Idiopathic Periureteral Fibrosis Demonstrating a Marked Polyuric Phase

WILLIAM MORTON GOLDBERG, M.D., F.R.C.P.[C], Hamilton, Ont.

PERIURETERAL fibrosis is a rare condition of unknown etiology in which there is fibrotic involvement, particularly of the ureters, although it may also include the aorta and other retroperitoneal structures.\(^4\) This pathologic situation results in a varied clinical picture, most often characterized by back pain, oliguria, anuria, weight loss, and at times vascular insufficiency of the lower extremities.\(^4\) \(^5\) The case presented in this report demonstrates the rare occurrence of a polyuric phase lasting many days that immediately followed a period of nearly complete anuria. To the author's knowledge, only one other case reported in the literature presented in a similar manner.\(^1\)

The patient, a 66-year-old white man, was admitted to hospital with a history of failure to pass urine for four days. He gave a one-year history of vague back pain off and on; this complaint had been investigated radiologically, with negative results. His back pain was not disabling; he carried on with his usual activities, and in fact the pain seemed to be lessening. He was feeling in his normal state of health until four days before admission, when he noted that he was not passing urine. He stated that perhaps once or twice a day he would pass at the most an ounce or so of urine, but certainly no more. On the third day of this episode he was seen by his family doctor, who catheterized him and obtained no urine. The next day he became nauseated, vomited occasionally, seemed to be a little drowsy and not quite as alert mentally; for this reason he was admitted to hospital. This patient had no specific complaints other than those noted above, and did not have any neurologic symptoms referable to his lower extremities. He had had no symptoms of vascular insufficiency. His past health had been quite good. A hemorrhoidectomy was his only previous operation. There was no pertinent family history.

On physical examination the patient appeared slightly apprehensive, was a little drowsy and looked chronically ill. His tongue was dry but there was no other abnormality of his head or neck. His ocular fundi appeared normal. His chest and heart were normal on examination. His blood pressure was 160/90 mm. Hg. His abdomen revealed no enlarged organs or masses. A rectal examination revealed that the prostate was only slightly enlarged. His extremities and skin were normal. His peripheral pulses were normal. There were no abnormalities on careful neurologic examination.

The patient's hemoglobin, white blood count, differential white blood count and sedimentation rate were within normal limits. The blood urea nitrogen was 101 mg. %; the serum calcium, phosphorus, sodium, potassium and chloride were all normal. An electrocardiogram and chest radiograph were perfectly normal.

The patient was catheterized in hospital, no urine was obtained and the catheter was left in place. He excreted no urine until approximately 10 hours after admission, when he suddenly started to void. In the next 12 hours he excreted 6000 c.c. of fluid; in the next 24 hours, 10,000 c.c. of fluid; and over the next two weeks, up to the time of his operation, he voided between 1500 and 6000 c.c. of fluid daily. His intake during this period was not excessive, and it never exceeded his output in a 24-hour period by more than 1000 c.c. Therefore the polyuria could not be accounted for on the basis of an increase in fluid intake alone. The specific gravity of the urine was 1.010 when measured on several occasions. His blood urea nitrogen over a four-day period dropped from 101 mg. % to a normal value of 18 mg. %. As well, he showed marked clinical improvement during this period.

An intravenous pyelogram showed evidence of bilateral hydrenephrosis of a minor degree with definite medial displacement of the lower third of both ureters. No obvious area of obstruction was revealed by this examination. Retrograde pyelography was carried out. Similar findings were observed, but there was a definite narrowing of both ureters at the junction of the middle and lower thirds with definite ureteral dilatation proximal to this area.

About 15 days after admission this patient was operated upon, and the right side was explored through the usual curved kidney incision. The upper third of the ureter was about three times as large as its normal size and thin-walled. It was followed downward. The middle third was encased in dense scar tissue, and there was a fair amount of edema in the surrounding tissue. The ureter was freed at this point and followed down to where it crossed the iliac vessels; in this area it appeared to be of normal calibre. At the latter location about an inch and a half of the ureter seemed to be encased in fibrous tissue, and this area felt much harder than normal. When the ureter was freed from this dense fibrous tissue, however, it did not decrease in size. For this reason, it was postulated that fibrous tissue might be invading the wall of the ureter; therefore this structure was opened and a No. 6 catheter was passed readily into the bladder. In passing a No. 10 T-tube, slight resistance was encountered in the fibrous area; the T-tube was left in place and the ureters were fixed laterally with three sutures. The patient was discharged from hospital with the T-tube in place, but it was removed approximately six weeks after insertion. The patient's subsequent course was uneventful.

He felt perfectly well when he was readmitted to hospital approximately three months after his initial operation for exploration of the left ureter. On approaching this structure, it was noted that there was considerable fibrosis in the same area described on the right side, and it was difficult at first to recognize.
the ureter. When it was recognized and freed, the most
constricted part of the ureter was found to lie in
apposition to the external iliac vessels. The ureter was
removed from this fibrous tissue and transferred to a
lateral position over the psoas muscle, and secured
with three sutures as on the right side. The roof and
floor of the former bed of the ureter were then
brought into approximation with sutures. The patient
was discharged from hospital and had an uneventful
postoperative course. When he was seen six months
after his last operation, he was free from symptoms.
An intravenous pyelogram at that time showed no
abnormality, and his blood urea nitrogen was perfectly
normal. The result of a Fishberg concentration test was
compatible with normal renal function.

**Discussion**

Bricker et al.2 in 1957 were the first to discuss the
abnormality in renal salt and water conserva-
tion which occurs after relief of acute urinary re-
tention. They studied several patients who had
lower urinary tract obstruction not due to peri-
ureteral fibrosis and who developed a marked
diuresis after release of the obstruction. They found
that these patients showed findings similar to those
of normal subjects during experimental osmotic
diuresis. This diuresis resulted primarily from de-
ivery into the urine of an excessively high per-
centage of the sodium and chloride which had been
filtered at the glomerulus. This defect in sodium
and chloride excretion could be related directly
to the suppression of the tubular reabsorption of
these elements; the defect was located primarily
in the proximal segment. This condition was there-
fore considered to represent a nephropathy due to
functional changes in the tubular epithelium, the
result of obstruction to the lower urinary tract.
The diuresis under these circumstances could not
be altered by the administration of pitressin, further
demonstrating that the defect was within the
tubules. Restoration to normal function occurred
from days to months after release of the obstruc-
tion.

In 1960, Knowlan et al.1 reported on a patient
who had a previous nephrectomy and later de-
veloped periureteral fibrosis of the remaining
ureter. This patient presented with a syndrome
of polyuria and polydipsia to a degree that a clini-
cal diagnosis of diabetes insipidus was made. This
polyuria was unresponsive to pitressin. The ureter
was freed surgically and the polyuria disappeared
within a few days postoperatively. This case re-
ported by Knowlan and his co-workers demon-
strated similar findings to those studied by Bricker
et al.2; it was the first reported case of partial
ureteral obstruction due to periureteral fibrosis that
showed this polyuric syndrome. Their patient must
have had enough physiologic damage to the renal
tubules, the result of partial ureteral obstruction,
to interfere with solute reabsorption and so result
in an osmotic diuresis.

The patient described in the present report must
have had complete bilateral obstruction of the
ureters at the onset; that is, when the symptom
of anuria was present. Thereafter, some release
of the obstruction must have occurred which led
to a massive diuresis; this was succeeded by a
moderate diuresis which continued until his opera-
tion two weeks later. While it cannot be proved,
at the time when partial obstruction existed the
patient must have had obstructive tubular nephro-
pathy which was the basis upon which osmotic
diuresis developed.

The clinical importance of the recognition of the
fact that periureteral fibrosis may present with a
polyuric phase which alternates with oliguria or
even anuria is illustrated by this case. If the patient
reported here had been examined for the first time
12 hours later, he would have been in the polyuric
phase, the history of anuria may have well been
doubted, and the diagnosis could have been missed.
He could well have been considered to be in the
end stage of chronic glomerulonephritis or pyelo-
nephritis with an obligatory polyuria. Therefore,
for any patient who presents himself with back
pain, azotemia and malaise, and has any disorder
of urinary output (even polyuria), the diagnosis
of periureteral fibrosis must be entertained. Addi-
tional symptoms described in such cases include
those of arterial insufficiency in the legs, anorexia,
nausea, vomiting, abdominal pain and even
jaundice.3 Therefore, in any patient exhibiting these
symptoms, an intravenous pyelogram should be
performed. The earliest evidence of periureteral
fibrosis in the pyelogram is a medial displacement
of the ureters; it is only later that actual obstruction
of the ureters will be encountered.4 If there is any
suggestion that this process may be present, retro-
grade pyelographic studies should be carried out
for further delineation of the problem. The results
of surgical treatment of this condition are excellent,
and if the ureters can be freed and laterally placed,
there should be no recurrence.

**Summary**

A case of idiopathic periureteral fibrosis demon-
strating a marked polyuric phase is presented.

The mechanism of this diuretic phase is discussed.
It is emphasized that, while this is an uncommon
condition, if it is not diagnosed and treated death
ultimately occurs, and if treated a complete cure is
usually obtained.

I wish to thank Dr. Harry Elliott for help in making
the correct diagnosis and for carrying out excellent surgical
treatment, and Dr. R. Morrison for referring this patient.

**References**

5. RICHES, E.: Editor, Modern trends in urology (second
series), Butterworth & Co., Ltd., London, 1960, Chapter
15, p. 159.