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Idiopathic Retroperitoneal Fibrosis

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Rezumat

Fibroza retroperitoneală idiopatică

Fibroza retroperitoneală idiopatică (F.R.I) a preocupat lumea chirurgicală începând cu anul 1948 când a fost publicat, de către J. K. Ormond, primul caz de stenoză ureterală consecutivă unui proces sclero-inflamator retroperitoneal de cauză necunoscută. Până în 1960 s-au raportat în literatură 82 de cazuri. Autorii prezintă 2 noi cazuri personale de FRI, localizate exclusiv în compartimentul pelvin (subperitoneal) al spațiului retroperitoneal. Este trecută in revistă literatura dedicată acestui subiect, se evocă teoriile etiologice actuale si se discută oportunitățile de tratament.

Cuvinte cheie: fibroza pelvină retroperitoneală, stenoză rectală

Abstract

The world of surgery has been concerned with idiopathic retroperitoneal fibrosis (RPF) ever since 1948, when J. K. Ormond published the first case of ureteral stenosis following a retroperitoneal scleroinflammatory process due to unknown reasons. Until 1960, 82 such cases were reported in the literature. The authors present two new cases of RPF, located exclusively in the pelvic (subperitoneal) compartment of the retroperitoneal space. The literature on this subject is reviewed, current etiological theories are mentioned and treatment opportunities are discussed.

Key words: retroperitoneal pelvic fibrosis, rectal stenosis

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Introduction

A rare condition, it became known due to J. K. Ormond, who, in 1948, described uterohydronephrosis following a ureteral obstruction in a retroperitoneal inflammatory mass (1). In 1960, the same author recorded the first 82 cases published until then. Afterwords, we can rarely find one or two cases published in the literature. It is clearly a rare pathology, with approximately 1.4 cases per 100,000 inhabitants. Autopsy histopathological descriptions show the existence of a fibrous tumour which occupies the retroperitoneal space between the sacral promontory and the renal vessels, encompassing the great vessels (the aorta and the inferior vena cava) and the iliolumbar ureters. This scleroinflammatory "muff" is relatively well defined. The fibrous tissue and the various degrees of lymphoplasmacytic inflammatory infiltrations prevail in the microscopic aspects. As the disease evolves, the inflammatory infiltrations are reduced, being replaced by fibrocollagenous structures. Real retroperitoneal sclerotic masses are created, which are capable of generating ureteral and rectosigmoid stenoses.

A particular case is that of subperitoneal sclerosis of the conjunctive tissue built around the pelvic organs. The first case thus located was published in 1998 (2). We will present two cases of pelvic subperitoneal fibrosis encountered in our clinic.

Before describing our cases, we will provide an overview of the nosologic framework of this pathology. Anatomopathologists describe the presence of a fibrous muff surrounding the great vessels – the aorta and the inferior vena cava – without, however, invading them. The muff consists of an unspecific, polymorphous reaction. In the later stages, the inflammatory tissue is replaced by the fibrosclerotic component. Initially, lymphoplasmacytic infiltrations prevail. The inflammatory cellular elements are scattered in a mass of fibrous tissue, located around the great vessels. The process covers the area between the renal pedicle and the sacral promontory. Exceptionally, the fibrosis goes down into the pelvis. At times, this fibrotic mass can contain areas of fibrinoid necrosis, lymphocytic infiltrations (B and T lymphocytes), epitheloid or gigantocellular structures, which suggest a sarcoidosis (A. Mzabi). Most of the time, a clear immunologic participation can be identified. Thus, antinuclear and anticytoplasmatic antibodies can be highlighted, among others. Finally, the immunoelectrophoresis can point out an increased activity of Th2 lymphocytes. At the same time, there can be a positive value of the CD4 marker, which is a glycoprotein specific to the surfaces of immune cells: helper T lymphocytes, macrophages, monocytes (4-6).

The clinical manifestations of the RPF are unspecific and appear relatively late. The diagnosis is established, on average, approximately 8 months after the onset of the disease. In the disease pattern, lumbar pains, phenomena of renal failure and the inflammatory syndrome predominate. Physical fatigue, weight loss and digestive discomfort can also be noted. Often, a marked increase of the VSH and of the values of the C-reactive protein is noted. In this clinical context, the diagnosis is established through imaging exploration: ultrasound is indicative, the CT and the MRI establish the diagnosis. Percutaneous image-guided biopsy is generally little used. Most often, surgical biopsy is used. The scanner shows the pyeloureteral dilatation, the hydronephrosis, the medial deviation of ureters, the preaortocaval fibrosclerotic mass. In what follows, we will present the cases we have encountered.

Case Reports

M.V. female, 37 years old. The patient goes to the doctor for diffuse pelvic pain. She has no notable personal pathological history. During the genital examination, the left appendage is not palpated, the uterine cervix is normal, but a hard, fixed tumoral mass can be felt, occupying the pouch of Douglas. The digital rectal examination reveals the existence of a left parameter fully occupied by a hard, woody mass. A pelvic MRI scan is carried out – diffuse tumoral process in the left parameter, with

diameters of 60/44/34 mm, with invasion of the pelvic peritoneum, in close contact with the left ovary, the uterus and the sigmoid, which has a slightly thickened, diffuse wall. Suspicious pelvic adenopathies. Colonoscopy – the video endoscope is introduced up to 18 cm from the anal opening. Normal-looking mucosa. An area of stenosis due to extrinsic compression is blocking the way. Under suspicion of a left adnexal neoplasia diagnosis, surgery is performed in August 2019. During surgery, an average amount of serosanguinous fluid is found (bacteriological examination – no germs, normal cytology). The intestinal ansae and the colic frame are much distended up to the level of the rectosigmoid junction, where a hard tumoral mass is detected, encompassing the left iliac vascular pedicle and the rectosigmoid junction. Biopsic material is sampled from the rectosacral space and from the epiploon. A colostomy in continuity is performed on the transverse colon, the sigmoid being unsuitable for this procedure. Biopsy – macroscopic – 1.5/1 cm violaceous-ash coloured tissular fragments. A 25/6 cm epiploon with small indurative areas on its surface. Microscopy – chronic inflammation with perivascular predominance, rare granulocytes, dense fibrosis and focal hematic infiltration, rare groups of mesothelial cells, on the surface of the fragment (*Figs. 1, 2*).

Favourable postoperative evolution. The MRI is repeated 2 months after surgery. It shows increased diameters and a diffuse altered signal of the cervicovaginal region. Bilateral parametrial and medial and left paramedial intervesicovaginal tissular infiltration. The MRI is repeated 8 months after surgery. A shrunk cervicovaginal oedema is observed. The thickening and the irregular outline of the superior vaginal wall persist; irregular fibrous infiltration extended between the cervicovaginal region and the region of the left appendage. The decrease of pelvic inflammatory infiltration, with some persisting perivaginal signal abnormalities also in the region of the left appendage. Signal abnormalities of the anterior rectal wall persist, being most likely inflammatory. The onset of a complex cystic formation on the left ovary, with a 47 mm diameter, well delimited, having a thick wall, septa and dependent sediment, without any solid component or restriction on the diffusion sequences – noted as functional cyst. The right ovary cannot be separated from the adjacent intestinal ansae. Common and external iliac nodal images stable from the perspective of size and number, without any adenopathy (*Fig. 3*).

The second case is that of a 66-year-old man, C.I. He was diagnosed in March 2013 with a stenotic, haemorrhagic middle rectal tumour. A year before, the patient had been admitted in a different surgical unit for constipation alternating with diarrhoea. Faecal

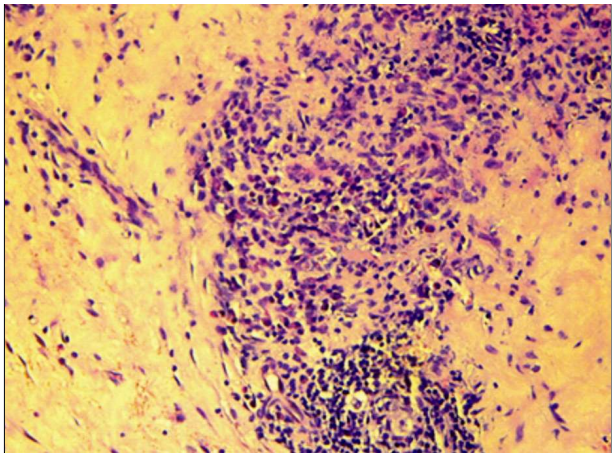

Figure 1. H&E stain, 20x lens, fibrosis and lymphoplasmacytic infiltrations mostly perivascular, with dispersed eosinophils

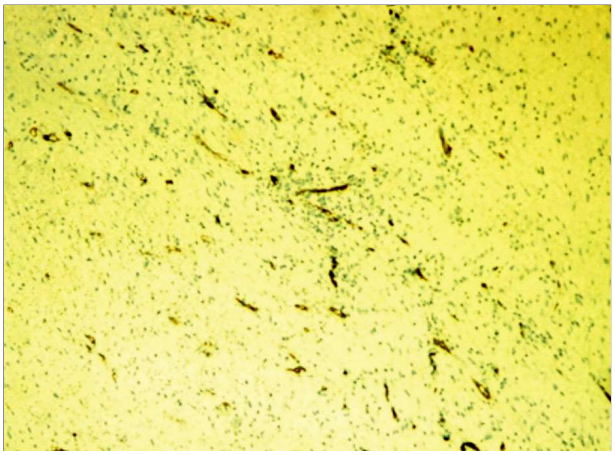

Figure 2. CD34 positive in vascular endothelial cells, negative myofibroblastic and inflammatory cells

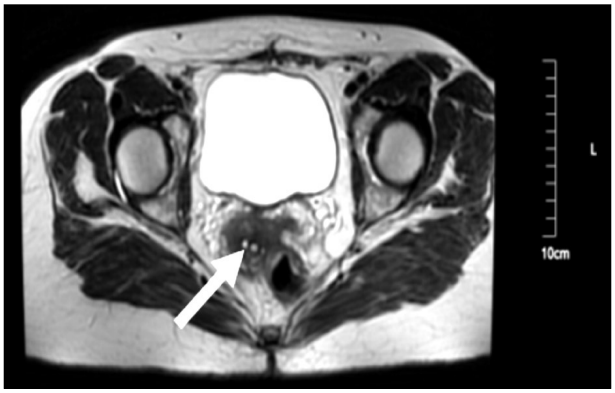

Figure 3. Follow-up MRI 8 months after surgery. The arrow indicates the retroperitoneal fibrous tumour

bacteriology tests were negative. At that time, a colonoscopy was performed – 10 cm from the anal opening there was a ulcerovegetative tumour occupying almost the entire lumen, which prevented the progression of the endoscope. The biopsy performed revealed an adenomatous hyperplasia, villous areas and unspecific inflammatory infiltration. This was followed by a CT, which identified an approximately 8 cm long spastic area at the level of the sigmoid. The lesion was 12 cm from the anal opening.

In those circumstances, a laparoscopic surgery was performed, proceeding to rectosigmoid resection and ultralow anastomosis, carried out mechanically. The postoperative evolution was eventless, with the exception of a diarrhoeic episode, interpreted as acute enterocolitis (negative clostridium difficile). Biopsy of the removed piece – rectal adenomatous hyperplasia, without neoplastic lesions.

One year after surgery, he addressed our clinic for occlusive phenomena, but also against the backdrop of faecal incontinence. The tumour markers CA 19-9, CEA were normal. During rectal touch – an area of tight stenosis of the coloanal anastomosis, with no tonus of the anal sphincter and faecal incontinence.

A colonoscopy was performed (03.2014) – the colonoscope was introduced up to 3-4 cm from the external anal opening, then it was blocked by a 4-5 mm diameter stenosis area. Several biopsies were sampled.

Histology – small fragments of mucosa with glands dissociated by the presence of fibrotic strips and a massive polymorphous lymphogranulocytic infiltration in the chorion. Immunohisto-chemistry – positive CEA in the superficial glandular epithelium, positive Ki67 in the basal glandular layers and in the normal lymphoid structures of the submucosa. There were no neoplastic lesions. The first blades were seen in another medical unit.

A CT was performed in March 2014 – a 5/4.3/7 cm iodophilic, spiculated tumoral mass, presacral and attached to the sacrum, which infiltrated the rectocolic anastomosis, the levator ani, extended to the right seminal vesicle and to the posterior side of the prostate (*Fig. 4*). The CT was repeated 5 months later (August 2014), revealing an increased process of pelvic fibrous infiltration.

As a consequence, a colostomy in continuity was performed on the transverse colon. In 2016, 2.5 years after the rectosigmoid resection, an MRI was performed – the bladder has flexible walls and no uretero-hydronephrosis. Small-sized rectal stump, with induration of the perirectal fat. Small-sized, visible low colorectal anastomosis.

Discussions

It is useless to underscore the rarity of this condition (1.3 cases/100,000). This category

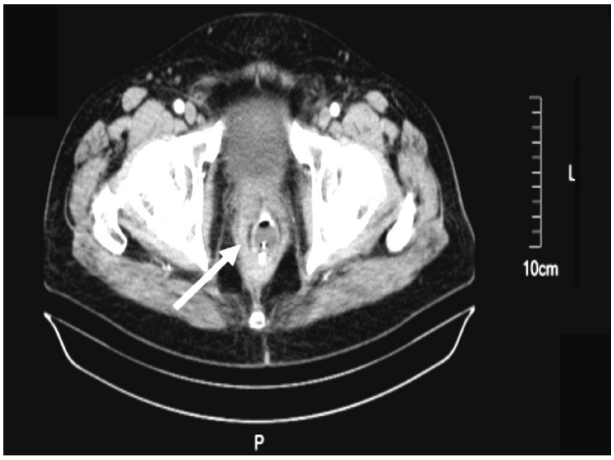

Figure 4. March 2014.The arrow indicates the RPF tumoral mass which envelops the clips of the laparoscopic low colorectal anastomosis

does not include the cases of RPF following radiotherapy, infections, medicinal treatments or other conditions. A large majority of authors insist on supposed "aortites" that would trigger a perivascular inflammation, "periaortitis", which would start the scleroinflammatory process (7-9). Others considered that this condition was associated with type 1 diabetes, with lupus erythematosus and even with myasthenia gravis. Finally, there are a lot of specialists who support the autoimmune theory: the presence of various antibodies, including antifibroblast antibodies, occasional identification of IgG4 (4,5).

In our cases, there were several issues of differential diagnostic. In situations similar to our cases, namely rectal stenosis due to a compressive pelvic block, the following differential diagnoses are necessary: GIST tumour, sarcomatous tumours (leiomyosarcoma), genital tumour, rectal neoplasias. Naturally, besides the aforementioned nosologic entities, the following can also be added: Chron's disease, lymphogranuloma venereum proctitis (Durand-Nicolas-Favre disease). However, pathologies such as Chron's disease or Durand-Nicolas-Favre disease have certain peculiar characteristics which we have not encountered in our patients (ulcerative, mucopurulent rectitis) and the onset of rectal stenosis occurs only towards the end of the pathological process. Only rectal and genital neoplasias are left to be discussed (the first observation), which, besides carcinomas, also include stromal tumours and leiomyosarcoma-like sarcomas.

During the first examination, the colonoscopy provided normal relations with respect to rectal intrinsic stenoses. The initial diagnosis during the second examination was that of rectal neoplastic stenosis, for which a rectosigmoid resection and an ultralow anastomosis were carried out. The neoplastic origin of the stenosis was not confirmed.

In order to correctly categorise this pathology in our patients, we have carried out several tests for markers which help make a distinction between GIST tumour, leiomyosarcoma and retroperitoneal fibrosis. The following markers were analysed: S100, CD34, EMA, Actin, CD117, cytokeratin AE1/AE3. All the analysed markers were negative in the fibroblastic cells. The histopathological and immunohistochemical aspects allowed us to rule out a stromal tumor or a leyomyosarcoma and therefore the diagnosis of retroperitoneal fibrosis was confirmed.

Conclusions

- Idiopathic retroperitoneal fibrosis (RPF) represents a challenge in establishing the diagnosis and the therapy to be followed.
- The surgical treatment is "palliative", solving the complications, while the fibrous lesions continue to evolve.
- The role histopatholgic examination and immunohistochemistry is to rule out a neoplastic infiltration of the retroperitoneal tissue, and to establish the diagnosis.
- RPF is a benign disease, with a "malignant" evolution.
- Subperitoneal fibroses stand out among RPF due to the fact that the ureterorenal area is not influenced.

Conflict of Interest

The authors declare no conflicts of interests.

Ethics Approval

The case study was done by following the ethical norms of scientific research and the principles anonymity a confidentiality.

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