RETROPERITONEAL FIBROSIS AS A RARE CAUSE OF LOWER LIMB CLAUDICATION

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The study presented a case of a 58-year-old male patient treated for retroperitoneal fibrosis, right hydronephrosis, and right common iliac artery stenosis and saccular aneurysm of the above-mentioned vessel. The patient was qualified for endovascular treatment. Stentgraft implantation was performed with good long-term patency during more than 3 years of follow-up. Complete relief of intermittent claudication was observed. However, the endovascular exclusion of the aneurysm did not influence the course of retroperitoneal fibrosis.

Key words: Ormond’s disease, retroperitoneal fibrosis, saccular aneurysm, iliac artery, endovascular, stentgraft

Retroperitoneal fibrosis (RF) is a disease entity that was first described in 1905 by the French urologist, Joaquin Albarran. In 1948, the histopathological description of the disease was published in English literature by the American urologist, John Kelso Ormond (1). The incidence of the disease is estimated at 1/200,000 to 1/1,000,000 (2, 3), divided into the primary and secondary form of the disease. The primary form (Ormond’s disease, idiopathic retroperitoneal fibrosis – IRF) accounts for 70% of cases (2). The above-mentioned disease entity is most often observed in male patients, average age of onset ranging between 40 and 60 years (4). The etiology remains unknown, although an unknown antigen might be responsible for the initiation of the inflammatory process. One of the hypothesis is that the antigen oxidizes lipoproteins from atherosclerotique plaques responsible for the inflammatory process in the aorta, which further spreads in the retroperitoneal space.

Considering the fact that most often, abdominal aneurysms are observed below the renal vessels and that the retroperitoneal fibrosis process begins to develop near the promontory, one may come to the conclusion that there is a correlation between the inflammatory process in the vessels and retroperitoneal space. The coexistence of other autoimmune diseases might be evidence of the common mechanism for the initiation of the process of fibrosis. Pathogenesis considers genetic factors, since 44% of patients are diagnosed with antigen HLA-B27 (5).

RF is characterized by limited or diffuse retroperitoneal space fibrosis, manifested by early clinical symptoms, such as blunt abdominal and lumber pain, as well as lower limb edema. Secondary RF might be observed in patients with primary tumors of the retroperitoneal space and in case of metastases, as well as a consequence of abdominal cavity radiotherapy, and abdominal aorta and urinary duct surgery. Some Authors mentioned drugs (metrysergid, beta-blockers, ergotamine) (4) as the possible cause of secondary RF. However, it seems justified that the hypothesis presented in literature data by Wołyniec (1) shows that it is only a risk factor of RF development. Due to unspecific clinical symptoms RF diagnosis is based on imaging examinations, such as CT.
and MRI. The characteristic feature of RF is the creation of a so-called “blanket” covering the aorta and inferior caval vein by a fibrous mass (6).

A similar picture might be observed in case of retroperitoneal space sarcomas, amyloidosis, and metastases, which should be considered during differential diagnosis. In consequence of urinary duct fibrosis one may observe hydronephrosis. Less frequently did we observe the infiltration of the iliac veins and inferior vena cava leading towards thrombosis, as well as the inflammatory process of retroperitoneal space arteries. Their stenosis, due to wall structure differences is rarely observed.

Conservative treatment of retroperitoneal fibrosis consists in the administration of immunosuppressive drugs (cyclophosphamid, azatioprin, glicocorticosteroids) and anti-estrogens (tamoxifen). Surgical treatment consists in the surgical decompression of fibrous urinary ducts or vessels. Their exist minimally invasive methods, such as implantation of double J catheters to the urinary ducts or vascular stenting.

The aim of the study was to present the rare cause of claudication, secondary to Ormond’s disease, arising from the inflammatory process of the right iliac artery.

**CASE REPORT**

A 58-year old patient with a history of arterial hypertension and smoking, one year since diagnosis of retroperitoneal fibrosis remains under outpatient nephrological control after right-sided nephrostomy, due to hydronephrosis as the first manifestation of IRF. After one year we observed intermittent claudication of the right lower limb and the patient was directed to vascular outpatient surgery. Doppler ultrasound showed stenosis of the right common iliac artery and a saccular aneurysm of the vessel. The patient was qualified for endovascular treatment and referred to The Department of Cardiosurgery and Vascular Surgery, GUMed.

Under local anesthesia the right femoral artery was punctured introducing a 6F catheter. Arteriography confirmed critical stenosis of the common iliac artery, 20 mm in length, and a saccular aneurysm (10x15 mm) located near the aortic bifurcation (fig. 1). A balloon catheter was introduced, 6 and 8 mm in diameter. However, after each angioplasty recurrent stenosis was observed (elastic recoil – associated with the inflammatory character of the lesion). The previous catheters were replaced by a 11F size, followed by a Jostent stentgraft, 39 mm in length (Abbott Vascular, Santa Clara, CA, USA). It was opened at the level of the iliac artery stenosis, simultaneously covering the saccular aneurysm of the common iliac artery. Control arteriography showed a positive effect and stenosis disclosure (fig. 2). After catheter removal manual pressure was introduced at the site of the puncture. The patient was discharged home the next day.

Two months after the procedure the physical examination revealed the presence of a pulse on the right lower limb, and angio-CT showed

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*Fig. 1. Angiography showing critical stenosis of the right common iliac artery with inflammatory infiltration. Proximally visible saccular aneurysm*

*Fig. 2. Stentgraft implantation to the right common iliac artery. Visible aneurysm exclusion and dilatation of the stenotic vessel*
Retroperitoneal fibrosis as a rare cause of lower limb claudication

Fig. 3. Angio-CT of the pelvis-frontal plane. Arrow 1 points to the distended right ureter, while arrow 2 to the implanted right common iliac artery stentgraft.

Fig. 4. Angio-CT of the pelvis-transverse plane. Arrow 1 points to the right common iliac artery with stentgraft implantation, while arrow 2 shows the perivascular inflammatory infiltration.

full patency of the iliac artery and saccular aneurysm exclusion. Shortly after the vascular procedure a double J catheter was introduced into the right ureter, in order to eliminate the nephrostomy. Several months thereafter, surgical release of the right ureter was necessary. Another angio-CT examination performed in September, 2012 showed a patent stentgraft and at the same time an infiltration of the right iliac artery and saccular distention of the distal segment of the aorta, as well as distention of the right ureter, up to 1 cm (fig. 3). Additionally, a 28 mm in diameter concrement was observed in the right renal pelvis. In December 2012, percutaneous nephrolithotripsy was performed. During the 45-month observation period stentgraft stenosis recurrence was not observed, nor novel stenotic lesions, despite the continuous process of retroperitoneal fibrosis (fig. 4). The patient to this day remains without symptoms of intermittent claudication.

DISCUSSION

Ormond’s disease may be manifested by a number of unspecific symptoms involving the urinary, vascular and gastrointestinal tracts. Rare symptoms include arterial stenoses manifested by intermittent claudication. Literature data presented cases of percutaneous intervention, considering patients with iliac vein (3) and inferior vena cava lesions (7). We only found one case report presenting the surgical intervention on the aorta and iliac arteries, due to inflammation, being associated with cocaine intoxication (secondary fibrosis) (8).

Considering the presented study patient isolated intermittent claudication symptoms of the right lower limb appeared one year after Ormond’s disease diagnosis. Therapeutic management in our case includes two options: open surgery with the excision of the stenotic vessel and surrounding inflammatory tissue and implantation of a vascular prosthesis, or the endovascular procedure with stentgraft implantation. Open surgery is burdened with the risk of damage to surrounding structures (duodenum, ureter or common iliac vein) and preparation of infiltrated tissues. Endovascular procedures are an interesting alternative in case of patients burdened with severe concomitant diseases for whom many hours of vascular surgery might prove to be too burdensome. Discussions are under way on how to reduce the inflammatory process in the retroperitoneal space by excision of the stenotic vessel. However, there is no conclusive evidence on the effectiveness of such management. Stone et al. mentioned the reduction in the thickness of the inflammatory process surrounding the aorta by approximately 50% after stentgraft implantation into the abdominal aneurysm, considering a group of 10 patients. However, the above-mentioned process is slower, as compared to that observed after open aneurysmal surgery (9).

Due to the inflammatory nature of the disease we decided to perform the endovascular procedure. The distant result concerning vascular patency and complete relief of intermittent claudication was good. Despite the parallel administration of glucocorticosteroids and anti-estrogens (oral Prednison 15 mg, once
daily, oral Tamoxifen 10 mg, once daily) the inflammatory process in the retroperitoneal space remained unchanged. Prolonged urine stagnation in the right kidney, due to ureter stenosis led to the creation of a concrement, which required surgical removal (fig. 5). The presented case as one of few describes the inflammatory process of the arterial vessel. Few studies described Ormond’s disease considering the inflammation of venous vessels.

CONCLUSION

Intermittent claudication of the lower limbs is one of the basic symptoms encountered in vascular surgery. However, when isolated, rapidly progressing intermittent claudication is rare. During differential diagnosis, especially in the presence of other extravascular symptoms, one should consider retroperitoneal fibrosis. The endovascular procedure provides an opportunity for effective treatment and relief of symptoms associated with lower limb ischemia. We observed no significant regression of the inflammatory infiltration involving the right iliac artery, despite the exclusion of the aneurysm and typical anti-inflammatory therapy.

REFERENCES


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