Spontaneous Bilateral Rupture of Kidneys in Two Patients with Poliarteritis Nodosa

POLİARTERİTİS NODOZALI İKİ HASTADA SPONTAN BİLATERAL RENAL RÜPTÜR

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SUMMARY

Poliarteritis nodosa is a vasculitic syndrome that involves medium-sized and small muscular arteries. It was first described by Kussmaul and Maier in 1866. There is a multisystem involvement such as nephritis, peripheral neuropathy, skin rash, asymmetric polyarthralgia or arthritis (Kidney involvement occurs in about 75% of patients with poliyarteritis). Renal infarction and rupture of intrarenal arteries may occur. Renal insufficiency is the cause of death in about 50% of fatal cases. Hypertension may also develop. In this report we present two cases of renal involvement of PAN resulting in bilateral perirenal haematoma due to spontaneous rupture of intrarenal arterial aneurysms. We presented two case reports that had been diagnosed as PAN. Both of two patients came to the Gülhane Medical Faculty, because of fever, fatigue, weight loss, muscular pain and lower abdominal pain. In our two cases, the diagnosis was established by renal US, CT and DSA. Spontaneous renal haemorrhage or rupture are dramatic complications of PAN, as in our two cases, may be the first sign of disease.

Key Words: Poliarteritis nodosa, Spontaneous renal rupture

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ÖZET

Poliarteritis nodoza (PAN), küçük ve orta çaplı damarları tutan vaskülitik bir sendromdur. İlk olarak 1866'da Kussmaul ve Maier tarafından tanımlanmıştır. Nefrit, periferik nöropati, deri döküntüleri, asimetrik poliartralji, artrit gibi multisistem tutulumu mevcuttur. PAN'lı hastaların yaklaşık %75'inde böbrek tutulumu söz konusudur. PAN'lı olgularda, renal infarktüs ve intrarenal arter rüptürü meydana gelebilir. Ölümle sonuçlanan olguların yaklaşık %50'sinde ölüm nedeni böbrek yetmezliğidir.

Bu makalede, renal tutulumu mevcut 2 PAN'lı hasta sunulmuştur. Olguların her ikisinde de intrarenal arter anevrizmasının spontan rüptürü nedeniyle, bilateral perirenal hematom oluşmuştur. Her iki hasta da hipertansifti. Her iki hasta, ateş, halsizlik, kilo kaybı, kas ağrısı ve alt batında ağrı şikayetleri ile Gülhane Askeri Tıp Akademisi İç Hastalıkları Kliniğine başvurmuştur. Bu olgularda tanı, renal US, CT ve DSA ile konmuştur.

Spontan renal hemoraji veya rüptür, PAN'ın dramatik komplikasyonlarındandır. Her iki olgumuzda olduğu gibi hastalığın ilk bulgusu olabilir. PAN'ın bu ciddi ve nadir komplikasyonu genellikle intraparankimal anevrizma rüptürü sonucu oluşur. Olguların çoğunda tanı operasyondan sonra konur. Ancak makalemizde sunulan her iki olguda da tanı, noninvaziv yöntemlerle konmuştur. İkinci olgu, tanı konduktan sonra operasyona alınmıştır. Nefrektomi örneğinin patolojik incelenmesinde mikroanevrizmalar saptanmış ve tanı kesinlik kazanmıştır.

Sonuç olarak, spontan renal parankimal hemoraji saptanan her hasta, PAN yönünden değerlendirilmelidir.

Anahtar Kelimeler: Poliarteritis nodoza, Spontan renal rüptür

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CASE REPORTS

Case I: A 20 year old male patient was referred to the Gülhane Medical Faculty. He had pain on left lower abdomen of sudden onset. He didn't describe any abdominal trauma. There were associated fever, fatigue, weight loss (5 kg a month) and muscular pain. On physical examination, he had fever 38.5^{o}C. The left lower abdomen was tender and dull to percussion on auscultation, there was 2/6 grade systolic murmur at the apex of the heart radiating to the left axillary region. There was no other abnormal findings apart from moderate hypertension (BP: 150/110 mmHg). The following investigations were performed.

According to the ARA (American Rheumatology Association) criteria, we decided that this patient had PAN with renal involvement resulting in bilateral perirenal haematoma due to spontaneous rupture of intrarenal aneurysms.

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Table 1. Blood and urine analysis

Urine	:Normal	
Lactate dehydrogenase	:12.8 m kat/L	(High)
Hemoglobin	:110 g/L	(Low)
Leukocyte count	:13x10^6 /L	
Erytrocyte sedimentation rate	:70 mm/h	(High)
HBsAg	:Positive	
Immunoglobulin G	:35.61 g/L	(High)

Table 2. Other procedures

Echocardiography	:2nd Degree mitral regurgitation
Abdominal US	:Bilateral perirenal haematoma
Abdominal CT	:Bilateral perirenal haematoma (Fig. 1,2)
Renal DSA	:Intrarenal multiple aneurysms

Table 3. Blood and urine analisis

Urine	:Normal	
Lactate dehydrogenase	:11.4 n kat/L	(High)
Hemoglobin	:54 g/L	(Low)
Leukocyte count	:22x10^6 /L	(High)
Erytrocyte sedimentation rate	:50 mm/h	(High)
HBaAg	:Positive	
Immunoglobulin G	:35.61 g/L	(High)
Haematocrit	:0.18 (%18)	(Low)

Table 4. Other procedures

Renal US	:Bilateral perirenal haematoma
Renal CT	:Bilateral perirenal haematoma
Renal DSA	:Multiple, small, aneurysms, involving the interlobar and arcuate arteries

Case II: The patient was a 23 year-old male who had fever, fatigue, weight loss (5 kg a month), diffuse muscular pain for three months. There was associated abdominal pain for two weeks. He didn't describe any abdominal trauma. On examination, he had fever of 38^{O}C, BP: 150/110 mmHg. The following investigations were performed.

We supposed this patient had urinary infection. Unfortunately, after the antibacterial therapy, there were no change about fever or other sign and symptoms. Because of that, he was hospitalized at Gülhane Medical Faculty. By the time, the patient's hemoglobin value and vital signs began to be worse. So surgeons decided to take an operation, that is left nefrectomy. Macroscopically, the weight of nefrectomy specimen was 160 g and the size of it was 12x6x5 cm (Figure 3).

Regarding to case I and II, in renal US and CT, there were bilateral perirenal heamatoma, in renal DSA, there were multiple, small aneurysms involving the interlobar and arcuate arteries.

DISCUSSION

The rupture of collecting system or renal paranchyme causes subcapsular or perirenal heamatoma. At the end, because of this heamatoma, spontaneous rupture of kidney occurs at patients with PAN. It was originally reported by Bonet and Later described by Wunderlich in 1856 and is also called spontaneous apoplexy of the renal capside. Schmitt first mentioned about spontaneous perirenal heamatoma that was caused by PAN at 1908 (4-6). After that time, only 18 cases that had complication of perirenal heamatoma were reported. This serious and rare complication of PAN, generally occurs because of the intraparanchymal aneurysms rupture (7,8). Most of the cases were diagnosed after operation. It had been diagnosed by non-invasive procedures at two patients that we reported. The second patient was taken on operation after it had been diagnosed and pathologic examination of nephrectomy specimen showed that there were micro-aneurysms. So it had certainly been diagnosed as PAN. We belive that every patient presenting with spontaneous renal paranchimal hemorrhage must be investigated for PAN.

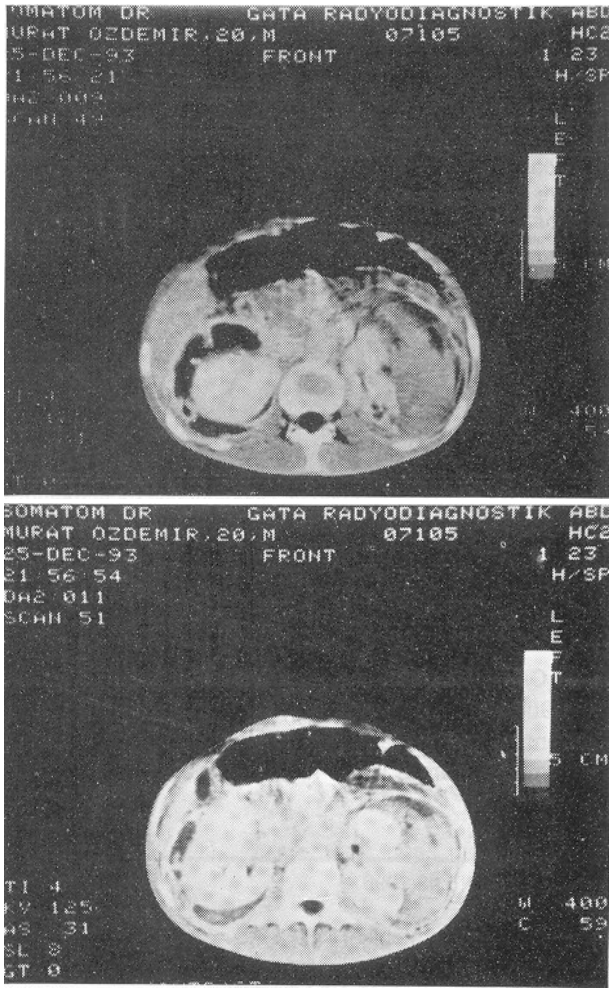

Figure 1,2. Bilateral perirenal haematoma (Abdominal CT).

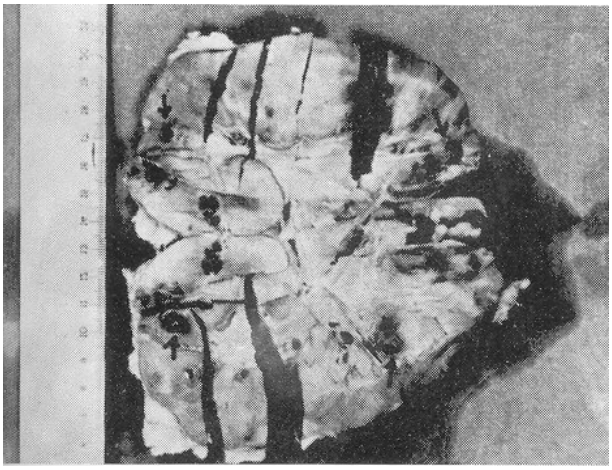

Figure 3. Nefrectomy specimen.

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