Isolated Unilateral Kidney Vasculitis

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Polyarteritis nodosa is a necrotizing vasculitis that primarily involves medium and small sized arteries. In most cases it is a multi-systemic disease, is rarely confined to a single organ, and if so is referred to as an isolated PAN.

Isolated PAN is a well-documented pathology involving various organs such as the gallbladder, the appendix, the female genital tract, and the prostate. Isolated PAN of a kidney is a rare condition and can be easily misdiagnosed. In the patient presented here, severe gross hematuria was the presenting symptom of isolated vasculitis of the right kidney and there was no clinical evidence of systemic involvement. To the best of our knowledge this is the first reported case of isolated kidney vasculitis presenting with severe hematuria.

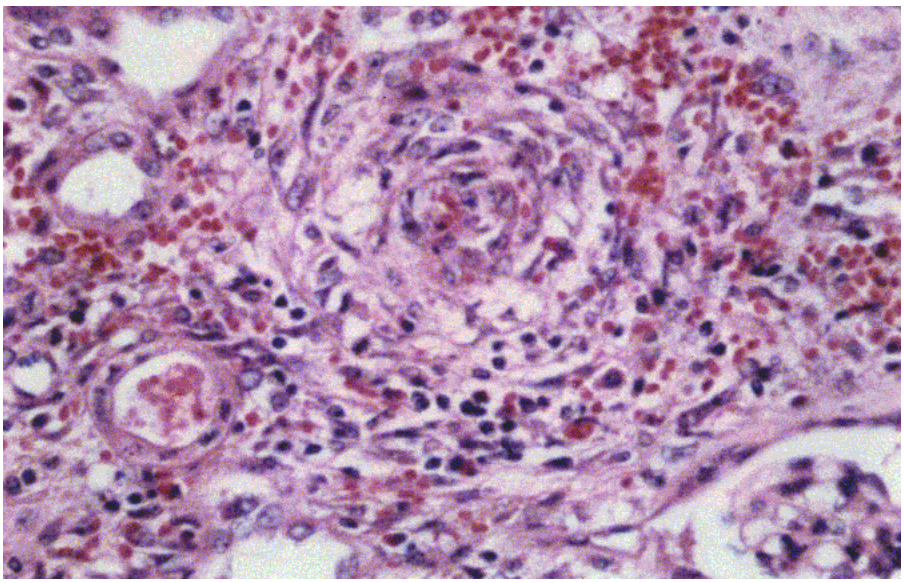

Figure A. Small vessel vasculitis, with prominent bleeding into the interstitial space and tubules. Intimal thickening, marked luminal narrowing and eosinophillic infiltration to the vessel wall are seen.

Case Description

A 67-year-old woman was referred to the emergency room because of a sudden and severe gross hematuria. Two years prior to her admission she had suffered from recurrent polyarthralgia, without any other systemic manifestations. X-ray studies showed no evidence of articular erosions and she was seronegative for rheumatoid factor and other autoantibodies. An infectious disease workup including serology for hepatitis B and C, parvoviruses and others was negative. During the 6 months prior to admission she felt well and had no active articular complaints. Upon admission she was mildly pale, but in a generally good condition. Her blood pressure and hemodynamic status were normal. No lymphadenopathy or other organomegaly was found. Her cell blood count, C-reactive protein, erythrocyte sedimentation rate, kidney and liver function analyses were all normal. All coagulation analyses including partial prothomboplastin time, lupus anticoagulant and anticardiolipin antibodies were normal.

Intravenous pyelography showed a poor nephrogenic and almost non-secreting silhouette (?) of the right kidney, whereas the left one was normal. During a cystoscopy, the bladder looked normal, but continuous oozing of fresh blood could be seen from the right ureter. A retrograde ureteroscopy showed that the bleeding originated from the upper part of the tract, and retrograde pyelography documented multiple infarcts of the right kidney. Due to the severe continuous hematuria, an emergency right nephrectomy was undertaken. She recovered rapidly and the hematuria ceased.

Macroscopically the kidney had widespread ischemic infarcts, whereas the histological examination revealed severe vasculitis involving mostly medium and small-sized arteries. There was prominent bleeding into the tubules along with an interstitial inflammatory infiltrate. Intimal thickening and a predominant eosinophilic infiltration of the vessel walls were seen in association with fibrinoid necrosis [Figure A].

A comprehensive evaluation to exclude possible systemic involvement was later performed. Multiple urinalyses were normal (no erythrocytes or casts were present), erythrocyte sed-

PAN = polyarteritis nodosa

imentation rate, complete blood count, and repetitive kidney and liver function analyses were all normal. A detailed immunological evaluation including antinuclear antibodies, antineutrophil cytoplasmic antibodies, C_3 and C_4 levels, and protein electrophoresis were all normal. Repeated coagulation studies including lupus anticoagulant, anticardiolipin antibodies, β2-glycoprotein1 and antithrombin 3 antibodies were all negative. Repeated analyses for antibodies to hepatitis B and C viruses were not found. Lung computed tomography scan was normal. Abdominal CT demonstrated normal liver and spleen and showed no lymphadenopathy. The patient's neurological status including the fundus was normal. Due to the patient's satisfactory physical condition, she was not treated with steroids or other immunosuppressive drugs. In the year since surgery the patient is in good health with no evidence of any systemic disease.

Comment

Isolated polyarteritis nodosa was first described by Plaut [1], who reported on focal arteritis in 22 appendices without any evidence of involvement other than that of the affected organ. PAN-like lesions confined to one organ are well recognized and have been reported in the gallbladder, the breast, the prostate, the epididymis, and the central nervous system. Idiopathic crescentic glomerulonephritis presenting as a renal-limited vasculitis (without systemic manifestations) has rarely been reported [2,3]. To the best of our knowledge the case described here is the first in which severe bleeding was present.

Immunohistochemical studies demonstrated that in both systemic and isolated PAN the inflammatory cells are composed mainly of T lymphocytes and macrophages. This suggests that isolated vasculitis is primarily an immune response, implicating mainly the cellular mediated response [4]. The causes of isolated or systemic vasculitis are unknown. Organs involved in isolated PAN are accessible to foreign antigens or infectious agents [5].

In kidney vasculitis, a retrograde exposure to an infectious agent could selectively induce local immunostimulation in one kidney without a systemic immunological reaction. However, no evidence for such an infection was found in our patient. The eosinophil infiltration observed in the histological examination is compatible not only with PAN, but also with Wegener's granulomatosis and with Churg-Strauss vasculitis. However, the lack of positive features for Wegener, such as granulomas or positive antineutrophil cytoplasmic antibodies, and the negative history of allergy/asthma suggest PAN to be the most likely diagnosis. One could consider that the patient's isolated PAN as part of rheumatoid arthritis (due to the history of polyarthralgia), however this does not seem likely due to the fact that these patients often have severe arthritis and very high rheumatoid factor titers. Therefore we presume that this is not a direct rheumatoid arthritis-associated isolated vasculitis. Moreover, the fact that our patient has been completely healthy during one year of follow-up, together with the absence of any American College of Rheumatology criteria for the classification of systemic vasculitis, points to the diagnosis of isolated unilateral kidney vasculitis.

Previously reported cases of isolated PAN usually resolved after the excision of the affected organ or following a short course of corticosteroids. Our case of isolated vasculitis is unusual due to the presence of severe bleeding, which resolved following unilateral nephrectomy without the need for immunosuppressive therapy.

References

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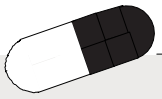
Capsule



Flu shot

The value of the annual flu vaccine is directly related to how well it can be determined which flu strain will actually predominate in the seasonal onslaught. Bush et al. have analyzed the molecular evolution of flu strains over recent years and arrived at a means to predict more accurately which strain will emerge in the next flu season. Their process, which involves identification of codons under positive selection to change, successfully predicted in retrospective tests the flu strains for the last 8 years.

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