Nephrol Dial Transplant (2002) 17: 281–282

Nephrology
Dialysis
Transplantation

Case Report

AA amyloidosis complicating an inflammatory abdominal aortic aneurysm

Ali Riza Odabas[1], Ramazan Cetinkaya[1], Yilmaz Selcuk[1], Cemal Gundogdu[2], M. Derya Onuk[3], Isa Ozbey[4] and Unsal Coskun[5]

[1]Department of Nephrology, [2]Department of Pathology, [3]Department of Internal Medicine, [4]Department of Urology Ataturk University, School of Medicine and [5]Department of Radiology, Maresal Cakmak Army Hospital, Erzurum, Turkey

Keywords: computed tomography; inflammatory abdominal aortic aneurysm; nephrotic syndrome

Introduction

Ureteral compromise by periaortic fibrosis secondary to abdominal aortic aneurysm was first described by James [1] in 1935. Subsequently, numerous articles appeared in the medical literature describing similar cases. Inflammatory abdominal aortic aneurysm (IAAA) is a clinico-pathologic entity that comprises about 5–10% of abdominal aortic aneurysms [2]. Characteristic features of IAAA are excessive thickening of the aneurysmal wall and a dense, inflammatory, fibrotic reaction in the retroperitoneum that incorporates adjacent structures. Retroperitoneal fibrosis is a fibrotic process with progressive encasement and compression of the retroperitoneal region from the renal pedicles to the pelvic structures. Fibrous encasement of the ureters eventually leads to ureteral obstruction, hydronephrosis and varying degrees of renal failure [3]. Patients with an IAAA frequently present with back pain, significant weight loss and elevated erythrocyte sedimentation rate, and computed tomography (CT) reveals a thickened, often calcified aortic wall and a mass of periaortic inflammatory tissue [4–6].

In this report, we describe a patient with amyloidosis who presented a different complication due to IAAA. According to our knowledge, IAAA with amyloidosis has never been reported.

Correspondence and offprint requests to: Ali Riza Odabas, Ataturk Universitesi Postanesi, PK: 26, 25171 Erzurum, Turkey. Email: alirizao@yahoo.com

Case

A 59-year-old man was admitted to hospital because of lower back pain, right-side hydronephrosis, and nephrotic syndrome. The patient had been well until 5 years earlier when he developed lower back pain. IAAA was detected that time in another hospital. Five months before admission he noted weight loss (8 kg in 5 months), fatigue, back pain, and pre-tibial oedema. The patient was a heavy smoker. There was no history of hypertension, renal disease, tuberculosis, or skin rash. The patient's family history was unremarkable. On physical examination, the systemic blood pressure was 110/70 mmHg, the temperature was 36.4°C, no rash or lymphadenopathy was seen. There was massive oedema of the ankles, feet, and sacral region. There were no other abnormal physical signs. The laboratory tests showed: erythrocyte sedimentation rate (ESR) 110 mm/h (Westergren), C-reactive protein (CRP) 4.2 mg/dl (normal value < 0.8 mg/dl), haemoglobin 7 g/dl, platelet count 450 000/mm^3, blood urea nitrogen 60 mg/dl, creatinine 4.6 mg/dl, albumin 1.7 g/dl, cholesterol 198 ;mg/dl, triglycerides 118 mg/dl. The serum electrolytes were normal. The 24-h urinary protein excretion was 6 g. Anti-nuclear and anti-DNA antibodies were negative. An ultrasound of the kidneys showed right-side hydronephrosis and enlargement of the abdominal aorta and a sonolucent halo outside the rim of calcifications. CT showed rapid intra-luminal and slightly delayed contrast enhancement of the inflammatory periaortic soft tissue encircling IAAA at the anterior and laterally (Figure 1). Nephrostomy was inserted into the right renal pelvis. An antegrade nephrogram showed minimal passage to the urinary bladder. Renal function did not improve.

A rectal biopsy was performed which was positive for capillary amyloid deposition. The amyloid proved to be potassium permanganate-sensitive characterizing it as AA amyloid [7]. The patient was referred for aneurysmectomy to another centre but it was later learnt that he again declined the operation.

© 2002 European Renal Association–European Dialysis and Transplant Association

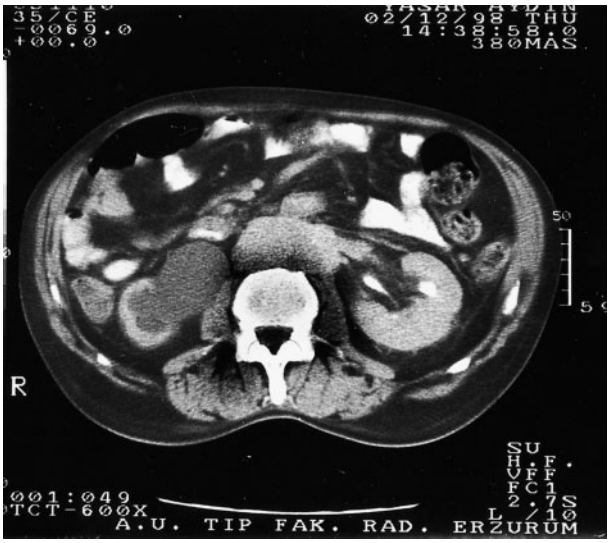


Fig. 1. In dynamic CT scanning rapid intra-luminal, slightly delayed enhancement of the inflammatory periaortic soft tissue and non-enhancement of the thick fibrous adventitia was seen.

Discussion

Although IAAA had been considered previously a distinct clinico-pathologic entity, current thinking is that it represents the extreme end of an inflammatory process leading to fibrosis, presents to a variable degree in all abdominal aortic aneurysm [8,9].

Several cases of retroperitoneal fibrosis associated with IAAA with or without ureteral involvement have been reported [8,9]. The inflammatory process involves the duodenum in $>90\%$, the inferior vena cava and left renal vein in $>50\%$, and the ureters in about 25% of cases [3]. The combination of abdominal and/or back pain, weight loss, an elavated ESR, and ureteral obstruction in the presence of an abdominal aneurysm is highly suggestive of the inflammatory type [2,4,5,9]. It is important to make the diagnosis of inflammatory aneurysm preoperatively as preparation and operative technique should be modified [4].

IAAA can be diagnosed by CT with high sensitivity; excessive thickening of the aneurysmal wall and conspicuous enhancement are the characteristic features leading to correct diagnosis [6,8]. Our patient presented with IAAA, showed by back pain, ureteral involvement, elevated ESR, and typical CT findings. We diagnosed it as an IAAA based on a symptomatic inflammatory reaction and the findings of ultrasound and CT.

The patient with IAAA presented nephrotic syndrome. He had proteinuria 6 g/day and rectal biopsy showed AA amyloidosis. AA amyloidosis is not a classic complication of IAAA. AA amyloidosis is mainly associated with long-standing infectious or non-infectious inflammation. AA amyloidosis is caused mostly by chronic rheumatic diseases, mainly adult rheumatoid arthritis and ankylosing spondilitis. Takayasu's disease and malignant retroperitoneal lymphoma may cause retroperitoneal fibrosis and AA amyloidosis. Takayasu's disease is a chronic, progressive inflammatory vasculitis of large and medium-sized vessels commonly seen in adulthood. The symptoms start before the age of 40 in Takayasu's disease [10]. In our patient, symptoms commenced at the age of 55, well beyond the accepted age presentation of Takayasu's disease. Also, contrary to Takayasu's disease, all arterial pulses were palpable. In malign retroperitoneal lymphoma, lymph nodes should be present in the abdomen. In our patient, no lymph nodes were identified in the thorax or the abdomen using CT.

We relate the IAAA and AA amyloidosis due to the severity and the long duration of the inflammation that the patient has been subjected to for 5 years without any treatment. IAAA was the most likely cause of the AA amyloidosis documented in this patient. Inherited, infectious, and other inflammatory diseases known to cause AA amyloidosis were excluded with reasonable certainty.

References

1. James TGI. Uremia due to aneurysms of the abdominal aorta. *Br J Urol* 1935; 7: 157–161
2. Sterpetti AV, Hunter WJ, Feldhaus RJ *et al.* Inflammatory aneurysms of the abdominal aorta: incidence, pathologic and etiologic considerations. *J Vasc Surg* 1989; 9: 643–650
3. Nitecki SS, Hallett JW Jr, Stanson AW *et al.* Inflammatory abdominal aortic aneurysms: a case-control study. *J Vasc Surg* 1996; 23: 860–869
4. Gans RO, Hoorntje SJ, Rauwerda JA, Luth WJ, van Hattum LA, Donker AJ. The inflammatory abdominal aortic aneurysm. Prevalence, clinical features and diagnostic evaluation. *Neth J Med* 1993; 43: 105–115
5. Crawford JL, Stowe CL, Safi HJ, Hallman CH, Crawford ES. Inflammatory aneurysms of the aorta. *J Vasc Surg* 1985; 2: 113–124
6. Liu CI, Cho SR, Brewer WH, Fields WR, Shaw CI. Inflammatory aneurysm of the abdominal aorta: diagnosis by computerized tomography and ultrasonography. *South Med J* 1987; 80: 1352–1354
7. Cotran RS, Kumar V, Robbins SL. Diseases of Immunity. *Robbins Pathologic Basis of Disease*, 4th edition. W.B. Saunders Company, London: 1989; 133–238
8. Rasmussen TE, Hallett JW Jr. Inflammatory aortic aneurysms. A clinical review with new perspectives in pathogenesis. *Ann Surg* 1997; 225: 155–164
9. Korzets Z, Witz M, Goldberg E, Rozin M, Lehmann J, Bernheim J. The patient with asymptomatic advanced renal failure obstructive uropathy caused by inflammatory abdominal aortic aneurysm. *Nephrol Dial Transplant* 1998; 13: 1835–1837
10. Ishikawa K. Patterns of symptoms and prognosis in occlusive thromboaortopathy (Takayasu's disease). *J Am Coll Cardiol* 1986; 8: 1041–1046

Received for publication: 13.3.01
Accepted in revised form: 18.8.01