CASE REPORT

Idiopathic retroperitoneal fibrosis

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KEY WORDS

azathioprine, chronic periaortitis, corticosteroids, retroperitoneal fibrosis

ABSTRACT

A 45-year-old woman was admitted to the hospital because of abdominal pain, fever, and weight loss. Laboratory tests performed on admission revealed raised inflammatory markers. Ultrasonography of the abdomen showed dilatation of the pyelocalyceal system of the left kidney, while computed tomography showed retroperitoneal concentric periaortic mass extending to common iliac arteries and entrapping the left ureter. We established the diagnosis of idiopathic retroperitoneal fibrosis (RPF). JJ catheter was placed in the left ureter, and treatment with corticosteroids and azathioprine was started. Follow-up examinations showed a gradual improvement, namely a progressive remission of the retroperitoneal mass and normalization of both erythrocyte sedimentation rate and C-reactive protein concentration. Case reports, including our paper, and small case series showed azathioprine to be particularly effective in the treatment of RPF.

INTRODUCTION Idiopathic retroperitoneal fibrosis (RPF) is a subtype of chronic periaortitis (CP). CP is a rare disorder characterized by the presence of a fibro-inflammatory mass usually surrounding the abdominal aorta and the iliac arteries. This mass may entrap the adjacent structures such as the ureters and the inferior vena cava. There are three main subtypes of CP including inflammatory abdominal aortic aneurysms (IAAAs), perianeurysmal retroperitoneal fibrosis (PRF), and idiopathic RPF. In idiopathic RPF the aorta is not dilated and the surrounding retroperitoneal mass may or may not encase the adjacent structures. Idiopathic RPF is also called nonaneurysmal RPF. In IAAAs and PRF aneurysmal dilatation of the aorta is present, and these two forms of CP are referred to as aneurysmal RPF. In IAAAs the periaortic mass does not involve the adjacent organs, while in PRF it causes obstructions. The pathogenesis of CP is not clear. Parums and Mitchinson proposed a hypothesis that CP is caused by an autoimmune response to some components of the atherosclerotic plaque, such as ceroid and oxidized low-density lipoproteins.[1,2] An alternative hypothesis is that CP is a manifestation of a systemic autoimmune disease. There is a growing body of evidence to support the latter hypothesis. First, CP may arise in the absence of severe atherosclerosis. Second, CP patients usually present constitutional symptoms, raised acute phase reactants and positive autoantibodies (the most common – antinuclear antibodies – are present in 60% of patients). Third, CP is frequently associated with autoimmune conditions affecting other organs. Fourth, CP was shown to be associated with *HLA-DRB1*03*, a genetic autoimmune marker associated also with such diseases as systemic lupus erythematosus, autoimmune thyroid disease, type 1 diabetes mellitus, and myasthenia gravis. Idiopathic RPF is a rare disorder, and available data on treatment outcomes are derived mainly from case reports and small case series. It is therefore important to report every single case of idiopathic RPF.

CASE REPORT In June 2008, a 45-year-old woman was admitted to the hospital because of abdominal pain, fever, and weight loss. Routine laboratory tests performed on admission revealed raised inflammatory markers (erythrocyte sedimentation rate [ESR] – 73 mm/h, C-reactive protein [CRP] – 42.7 mg/l). Other values, including serum creatinine and blood urea nitrogen, were within normal ranges. Abdominal ultrasonography showed

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Received: May 19, 2009.
Revision accepted: July 3, 2009.
Conflict of interest: none declared.
Pol Arch Med Wewn. 2009; 119 (10): 677-679

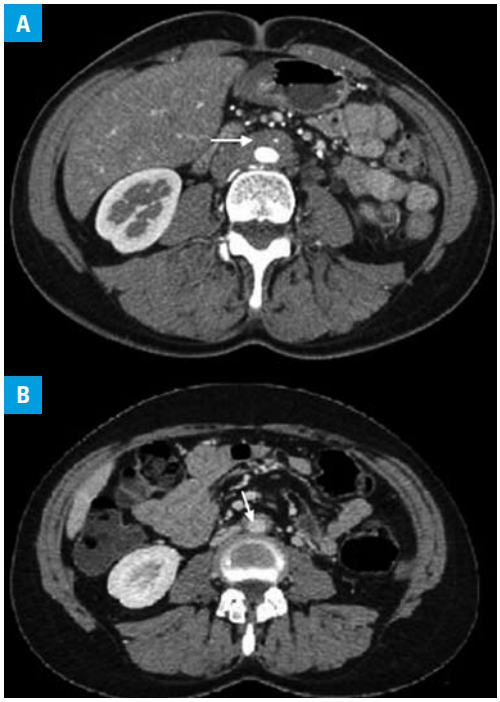

FIGURE Regression of the pathological tissue (arrows) surrounding the aorta in computed tomography
A CT performed on admission
B CT performed at 6 month

dilatation of the pyelocaliceal system of the left kidney, while abdominal computed tomography (CT) revealed retroperitoneal concentric periaortic mass starting 23 mm below the origin of the right renal artery, extending longitudinally to common iliac arteries and entrapping the left ureter (FIGURE). On the basis of the clinical, laboratory and radiological data, a diagnosis of idiopathic RPF was established. JJ catheter was placed in the left ureter and the treatment with corticosteroids (prednisone – 60 mg/day) and azathioprine (150 mg/day) was started. JJ catheter was removed after 6 months. A control ultrasonography performed after removing ureteric catheter showed a normal left ureter and pyelocaliceal system of the left kidney. Corticosteroids were gradually tapered off. At present, the patient continues a low-dose corticosteroid therapy (prednisone – 10 mg/day) combined with azathioprine (150 mg/day). The follow-up CT scans at 3 and 6 months showed a gradual remission of the retroperitoneal mass (FIGURE). Routine laboratory tests performed at the last follow-up visit (after 9 months) showed normal ESR (<10 mm/h) and CRP (<5 mg/l).

DISCUSSION The clinical picture of idiopathic RPF is characterized by abdominal, back, or lumbar pain, usually associated with constitutional symptoms such as fever and weight loss. Our patient presented all these symptoms. The most common severe complications of idiopathic RPF are hydronephrosis and renal failure (secondary to ureteric obstruction). It is estimated that about 75% of RPF patients have renal impairment at diagnosis. Management of idiopathic RPF is mainly based on corticosteroids and immunosuppressive drugs. In the case of ureteral involvement procedures relieving obstruction are performed. The placement of ureteral stents or nephrostomy is usually sufficient, although surgical ureterolysis may be necessary in advanced stenosis. Given the lack of randomized controlled trials, recommendations for the treatment of idiopathic RPF are yet to be established. At present, terapheutic decisions are made mainly on the basis of small case series and case reports. The results of Fry et al.[3] indicated that idiopathic RPF may be effectively treated with corticosteroids (without immunosuppressive drugs). However, Maillart et al. showed that corticosteroids combined with immunosuppressive drugs are more effective than corticosteroids alone (efficacy of 97% and 70%, respectively).[4] Additional rationale for the combined use of steroids and immunosuppressive agents is the possibility to reduce and taper the dose of steroids more quickly. Immunosuppressive drugs, such as azathioprine, cyclophosphamide, methotrexate, cyclosporine, and mycophenolate mofetil (MMF), were shown to be effective in RPF patients.[5-8] Recent studies suggest that MMF is becoming one of the most frequently used immunosuppressive drugs in RPF. Yet case reports, including our paper, and small case series show that azathioprine is a highly effective agent in the treatment of RPF. In Moroni et al. all patients (6 out of 6) responded to azathioprine therapy.[7] In our department steroids combined with azathioprine are considered as a first choice treatment in mild to moderate RPF cases. It should be emphasized that MMF is several times more expensive than azathioprine while the safety profiles of both drugs seem similar.[9] Hence, it seems that steroids combined with azathioprine may be used as a first-line therapy in mild to moderate RPF until randomized controlled trials prove the superiority of MMF (or other immunosuppressive drugs) over azathioprine. In severe RPF a combined therapy with corticosteroids and intravenous pulses of cyclophosphamide was shown to be particularly effective.[10]

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OPIS PRZYPADKU

Idiopatyczne włóknienie zaotrzewnowe

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azatiopryna, kortykosteroidy, przewlekłe zapalenie okołoaortalne, włóknienie zaotrzewnowe

STRESZCZENIE

45-letnia kobieta została przyjęta do szpitala z powodu bólu brzucha, gorączki oraz utraty masy ciała. W badaniach laboratoryjnych wykonanych przy przyjęciu stwierdzono podwyższone wartości wskaźników stanu zapalnego. Badanie ultrasonograficzne jamy brzusznej wykazało poszerzenie układu kielichowo-miedniczkowego lewej nerki, natomiast w tomografii komputerowej stwierdzono obecność zaotrzewnowego koncentrycznego nacieku okołoaortalnego przechodzącego na tętnice biodrowe wspólne oraz obejmującego lewy moczowód. U chorej rozpoznano idiopatyczne włóknienie zaotrzewnowe. Do lewego moczowodu założono cewnik JJ oraz zastosowano leczenie skojarzone kortykosteroidami i azatiopryną. W badaniach kontrolnych obserwowano sukcesywną poprawę wyrażającą się stopniowym zmniejszaniem się okołoaortalnej masy tkankowej oraz normalizacją odczynu Biernackiego i białka C-reaktywnego. Zarówno nasz, jak i inne opisy przypadków oraz badania na niewielkich grupach pacjentów wykazały dużą skuteczność azatiopryny w leczeniu włóknienia zaotrzewnowego.

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Praca wpłynęła: 19.05.2009.
Przyjęta do druku: 03.07.2009.
Nie zgłoszono sprzeczności interesów.
Pol Arch Med Wewn. 2009; 119 (10): 677-679