Tumors of the Urachus

Report of Five Cases

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• **Reports of three patients with malignant and one with benign urachal tumor are presented.**

Survival of one patient in good health 11 years after removal of adenocarcinoma of a urachal cyst with vesical invasion is reported.

Methods of diagnosis and treatment are discussed.

TUMORS OF THE URACHUS are relatively rarely reported in the literature. It is the purpose of this paper to present three additional patients with malignant and one with benign urachal tumor, and an 11-year follow-up of another patient, one of three previously reported,[14] who had adenocarcinoma of a urachal cyst with vesical invasion.

In February, 1953, Slater and Torassa[19] found reports of 70 cases of carcinoma of the urachus in the literature and added one of their own. Since then, Begg,[2] Pollock,[13] Bobrow[3] and Faulkner and coworkers[5] have increased the number to 75. The authors' three additional cases bring this total to 78, of which six have been under their personal observation.

The allantois extends from the cloaca to the placenta, leaving the embryo through the umbilical cord. The abdominal portion is called the urachus (Figure 1). Later it is intimately associated with the peritoneum. In normal development, the bladder and urachus separate from the cloaca, and the urachus then becomes obliterated. However, at any point in the urachus, epithelial-lined spaces may persist and later become cystic or malignant. If bladder neck obstruction exists the urachus may remain patent. The tumors may be transitional or revert to the cuboidal-columnar anlage and bring about mucinous adenocarcinoma.

REPORTS OF CASES

CASE 1. A white man 25 years of age was admitted to Scripps Hospital July 14, 1953, and was discharged July 31, 1953. He complained of a slow urinary stream all of his life. In 1946, dilatation of a urethral stricture and cystolithotomy were done in a government hospital. In 1952, in another government hospital, dilatation of the stricture was again carried out. Cytoscopy revealed no other cause for hematuria, which began in November, 1952. When seen by the authors in June, 1953, the patient complained of a slow stream, intermittent hematuria, recurrent dysuria and epididymitis. Upon physical examination, the only abnormalities noted were a transverse suprapubic scar, thickening of the left epididymis and inflammatory induration of the prostate.

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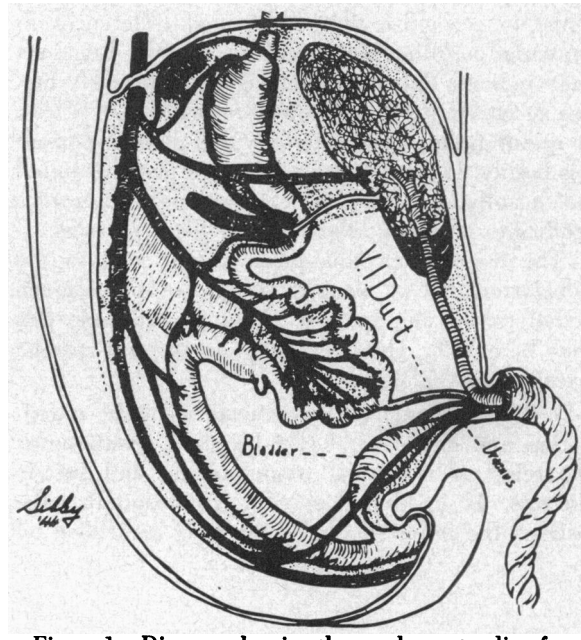

Figure 1.—Diagram showing the urachus extending from bladder to umbilicus. Bladder still connected to cloaca. (From Sibley, W. L., Cyst of Urachus, Arch. Surg., 79:465-468, 1950.)

The prostatic secretion and the urine contained many leukocytes. The urine contained many Gram negative bacilli.

Excretory urograms were normal. Cystourethroscopic examination showed a bulbous stricture to the size of a No. 14 (French) catheter, Grade I intraurethral lateral lobe prostatic hypertrophy, and a posterior commissure of Grade IV enlargement. On the posterosuperior wall of the bladder was a flat tumor, 2.5 x 3 cm. in dimension, having a papillary texture and appearing to be Grade II or more in degree of malignancy.

On July 15, 1953, internal urethrotomy, transurethral prostatectomy, and biopsy of the tumor were performed. Upon examination of a frozen section of the tumor adenocarcinoma was diagnosed

Figure 2.—Tissue section of tumor removed in Case 1 (×50). Note mucus glands.

and suprapubic removal of the tumor-bearing bladder wall, the overlying peritoneum and inferior portion of the urachus was carried out immediately. There was invasion of the peritoneal surface, but no distant metastatic lesions were present. Since the tumor was entirely inferior, the peritoneum in the midline and the navel were not removed. The wound was closed in the usual manner and the bladder was drained with a urethral catheter.

The pathological report was: "(1) Colloid adenocarcinoma, Grade I, arising in adenoma of urachus. (2) Benign prostatic hypertrophy" (Figure 2).

Postoperatively the patient did well, and was discharged on the 16th day. Dilatation of the stricture was carried out regularly thereafter. Occasional relapsing infection responded well to treatment. Cystoscopic and general examination in February, 1955, showed no recurrence.

CASE 2. A white male patient, 51 years of age, was admitted to Mercy Hospital July 5, 1951, with complaint of intermittent hematuria of four months' duration. There were no other symptoms.

No abnormalities were noted upon physical examination. The urine contained 10 to 12 erythrocytes per high dry field.

Cystoscopic examination and retrograde pyelography showed the presence of a 1.5 cm. broad flat tumor in the vertex of the bladder. The report on a biopsy specimen was "mucinous adenocarcinoma."

On July 6 the umbilicus, a full thickness of abdominal wall with attached peritoneum and a cuff of bladder including the tumor were removed en bloc. There was no tumor except at the urachal

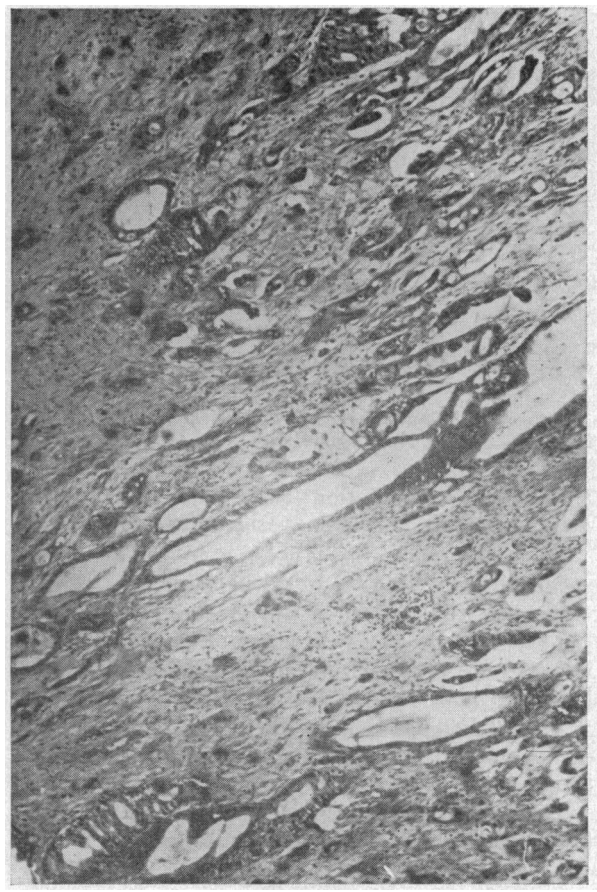

Figure 3.—Tissue section of tumor removed in Case 2 (×50). Note mucus glands.

attachment to the bladder where wrinkling of the peritoneum suggested invasion. The bladder was closed easily, but approximation of the peritoneum and fascia was difficult.

The pathologic diagnosis: "Adenocarcinoma of bladder, Grade II (Urachus)" (Figure 3).

The patient did well and was discharged July 18. Cystoscopic examination was done on October 11, 1951, and no abnormality was noted. On January 8, 1952, local recurrence in the wound was evident. That lesion was excised, but there were numerous tumors in the peritoneal cavity. The patient died September 22, 1952. At autopsy no other primary tumor was observed.

CASE 3. A negro woman, 56 years of age, was admitted to Mercy Hospital August 22, 1951, with complaint of soreness about the navel, low backache and diarrhea of four weeks' duration. In the navel was a hard, button-like growth 2 cm. in diameter. There was no ulceration. In the left lower quadrant of the abdomen there was a palpable mass extending to the midline in the region of the bladder. Upon bimanual examination it was noted that the mass extended beyond the midline, and rectal examination suggested high annular constriction.

Results of examination of the urine were within normal limits. Excretory urograms were normal.

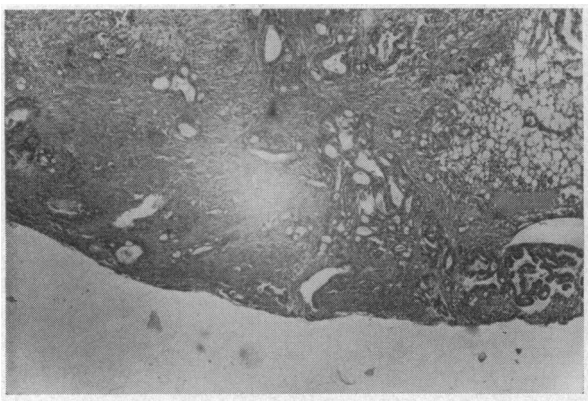

Figure 4.—Tissue section of umbilical tumor removed in Case 3 (×50). Note mucus glands.

Cystoscopic examination showed an extravesical mass to the left, and intact vesical mucosa.

No abnormality was observed at biopsy of a rectal specimen from the level of involvement.

At operation the navel and the tumor in it were removed. Upon peritoneal exploration, metastatic lesions were observed on bowel surfaces, omentum, liver and both ovaries. The ovaries were removed.

The pathologic report: "Primary colloid adenocarcinoma of umbilicus (urachus) with metastasis to omentum and bilateral metastasis to ovaries."

The patient died December 10, 1951. Permission for postmortem examination was refused.

CASE 4. A white man, aged 45 years, was admitted to Scripps Hospital May 5, 1953, with complaint of a foul, purulent discharge from the navel, first noted in 1933, but recurring April 12, 1953. There was moderate pain about the navel and a pulling sensation in the navel on urination. These conditions had been noticed at the time of the episode in 1933 also. Hair was noted in the material discharged from the navel. In 1927 a dermoid cyst had been removed from the area of the right twelfth rib.

Upon physical examination, inflammation was observed about and below the navel. The urine and prostatic fluid were normal. Mixed bacteria, but no tubercle bacilli, were seen upon examination of material discharged from the navel.

Excretory urograms and retrograde air and opaque cystograms were normal. A flexible hollow probe was introduced through the navel for a distance of 4 cm. Upon injection of 2 cc. of iodochloral, a cavity 2.5 cm. in diameter below and to the right of the navel was visualized (Figure 5).

Cystourethroscopic examination revealed only mild posterior urethritis. No abnormalities were seen in barium contrast roentgenographic studies of the entire gastrointestinal tract.

At operation the navel was circumcised and the incision was carried to the symphysis. The rectus fascia was opened and the peritoneum was opened above the navel. Adhesions were freed and the navel, the tumor, attached peritoneum and rectus

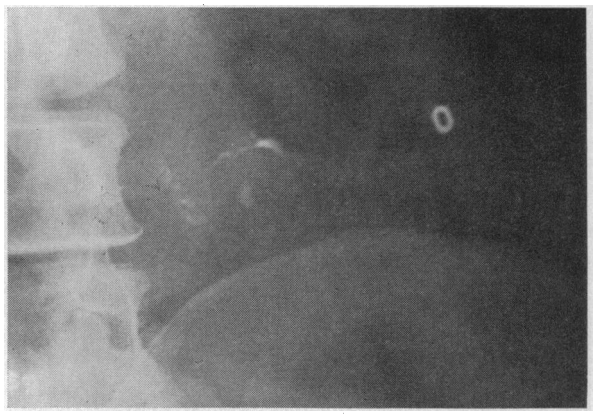

Figure 5.—Left oblique radiograph of abdomen in Case 4 after injection of iodochloral through navel sinus. Note deep cyst outline. The "O" marks navel.

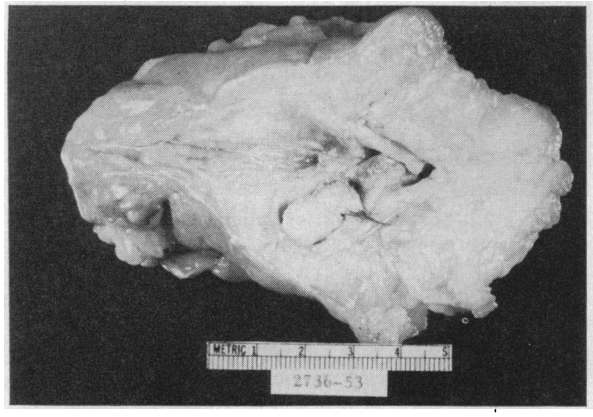

Figure 6.—Gross specimen of tumor-cyst removed in Case 4. Note cyst surrounded by mass of fibro-fatty tissue.

muscle were excised along with the urachus and the dome of the urinary bladder. Grossly the tumor appeared malignant (Figure 6). Repair of the bladder was routine, but the peritoneum and abdominal wall were closed with difficulty.

The pathologic diagnosis was: "Chronically inflamed urachal cyst containing acutely inflamed ulcerated polyps and hair mixed with debris." (See Figure 7.)

The patient was alive and well in February, 1955.

CASE 5. In this case (previously reported[14]) a white woman 38 years of age had radical excision of a urachal cyst containing colloid carcinoma and invading the bladder (Figure 8). Radon seeds were placed in the bladder wall and the rectus muscle. The patient was alive and well when examined cystoscopically some ten years after operation. In January, 1955, she reported herself to be well.

DISCUSSION

The symptoms of urachal tumors and cysts may be classified as follows: (1) Local—tumor and infection about or below the navel (Case 4); (2) Peritoneal irritation, when infected; (3) Gastro-

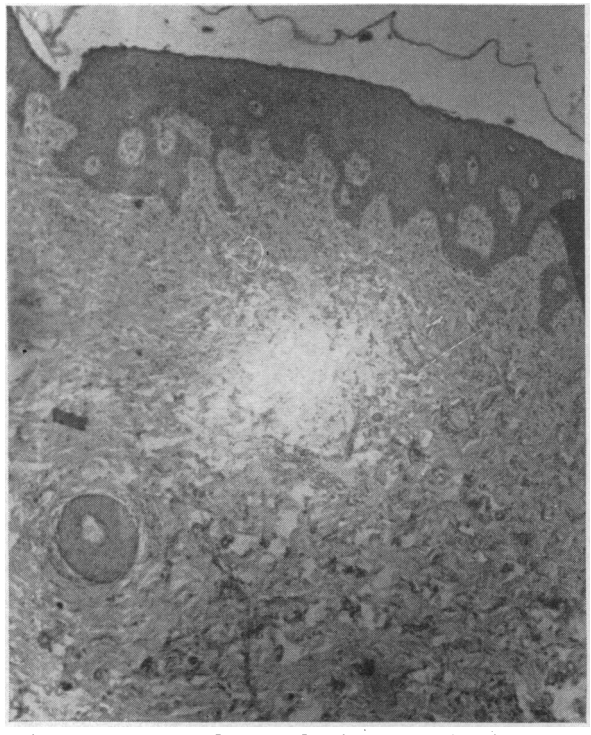

Figure 7.—Microphotograph of section of wall of cyst removed in Case 4 (×50). Note epidermoid lining and hair follicles.

intestinal in the presence of adherence of bowel, or in metastases to bowel or liver (Case 3); (4) Vesical, including dysuria, hematuria or pain referred to navel on urination (Cases 1, 2 and 4); (5) Genital, as in women with palpable midline tumor of mistaken pelvic origin (Case 5). Specific symptoms demanding consideration of urachal disease are suprapubic tumor, discharge of material from the navel, and pain in the navel on urination.

Essential examinations in cases in which urachal abnormality is suspected include: Physical examination, including bimanual; upper gastrointestinal and colon barium contrast x-ray study; sigmoidoscopic, cystoscopic and pyelographic examination; radiopaque visualization of draining umbilical sinuses (Figure 5); and transurethral biopsy of tumors in vesical vertex. Not all of these procedures were carried out in the group of patients herein reported upon. Routine application, however, would eliminate diagnostic error and uncover possible complicating factors.

Physical signs suggestive of urachal disease are inflammation of and drainage from the navel (Case 4). Tumor may be present in the umbilicus or at any point down to the symphysis (Cases 3 and 5). The tumors are usually midline. Upon bimanual palpation, preferably under anesthesia, the physician may discover unsuspected masses. Signs of peritoneal inflammation may occur with infected cysts

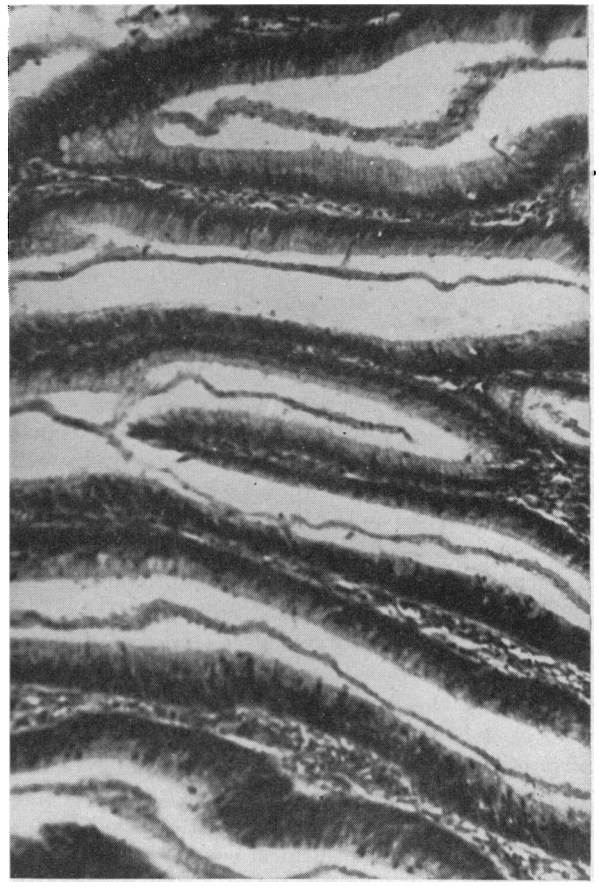

Figure 8.—Microphotograph of tissue section of tumor removed in Case 5 (×150). Note columnar mucus cells.

because of their intimate relationship to the peritoneum; some cysts or tumors may be almost intraperitoneal.[15]

Cystoscopy may reveal normal findings (Cases 3 and 4), even in the presence of hematuria, as the tumor may arise above but drain into the bladder through a patent duct.[2] An extravesical mass, with or without bullous mucosal edema, may be seen. Usually a flat infiltrating tumor is seen in the vertex (Cases 1, 2 and 5). It is most often glistening and medullary but may be papillary (Figure 8) (Cases 1, 2 and 5). Most such tumors are about 2 to 4 cm. in diameter, but some are very extensive. At times the extravesical cyst depresses the vertex of the bladder as in pregnancy.

Radiographic studies are helpful. Usually studies of the upper tract are negative, but large cysts, with or without tumors, may displace the ureters laterally by traction on the transversalis fascia. Extremely large cysts, filling the pelvis, may cause hydroureter above the brim of the true pelvis. Cystograms will reveal superior compression and possibly lateral displacement of the bladder in the presence of large tumor-cysts. Roentgen visualization of cysts by in-

jection of radiopaque material into the draining umbilical sinus will settle the diagnosis (Figure 5).

Differential diagnostic possibilities include: Inflammatory and malignant lesions of the sigmoid colon; tumors and cysts of internal female genitalia; primary tumors of the bladder; and disease of the vitelline duct when the primary findings are in the umbilicus (Figure 1). Following the suggested seven diagnostic steps should eliminate error even if the biopsy is reported as carcinoma of the rectosigmoid.

The treatment is always radical excision, even in the case of benign cysts, as they may become malignant (Case 4). Theoretically this should include the navel, urachus, adjacent peritoneum and a cuff of the bladder. The authors recommend this for lesions beginning in the navel, and feel it is best in all cases. However, in the group of cases herein reported, two of the survivors had only the bladder cuff and proximal urachus removed. When the umbilicus and upper urachus are removed, difficulty in closing is certain (Cases 2 and 4).

Whether the lesion is primarily umbilical or vesical, it is mandatory to open the peritoneum for inspection. Umbilical lesions may be found to be due to vitelline duct persistence (Figure 1) and have connection with the bowel. With the latter in mind it is wise to have general surgical consultation. In any case, opening the peritoneum permits exploration for metastatic lesions, division of local adhesions and wide excision of the disease.

Radiation therapy is of doubtful value. However, the patient, who is living 11 years after operation, had radon implanted in the bladder and the abdominal wall, and although this delayed healing, it may have helped survival (Case 5).

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