HISTORY OF MEDICINE

RETROPERITONEAL FIBROSIS WITH INTRA-URETERAL INVOLVEMENT

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INTRODUCTION

In three previous papers, the subjects of retroperitoneal fibrosis, the genitourinary complications of surgery for abdominal aneurysms, and the genitourinary manifestations of abdominal aneurysms were fully discussed [1-4]. In this paper, the little known subject of intraluminal fibrosis of the ureter will be discussed [5].

HISTORICAL

Our knowledge of retroperitoneal fibrosis, also called periureteral, periarterial, or perianeurysmal fibrosis [6-7], has evolved with time [5]. Albarran (1905) was the first to describe “renal retention by periureteritis,” and to devise the surgical operation of liberation of the ureter [8]. The first article concerning the association of ureteral obstruction and abdominal aneurysm was not published until 1931 [9]. In the USA, it was the article of Ormond (1948), which established the new entity of “retroperitoneal fibrosis” [10].

REAL ETIOLOGY

It used to be thought that periureteral fibrosis was due to methysergide. In rare instances other causes were reported, such as a tumor or tuberculosis [6]. However, since the work of Mitchinson (1972) [12, 14], it has become evident that periarterial fibrosis is a reaction to an insoluble antigen, a ceroid, which originates in atheromata and which produces antibodies that circulate in the blood and that are mainly IgG [15-16]. Because the fibrosis has been imaged by gallium scanning, some believe it to be a true inflammation [17]; and English authors designate the condition as “inflammatory aneurysm” [18-19]. Although generally defunct, the theory that the fibrosis is the cause rather than the effect of the aneurysm is still held by a very small number of physicians [7, 20].

Case report

A 65-year-old Greek male (CK # 3163) was admitted in February 1982 with renal failure (Blood urea nitrogen 42 mg%, creatine 4.5 mg%) and 50 red cells in the urine, from an aortic aneurysm causing bilateral ureteral obstruction. He was first treated with bilateral ureteral stents followed, one month later, by ureterolysis and peritonealization of the ureters. In 1983, the right hydronephrosis recurred. An attempt at stenting the right ureter failed, even with a 5 F stent. In May, resection of the abdominal aneurysm with left to left axillofemoral and left to right femoro-femoral grafting were performed; during this procedure, the right ureter was explored and the short segment of ureter overlying the right iliac artery was found to be markedly narrowed, this segment was resected and the ureter reanastomosed over a stent. Microscopic examination showed that the fibrosis had invaded the ureteral wall (Figure 1).

CLINICAL

The fibrosis envelops the retroperitoneal structures such as the retroperitoneal system of arteries (aorta and the iliac arteries), the ureter, and the sympathetic network of nerves, and strangles them; it usually bifurcates and follows the iliac arteries. The ureters are characteristically seen on röntgenograms to be deviated medially and are frequently obstructed. This fibrosis can sometimes even compress the vena cava [21], and it can extend up to the region of the duodenum [22]. It is seen in about 7% of the cases of aneurysms [23-24], and is more frequent in diabetic males.

In 15% of the cases [25] this fibrosis does not remain periureteral but actually invades the ureteral wall, making it difficult or even impossible to catheterize the ureter, or to surgically dissect it and liberate it from the fibrous tissue engulfing it [25-30], a change in treatment plan may become necessary [29]. Sixteen such cases were reported in 1967 [31].

Liberation and peritonealization of the entrapped ureter does not seem to stop the ongoing fibrosing process which seems to continue inexorably until the ureteral lumen becomes completely occluded.

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Treatment

The initial treatment of ureteral obstruction caused by peri-arterial fibrosis is with steroids [32-34]. If this fails, a double J stent could be used; this is however fraught with the risk of the usual complications of the use of stents [35]. When the fibrosed segment of ureter is short,
The fibrosis has invaded the wall of the ureter and caused narrowing of its lumen. It can be resected surgically and the ureter reanastomosed.

Ureterolysis is however the most commonly employed form of surgery. When intraluminal fibrosis exists, partial excision of the involved ureteral segment becomes necessary; the defect could be repaired by end-to-end anastomosis, by utilizing a spiral strip of the ureter, small bowel substitution, or by iatrogenic nephropotomy; longitudinal splitting of the ureteral wall down to the mucosa may become necessary [36].

CONCLUSION

The use of the words “periureteral, periarterial, or perianeurysmal fibrosis” to describe a lesion that may become intra-ureteral is a misnomer. The term “retroperitoneal fibrosis” is more appropriate and more factual.

After the pioneering work of Mitchison, one can no longer speak of methysergide as the etiological factor.

Faced with the possibility of intra-ureteral fibrosis, the surgeon has been forced to devise new approaches to the surgical treatment of this condition.

REFERENCES

27. Skeel DA, Shols GW, Sullivan MJ, Witherington R. Retroperitoneal fibrosis with intrinsic ureteral involve-