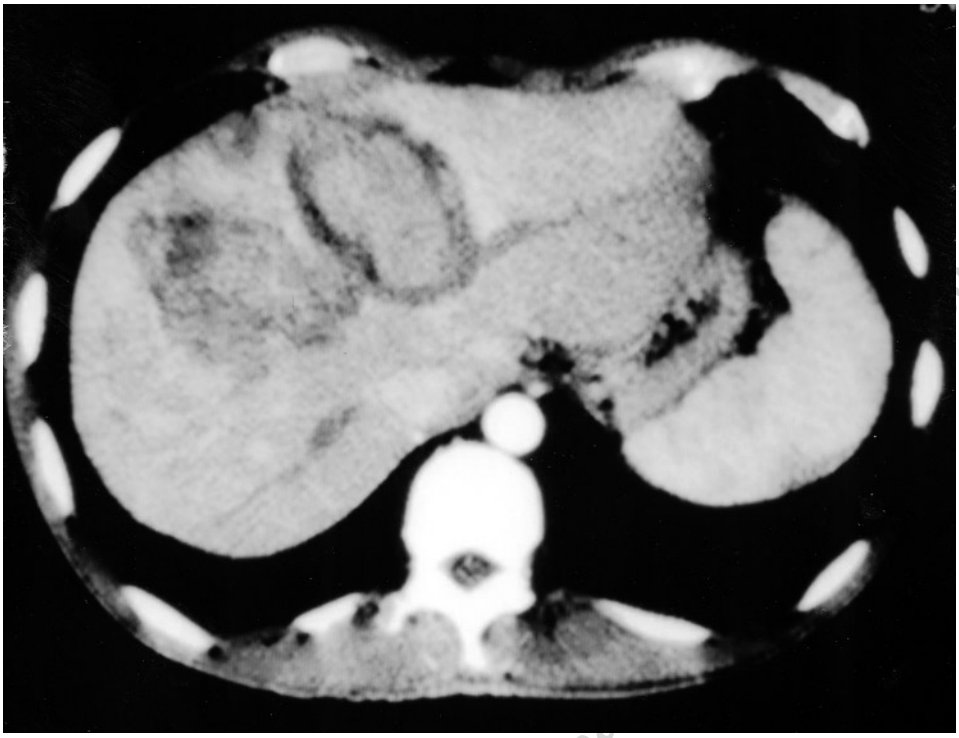

Figure 1. Helical computed tomography of the liver after bolus injection of contrast agent: multiple hepatic hematomas (arrow) with spontaneous increased signal intensity and no tumoral enhancement on the arterial phase.

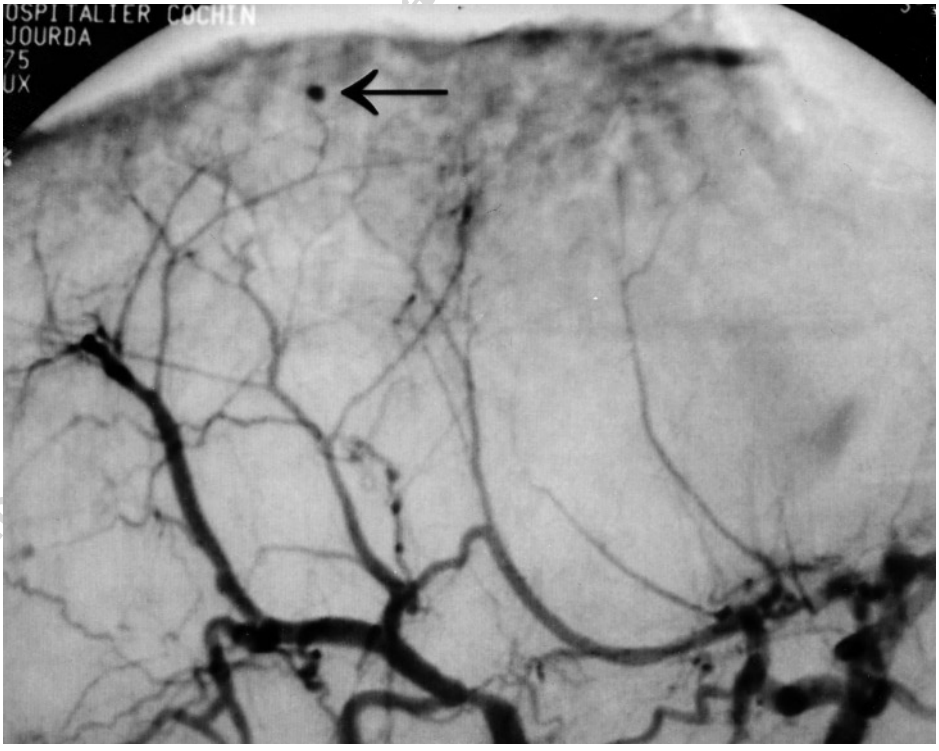

Figure 2. Selective angiography of the hepatic artery: small aneurysms on distal branches.

tions and aneurysms, is characteristic of PAN. Hematoma is an exceptional complication. This is the first reported case of successive hematomas of the renal and GI organs of a single patient.

Kidney involvement in PAN generally consists of proteinuria, hematuria, renal insufficiency, and malignant hypertension due to arcuate, intralobular renal artery lesions. Perinephritic hematoma associated with PAN was first described in 1908, and 55 to 60 cases have now been reported[3]. Spontaneous subcapsular and perirenal bleeding secondary to PAN may occur in 4 to 13% of cases[3], with an unexplained high prevalence (15% of PAN patients having perirenal hematoma) in Turkey. The other possible causes of spontaneous perirenal hematomas are renal cell carcinoma in more than 50% of cases, angiomyolipoma, and polycystic kidneys[4]. In most of these cases, PAN has already been diagnosed. In contrast, in our case, hematomas were the initial sign, resulting in delayed diagnosis. PAN generally occurs in young men with a recent history of hypertension, flank or abdominal pain, and anemia[5]. In most cases, bleeding is secondary to rupture of a microaneurysm. The prognosis is poor, and more than 50% of the patients are likely to die from the condition[6].

Intrahepatic hematoma is rare, with only 9 cases reported to date[7]. The mean age of these patients was 33 years (20–77), and 7 of these 9 patients were men. In 3 patients, PAN was diagnosed before the hematoma event, and 4 patients had PAN symptoms before this event. In 8 of these 9 cases, hematomas were located in the right hepatic lobe, as in our case, the remaining case having a hematoma in the left lobe[7]. The other possible causes of spontaneous intrahepatic hematoma are benign tumors, which account for 24% of hematomas, malignant tumors, which account for 35% of hematomas, vascular abnormalities, thrombocytopenia, and vasculitis. None of these patients died from hepatic hemorrhage.

Medical treatment of severe PAN includes corticosteroids and cyclophosphamide. However, in cases of life-threatening visceral bleeding, some authors have reported the use of selective embolization[5] or radical surgery. Selective embolization is of potential value because it can be performed at the same time as arteriography if active bleeding is detected. The mortality rate for patients undergoing radical surgery is high (50%); better results have been reported with embolization, but there are too few cases for firm conclusions to be drawn[5]. Bleeding was not severe in our patient, and medical treatment was initiated without surgery.

The gold standard for PAN diagnosis is the pathoanatomic observation of necrotizing inflammation of a medium size artery. However, even if not pathognomonic, the most common type of vasculitis encountered on angiography is PAN. Indeed, for cases of vasculitis in which PAN was suspected, angiography was shown to have a sensitivity of 89%, a specificity of 90%, and, above all, a negative predictive value of 98% in a retrospective study of 748 renal angiographies[8]. The main arterial characteristics are stenoses, occlusions, and aneurysms of various sizes, numbers, and distributions: microaneurysms are the most widely recognized lesion typical of classic PAN. Although rare, visceral aneurysms are observed in other diseases, such as severe hypertension, rheumatoid arthritis, systemic lupus erythematosus, temporal arteritis, renal carcinoma, and fibromuscular dysplasia[8]. For our patient, hereditary connective tissue disorders were first suspected, but the review of the various previous histological analyses did not support this hypothesis. Moreover, the context of fever, weight loss, and blood inflammation was evocative of an inflammatory disease, and the discovery of microaneurysm was then suggestive of PAN, even though he did not fulfil the classification criteria of the American College of Rheumatology[9].

We describe the exceptional case of a 28-year-old man with successive spontaneous visceral hematomas of the kidney, bladder, and liver. Arteriography was performed for a recent spontaneous hepatic hematoma. Microaneurysm was detected, allowing the diagnosis of polyarteritis nodosa and the prescription of appropriate treatment. PAN diagnosis may thus, in rare cases, be revealed by spontaneous visceral hematomas.

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