Periaortitis: The Importance of Early Diagnosis

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Chronic periaortitis is a rare disease affecting the abdominal aorta, usually below the level of the renal arteries. Other terms for periaortitis are: idiopathic retroperitoneal fibrosis, inflammatory aneurysm of the abdominal aorta, and perianeurysmal retroperitoneal fibrosis. The most serious complication of retroperitoneal fibrosis is urethral obstruction and renal failure; early diagnosis may preserve organ function and may prevent all the complications caused by fibrosis [1].

Patient Description

A 58 year old man was admitted with a continuous dull abdominal pain that started 2 weeks previously. The pain was radiating to the left inguinal zone. He had erectile dysfunction and constipation, had lost 8 kg in the last month, and felt weakness and fatigue. He was a heavy smoker (60 pack-years); he had arterial hypertension stage 1 for 10 years and was treated with disothiazide and nitrates. Six months before admission he was involved in a car accident with blunt abdominal trauma.

On admission his chest X-ray, electrocardiogram and abdominal ultrasound were normal. He underwent gastroscopy (mild duodenitis) and colonoscopy (normal). Chest computerized tomography was normal, but the abdominal CT demonstrated atherosclerotic changes of the abdominal aortic wall with calcifications and ulcerations in the abdominal lumen, and a 5 cm hypo-dense infiltrate that surrounded the abdominal aorta just below the origin of the renal arteries [Figure].

He had mild normocytic anemia (hemoglobulin 12.5 g/dl), elevated erythrocyte sedimentation rate (100 mm/hour), C-reactive protein 7.5 mg/dl, and positive rheumatoid factor. Antinuclear antibody and the perinuclear and cytoplasmic antineutrophil cytoplasmic antibodies were negative. Renal functions and liver enzymes were normal.

Prednisone 60 mg/day was started and led to a clinical and laboratory improvement. The pain disappeared, he stopped losing weight, regained his appetite, and there was an improvement in his general mood. The erectile dysfunction and the flank pain disappeared completely. ESR decreased from 100 to 30 mm/hour and the CRP decreased from 7.5 to 0.07 mg/dl. Abdominal CT performed 2 months later demonstrated that the mass surrounding the aorta had almost disappeared.

Abdominal CT shows atherosclerotic changes of the abdominal aortic wall with calcifications and ulcerations. The arrows point to the area of involvement. An infiltrate (hypo-dense mass) surrounds the aorta below the renal arteries.

ESR = erythrocyte sedimentation rate
CRP = C-reactive protein

Comments

Chronic periaortitis is a rare disease that occurs in middle-aged adult men without ethnic predisposition or familial clustering. The disease follows a chronic-relapsing course, and is characterized by the periaortic deposition of collagen that often obstructs the ureters and other abdominal organs. The initial signs and symptoms are often non-specific. A dull, poorly localized abdominal pain occurs in 80% of cases. Systemic symptoms like fatigue, weight loss and low grade fever are noted in 40–80% of patients. Renal failure and uremic syndrome occur in advanced disease. Laboratory analysis demonstrates elevated acute phase reactants (ESR and CRP) [2].

In most cases the disease is idiopathic (75%) [3]. Other causes of retroperitoneal fibrosis are malignancy (metastasis, lymphoma), drugs (bromocriptine, beta blockers, hydralazine), retroperitoneal injury (hemorrhage, radiation, rupture of abdominal organ), infections (tuberculosis, histoplasmosis) and mesenteric panniculitis [1].

The pathogenesis of the disease is still unknown. Several studies have suggested that periaortitis is caused by a local autoimmune response to antigens in the atherosclerotic plaque. However, the constitutional symptoms and the association with other connective diseases are suggestive of a systemic inflammatory disorder. Vaglio and Buzio [2] showed that 10 of 16 patients with periaortitis had a positive antinuclear antibody and some other immunologic markers like rheumatoid factor (3 patients),

antineutrophil cytoplasmic antibodies (3 patients) and antithyroid microsome and antithyroglobulin antibodies (3 patients). These data suggest that periaortitis may be an expression of a systemic autoimmune disease [1,4,5].

A few cases have demonstrated that periaortitis is associated with small and medium-size vessel vasculitis. It may be a presentation of Wegener's granulomatosis or temporal arteritis. Retroperitoneal fibrosis may be associated with ankylosing spondylitis or autoimmune thyroiditis [1,5]. In our case, the patient had been involved in a car accident, with blunt trauma to the abdominal aorta, and we suspected that the trauma might have caused an inflammatory response that was reflected by an intense local and general inflammation, which was resolved after initiation of steroid treatment.

The diagnosis is reached by CT or magnetic resonance imaging. Biopsy is necessary to exclude other disease such as malignancy or infections. In our patient, biopsy was not done because of the location of the mass that was surrounding the abdominal aorta, and the approach was too risky.

With regard to management, steroids are the principal therapy, although there is no consensus regarding the dosage and duration of therapy. Steroid-sparing agents like azathioprine, cyclosporine or methotrexate have been described. In steroid-resistant patients the combination of steroids with tamoxifen has been used successfully [1].

Since the disease has a chronic-relapsing course patients should be closely monitored after diagnosis for signs of reactivation of the disease, especially after tapering down of steroid treatment. In advanced cases, surgery – for example, ureterolysis, uretral stent insertion or nephrostomy – is effective and considered safe [1]. In our patient, an early and aggressive approach prevented irreparable deterioration and the development of retroperitoneal fibrosis. We believe that early diagnosis and treatment may prevent suffering and an irreversible outcome.

References

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