Association of retroperitoneal fibrosis, proctitis and rectal stenosis

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Summary

Retroperitoneal fibrosis (RF) in most cases is 'idiopathic'. In the case reported, RF could be the result of a proctitis that followed a haemorrhoidectomy.

Introduction

Retroperitoneal fibrosis (RF) is a fibrotic process in the retroperitoneum, that often causes ureteral obstruction. This condition, first described by Albarran in 1905, became an established clinical entity in 1948 owing to the work of Ormond. Studying the origins of this illness, Koep and Zuidema (1977) reviewed 481 cases of RF, of which 68% were considered idiopathic. This study presents a case of RF following a proctitis which ended in rectal stenosis.

Case report

A 67-year-old man was admitted complaining of burning nocturnal lumbar pain, localized at the 2 last lumbar vertebrae and which spread out anteriorly; intermittent fever (37.5-38°C); loss of weight (7 kg in 2 months); anorexia and periumbilical pain. An ESR performed just before admission was 100 mm/hr. Before admission, the patient had occasionally used analgesics (pyrazolone), corticosteroids and anticholinergic drugs, without improvement of his symptoms.

Two years previously he had undergone surgery for haemorrhoidectomy, with subsequent proctitis and rectal stenosis; this last was treated with periodic dilatations. No other important element was found in his physiological and family history.

The clinical examination showed no abnormality.

The laboratory tests showed a mild microcytic anaemia (Hb was 11 g/dl, MCV 80 fl), a high ESR (116 mm/hr), an elevated blood urea (5.98 mmol/l,

haematuria, an increased concentration of α_2 -globulins (18%) and a reduction of albumin (38%) on the protein electrophoresis, a reduction of creatinine clearance (35 ml/min) and a mild iron deficiency. Auto-antibody screen (to the nuclei, DNA and mitochondria) and immunoglobulins levels were normal; cryoglobulins, L.E. cells and Bence-Jones protein were absent.

Radiology of the lumbar spine showed no signs of metastatic lesions; urography showed delayed elimination of the contrast agent by the left kidney and subsequently severe left ureteropylectasia due to external ureteral obstruction in the middle third of probable retroperitoneal origin. The renal scintigram showed an irregular accumulation of the radio-isotope in the left kidney, without evidence of defects suggesting neoplastic disease.

A CT-scan of the pelvis confirmed left hydronephrosis, caused by a widespread process with

FIG. 1. CT-scan of the pelvis showing left hydronephrosis; rf = retroperitoneal fibrosis; a = aorta; rk = right kidney; lk = left kidney with hydronephrosis; b = bowel.

boundaries involving the left ureter, and also involving the aorta, inferior cava and iliac arteries.

Exploratory laparotomy revealed dense retroperitoneal fibrous tissue, mainly extended on the left abdominal wall, with a pseudo-sarcomatous appearance; multiple biopsies showed RF. Left nephrectomy was performed.

Histology of the removed tissues showed hydropnephrosis and subacute pyelitis in the kidney, a fibrous process with chronic follicular and granulomatous inflammation in the retroperitoneum, and a reactive lymphadenitis in the lymph nodes. The ureter was wrapped in dense fibrous connective tissue, sprinkled with inflammatory elements, similar to those described above.

The postoperative period was uneventful. Treatment with corticosteroids started immediately after surgery. ESR, serum creatinine and blood urea decreased rapidly. Fever and pain disappeared. The patient was discharged after a few days in good condition.

Discussion

It is thought that the RF in this case could be the result of a pathological process not previously described, namely an extension of proctitis to the retroperitoneal space (including lymph nodes), possibly with the migration of micro-organisms. On the other hand, the tendency for developing sclerosing responses, probably as a manifestation of a non-inflammatory or, more probably, inflammatory systemic disease (collagen-vascular-like process) could already be recognized in the rectal stenosis that followed the haemorrhoidectomy (see histological changes in the experimental model of 'animal-induced hypersensitivity'; Goddard, 1947).

From a therapeutic point of view, the result of combined surgery and corticosteroid therapy in this patient must still be evaluated in time. It is interesting to note that Jones *et al.* (1970) obtained remission of this disease in all their patients by this therapeutic procedure. Ross and Goldsmith (1971) also had good responses. These results must lead physicians to more careful research of RF in patients with lumbar or abdominal pain, considering the strong possibility of therapeutic success.

Appendix

Reported causes of retroperitoneal fibrosis

(a) malignant tumours: peri-ureteral metastasis (Grabstald and Kaufman, 1969); primary retroperitoneal tumours, serotonin-producing carcinoid tumours (Morin and Zuerner, 1971);

(b) any type of retroperitoneal lesion stimulating the retroperitoneum: haemorrhages with subsequent haematomas; Henoch's purpura (Cerny and Scott, 1971; Hacke, Utz and Woolner, 1962); factor VII

deficiency (Popham and Stevenson, 1960); abdominal aneurysms (Charnock, Riddel and Lombardo, 1961); trauma in the suprapubic region (Webb and Dawson-Edwards, 1967);

(c) Crohn's disease, intestinal diverticulitis, retroperitoneal appendicitis (Harlin and Hamm, 1952);

(d) peri-pelvic urinoma subsequent to urinary obstruction (Jones *et al.*, 1970);

(e) radiotherapy (Koep and Zuidema, 1977);

(f) pelvic surgery, with sectioning of the ureter followed by extravasation of urine (Koep and Zuidema, 1977);

(g) primary infections of the genito-urinary tract, with transmission of micro-organisms to the retroperitoneum (Winsbury-White, 1933);

(h) histoplasmosis, often associated with mediastinal and retroperitoneal fibrosis (Wieder and Rabinowitz, 1977);

(i) use of certain drugs, e.g. methysergide (Litz and Rooke, 1965); vaso-active drugs (Stecker *et al.*, 1974; Saxton *et al.*, 1969); methyldopa (Iversen *et al.*, 1975);

(j) association with auto-immune disorders: vasculitis (Hellstrom and Perez-Stable, 1966); Weber-Christian's panniculitis (Mitchinson, 1965); scleroderma (Mansell and Watts, 1980); processes of sclerosing fibrosis as fibrotic orbital granuloma, Riedel's struma, sclerosing cholangitis, mediastinal and retroperitoneal fibrosis (Arger, Stolz and Miller, 1973; Bartholomew *et al.*, 1963; Gleeson, Taylor and Dowling, 1970; Kittredge and Nash, 1974; Mitchinson, 1970; Palmer, Wolfe and Kostas, 1978).

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