Unilateral Complet Renal Artery Occlusion Due to Polyarteritis Nodosa : An Unusual Presentation

POLİARTERİTİS NODOSA'YA BAĞLI TEK TARAFLI KOMPLET RENAL ARTER OBSTRÜKSİYONU : NADİR BİR OLGU

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Summary

Polyarteritis nodosa is a disease of small and medium-sized arteries. It was first described by Kussmaul and Maier in 1866. Polyarteritis is an uncommon disease. The annual incidence rate for PAN-type systemic vasculitis in a general population is 4.6-77/1 000 000. Polyarteritis may present in a variety of ways. There is a spectrum of severity from mild, limited disease to progressive disease which may be fatal. Cutaneous lesions occur in 25-60% of patients with polyarteritis. Renal infarction and malignant hypertension are commonly found in patients with renal involvement. In this report, we present a case of classical polyarteritis nodosa (PAN) with unilateral complet renal artery occlusion, resulting in renal infarction and malignant hypertension. Complet renal artery occlusion is a rare severe complication of PAN. It is important to make diagnosis quickly, as untreated disease may progress with time to involve organs, and the extent of their involvement determines the outcome. Polyarteritis should be suspected in such a patient that was described by us.

Key Words : Polyarteritis Nodosa, Renal Artery Occlusion, Malignant Hypertension

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Özet

Poliarteritis nodosa (PAN), küçük ve orta çaplı damarları tutan bir hastalıktır. İlk olarak 1866'da Kussmaul ve Maier tarafından tanımlanmıştır. PAN, seyrek görülen bir hastalıktır. Yıllık insidensi 4,6-77/1 000 000 olarak bilinmektedir. PAN klinikte değişik tablolarla seyredebilir. Klinik seyri ; hafif sınırlı hastalıktan, fatal seyredebilen progressif ciddi hastalığa kadar geniş bir spektrum gösterebilir. Olguların %25-60'ında cilt lezyonları gözlenir. Böbrek tutulumu mevcut olgularda sıklıkla renal enfarktüs ve malign hipertansiyon görülür. Renal arterin tam tıkanıklığı, PAN'ın nadir görülen ciddi bir komplikasyonudur. Tanıyı zamanında koymak son derece önemlidir. Tedavi edilmeyen olgular zaman içinde ilerleme gösterir. Hastalığın tutulum yaygınlığı ise prognozu belirler. Tanımladığımız olguya benzer hastalarda, ön tanıda mutlaka PAN düşünülmelidir.

Anahtar Kelimeler : Poliarteritis Nodosa, Renal Arter Tıkanıklığı, Malign Hipertansiyon

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Case Report

A 21 years old male was referred to our department with cutaneous lesions (palpable purpura at lower extremities and glans penis necrosis) in May 2000. These cutaneous lesions were lasted two months. There was no systemic sign and symptoms. The patient did not use any medication before admission. The results of all tests for systemic involvement (urine analysis, leucocyte and platelet count, routine serum biochemistry, viral serology, ANA, serum Ig levels etc.) were normal, except for a finding of normocytic anaemia with normal serum iron levels. Physical findings showing palpable purpura and glans penis necrosis suggested a positive diagnosis of cutaneous necrotizing vasculitis. Skin biopsy of cutaneous lesions at lower extremities confirmed the diagnosis of leukocytoclastic vasculitis.

A diagnosis of cutaneous vasculitis without systemic involvement was made, and the patient started on a regimen of azathiopurine and oral corticosteroids. He responded very dramatically to

Table 1. Blood and Urine Analysis

Urine	: 1+ proteinuria
Hemoglobin	: 12.7 g/dl
Leucocyte count :	: $9.3x10^3$/dl
Erytrocyte sedimentation rate	: 114 mm/h (abnormal)
HbsAg	: Negative
Anti-HIV	: Negative
ANA	: Negative
Antitrombin III	: 25.9 mg/dl
PTZ	: 0.946 INR
Ig G	: 2113 mg/dl (abnormal)
Ig A	: 358 mg/dl
Ig M	: 270 mg/dl (abnormal)

therapy, with all cutaneous lesions returning to normal within 3 weeks. The patient used these medications for three months.

The patient was admitted acutely with headache in September 2000. On admission, he looked very ill. He was hypertensive with a blood pressure of 220/130 mmHg. Physical examination was unremarkable except for a 2/6 systolic murmur at the apex of the heart radiating to the left axillary region. Fundus examination (ophtalmology consultation) was confirmed grade III hypertensive retinopathy. There was no other abnormal findings apart from severe hypertension (BP:220/130 mmHg). Urine analysis showed proteinuria. The other results including urine analysis were between normal limits. BUN and serum creatinine values were also between normal limits. The following investigations were performed :

His current medications are prednisolone 60 mg/day, methyle-dopa 1 g/day, doxazosin 4 mg/day, metoprolol 100 mg/day, amlodipine 10 mg/day and clopidogrel 75 mg/day. His blood pressure came under control. After urologic consultation, it was decided that the only radical therapeutic option for this patient with renovascular hypertension was nephrectomy.

Discussion

The systemic vasculitides comprise a heterogeneous group of clinicopathologic entities characterized by inflamation and damage to blood vessels (1). Polyarteritis may present in a variety of ways. Virtually any organ may eventually be affected. In some cases, single organ involvement may be present alone and may remain limited, including isolated involvement of skin, peripheral nerves and visceral organs. Kidney involvement occurs in 75% of patients with PAN (2,3).

Occlusive vasculopathy potentially complicates most vasculitis with vasoconstruction, thrombosis and vascular cell proliferation (2). Classic PAN is usually characterized by vascular nephropathy without glomerulonephritis about 35% of all cases. Multiple renal infarctions, the consequence of vascular nephropathy, produce renal failure in classic PAN. Hypertension develops as a result of renal artery or less commonly, glomerular involvement. Renal infarction and malignant hypertension may frequently occur. Renal vascular hypertension is a late complication that can cause deaths in PAN. These vasculopathic complication of PAN are due to the effects of arterial inflammation (2,4).

Hoover et al reported a case of PAN involving only the main renal arteries. They presented this case as an unusual variant of PAN limited to both

Table 2. Other Procedures

Skin Biopsy at lower extremies	:	Leukocytoclastic vasculitis
DTPA renal scintigraphy (with captopril)	:	Left renal function decreased (%17)
DMSA renal scintigraphy	:	Impaired glomerular function, intrarenal stasis, left hypoplastic kidney, left renal paranchimal defect
Renal doppler ultrasonography	:	Supportive findings of renal vasculitis (lasting intrarenal artery acceleration time, decreased PI and RI)
Selective renal angiography	:	Complet left renal artery occlusion (collateraly circulation by lumbal arteries) (Figure 1,2,3)
Cranial MR angiography	:	Normal findings
Fluorescein fundus angiography	:	Supportive findings of hypertensive retinopathy

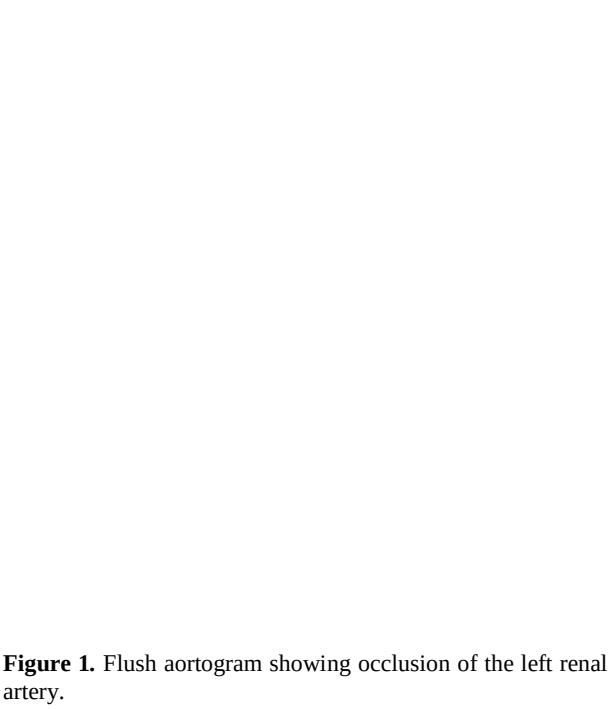

Figure 1. Flush aortogram showing occlusion of the left renal artery.

main renal arteries (5). Our case was presented by the left main renal artery occlusion. There was no any other arterial involvement. So renal functions wasn't impaired, but there was malignant hypertension and left renal regional infarction. Dasgupta et al reported a case of classic PAN who also had all laboratory features of the antifosfolipid syndrome (APS) (6). The association of APS with PAN is a rarity, although it has been described with PAN as well as a few other vasculitides. (7). So such a patient who had main arterial and venous thrombosis must be evaluated by a diagnostic approach to PAN and vasculitides. If we regarded our patient, we didn't observe any clinical and laboratory features of APS including strokes, migraine, livedo reticularis and thrombocytopenia in the presence of prolonged APTT etc.

There is often a long delay in diagnosis, often months and it is a problem that should be considered in many multisystem diseases which are otherwise diagnostically difficult and unexplained. Sometimes, generalized inflammation of medium and small arteries that leads to thrombosis or aneurysmal dilatation may occur. And the clinical course may be complicated by recurrent bleeding due to ruptured aneurysm and death at the early period of disease. Siegel et al reported such a case of PAN that was characterized by acute abdomoinal pain and death within 48 hours (8). Mas et al

Figure 2. Aortogram showing occlusion of the left main renal artery.

Figure 3. Selective left renal angiogram showing complet occlusion.

reported two cases of PAN that was complicated by spontaneous bilateral rupture of kidneys (3).

In conclusion, in patients with known necrotizing cutaneous vasculitis without any systemic involvement, clinical course may remain limited. But after a time, it may present in variety forms of systemic involvement. So such a patient must be carefully monitored. Our case is an unusual presentation of PAN that is limited to one main renal arteries.

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