Idiopathic Retroperitoneal Fibrosis – Treatment choices

A. J. Linhares Furtado

Prof. Catedrático de Cirurgia, Jubilado, da Fac. de Med. da Universidade de Coimbra

Resumo

Apresentam-se três casos de fibrose retroperitoneal idiopática (F.R.P.I.),em dois dos quais, o tratamento exclusivo foi a intubação uretérica definitiva, ao longo de 32 e de 20 anos, com tubos de "silastic" por via aberta, segundo técnica ilustrada neste trabalho. Não se registaram deposições litiásicas e a tolerância foi excelente. No terceiro doente utilizou-se a mesma técnica, alterada após 6 anos para duplos J, sendo substituídos periodicamente (tempo total de seguimento 18 anos). Na literatura ao nosso alcance não se encontram observações seguidas por tão longo tempo. Estes casos evoluíram de forma excelente, parecendo muito provável que, na ausência de outros factores capazes de comprometerem a função renal, a manutenção de uma boa drenagem renal assegure a preservação funcional dos rins.

O A. defende que a terapêutica inicial da F.R.P.I., exceptuando provavelmente as crianças, deve consistir na colocação bilateral de duplos J de material biocompatível (o "silastic" mostrou-se excelente). Na grande maioria dos casos, a restauração de boa drenagem pielo-calicial acompanha-se do desaparecimento das dores. O recurso a terapêutica médica – corticosteróides, tamoxifeno, imunossupressores – deve reservar-se para os casos nos quais persistam dores ou outras complicações – essencialmente vasculares. Nestas últimas situações as técnicas endovasculares oferecem uma solução habitualmente definitiva, isoladamente ou associadas a terapêuticas médicas.

Em conclusão, a cirurgia aberta para tratamento da F.R.P.I., quer na obstrução uretérica (de longe a mais frequente), quer nas complicações vasculares, deixou, praticamente, de se justificar.

Summary

Three cases of I.R.P.F. are presented here, two of which were treated exclusively through definitive ureteric stenting, during 32 and 20 years, using an open technique also described in this paper. Tolerance and preservation of renal function were excellent, and no lithiasis deposits have appeared. A third patient was initially treated in the

same way, but the "silastic" tubes were replaced 6 years later by double J's periodically changed (total follow-up time was 18 years). No other cases of such long term observations were reported in all the literature that we had access to.

These cases have evolved in a very positive way and they allow us to conclude that the upholding of kidney drainage ensures functional preservation of the kidneys, provided other factors, capable of affecting the renal function, are absent (intrinsic renal disease, hypertension, cardiac failure, etc.).

In the Author's view, the initial treatment of choice for symptomatic I.R.P.F. in the adult patient should be the endoscopic insertion of bilateral stents made of proven biocompatible material ("silastic" showed to be excellent). In most cases, restoring good pielocalicial drainage makes the pain disappear. Medical treatments - corticosteroids, tamoxifen or immunosupressor- should be offered to patients in which, despite good kidney drainage, pain or other complications (essentially vascular ones) persist. If there is no response to medical treatment (which may include anticoagulation), endovascular prosthesis should be inserted.

In conclusion, open surgery for treating I.R.P.F. is no longer justified, both in ureteric obstruction cases (the most common ones) and in vascular complications.

Introdution

Idiopathic Retroperitoneal Fibrosis (I.R.P.F.) or Ormond's disease is a very rare inflammatory disease, which appears mainly in men. Reasonable evidence supports [1,2] an immune etiology, but the designation –idiopathic– stands on clinical practice. Its extension and clinical manifestations are varied. Most cases are diagnosed because of abdominal, flank or back pain or else acute or progressive renal failure. Symptoms related to vascular pathology (edema, hypertension, hematuria) and other forms of presentation are very rare [3,4,5,6,7]. Although the disease may be associated to one or more of a variety of other pathological conditions or plausible contributing factors [8,9], the impossibility of establishing a relation between cause and effect is crucial in classifying fibrosis as idiopathic. The medial and almost symmetrical displacement of both lumbar ureters is a classical sign. A T.A.C. scan or M.N.R. imaging clearly shows the extension of the fibrosis, involving the ureters and the retroperitoneal vessels [4]. The biopsy definitively establishes the diagnosis [10]. A wide variety of non surgical and surgical treatments have been proposed. However, most reports regard short or medium term follow-up. Three cases of long term use of ureteric stenting, with excellent results, are herein reported, and the main therapeutic choices are discussed.

Brief cases reports:

Case I

M.S.S. 45 year old man with anuria evolving for two days and unresponsive to diuretics; generalized edema, confusion, serum creatinine 9 mg/dL, moderate anemia, slightly elevated blood pressure, non painful abdominal distention, normal rectal examination; radionuclide renal scan indicating bilateral obstruction; no apparent lithiasis on abdominal Rx. Bilateral ureteric catheterization gave place to important urinary drainage and rapid clinical and biochemical improvements; repeated Rx –typical medium displacement of both ureters. Infraumbilical laparotomy confirmed extensive retroperitoneal fibrosis. Definitive insertion of Fr 7 Ch "silastic" tubes in both ureters was carried out according to the technique shown in fig. I. This patient was seen recently (32 years later), suffering from a T4 non-obstructive prostate cancer (PSA 230 ng/ml), with a very good general condition, serum creatinine of 1 mg/dl. Plain abdominal Rx: no deposits on the "silastic" tubes.

Case II

J.M., mail, 56 years old, obese, severely hypertensive for a long time; fatigue; lower limbs edema, oliguria rapidly progressing to complete anuria; left ventricle hypertrophy; serum creatinine - 5 mg/dl. Bilateral ureteric catheterization immediately followed

by intense polyuria and clinical improvement, except for persistent hypertension, although it became better responsive to adequate medication. Creatinine stabilized to 1,7 mg/dl in four days. Laparotomy confirmed very extensive I.R.P.F. Bilateral ureteric implantation of silastic tubes was carried out as in case I. Followed-up for 20 years, during which renal failure progressed very slowly, without obstruction. The patient complied poorly to antihypertensive medication, nutritional recommendations and physical restraints. Progressive congestive cardiac failure leading to MOF and death at the age of 76.

Case III

H.A.L., healthy man, 34 years old; first complaint - steadily increasing lumbar pain. C.A.T. scan suggested extensive I.R.P.F. Right kidney function severely impaired. Diagnosis confirmed through laparotomy and biopsy. "Silastic" tubes were placed in both ureters as in the previous cases. No improvement in right kidney function, in spite of no obstruction; good left kidney function. Relaparotomy one year later: extensive and thick fibrosis constricting the aorta, the iliac veins and the vena cava which were extremely difficult to isolate (aorta torn twice); right renal artery almost totally collapsed due to fibrosis, severely damaged during attempts to dissect it. Because of the extension of the fibrosis to the pelvis, auto-transplantation was deemed unfeasible. Six years later, "silastic" tubes were removed by another Urologist, because of small deposit on the knot which secured them; "silastic" tubes showed no incrustations at all. Double J stents were inserted and changed every 12 months. With 50 years old, right lower limb edema. In 2004 serum creatinine was 1,4 mg/dL. GFR (left kidney) was 45 ml/min/1,73 m2. The edema subsided on intermittent corticosteroid therapy alone. Apart from initial periods of hospital admission, the patient has been fully active during the 18 years following the diagnosis of the disease.

Discussion

I.R.P.F. is a rare pathological condition, whose diagnosis became easier thanks to the contributions of C.A.T. scans or M.N.R. These imaging procedures provide a very high diagnostic probability, although biopsy remains the ultimate evidence for a definitive diagnosis.

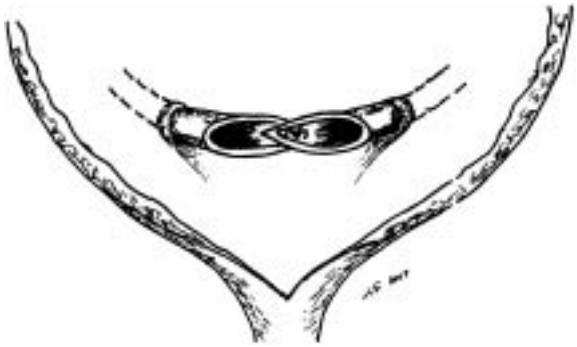

Fig. 1 - Stenting of both ureters using straight 7/8F.Ch silastic tubes cut to the needed length; 3 to 5 holes were made on the upper ends; the lower tips were cut obliquely and stuck to the trigonal muscle with a deep stitch of non-absorbable monofilament 3/0.

There are important questions concerning the best treatment choices, when dealing with the individual cases. A number of medical treatments have proved to be effective not only in reducing pain, but also in diminishing the amount of fibrosis. Corticosteroids, tamoxifen and immunosupressors are the most effective medical therapies [11,12,13,14]. For most published cases, however, the follow-up periods were relatively short and the responses were not uniformly good. The undesirable effects of these therapies are not negligible, and the economic and discomfort burdens are significant. Frequent monitoring of the disease course is strongly advised, in order to detect early impending ureteric obstruction [10]. The vast majority, if not all of the consequences of I.R.P.F., stem from entrapment and compression of the ureters and vessels. In rare cases the fibrosis may reach the mediastinum and cause obstructions which could demand special treatments [7]. However, the most common and dreadful complication is renal damage, a direct consequence of ureteric obstruction.

The most frequently performed surgical treatment has been ureterolysis, whether followed or not by intraperitonealization or simple lateralization of the ureters, sometimes accompanied by wrapping the ureters with vascularized omentum or local fat; open or laparoscopic approaches may also be used [15, 16,17]. To our knowledge, no long-term reports on the effectiveness of these operations are available. The follow-up of the three cases herein reported spans 18 to 32 years. They decidedly suggest that the renal function will be preserved as long as the ureteric obstruction is avoided and the renal vessels are

spared (which is the usual common situation). The excellent biocompatibility and flexibility of "silastic" allows long-term stenting with little or no risk of solid deposits. The vesico-renal reflux inherent to stenting is not deleterious to the adult kidneys and no discomfort or urinary infections were reported by those patients (nor by others submitted to the same operation for other causes). We suggest annual monitoring consisting of abdominal Rx, radionuclide renography and creatinine clearance. At the time, when those patients were operated there were no double J's silastic stents. Thus, the open approach that has been used is no longer required, except for differential diagnosis in doubtful cases. In our experience, nephrostomy was never needed. Indeed, the ureteric catheterization was easy in all cases (a fact well known for long). For all procedures, general or more often subarachnoid or epidural anesthesia were used.

Although there are reports mentioning spontaneous resolution [18,19], we do not support therapeutic abstention, given that some degree of ureteral obstruction is always present at the time of diagnosis, in the overwhelming majority of cases.

Vascular complications are dealt with by endovascular angioplasty or endovascular prosthesis [20,21, 22]. Anticoagulant treatment may be used alone or added to other medical therapies or to stenting.

Contrary to others, [23] our view is that ureterolysis is no longer the treatment of choice (if needed at all). Ureteric indwelling stenting must be the first approach for treating I.R.P.F. In some rare cases, mainly if pain persists, stenting should be complemented intermittently with steroids, tamoxifen or immunosuppressive drugs, according to the therapeutic responses.

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