RHABDOMYOSARCOMA OF THE PROSTATE IN CHILDHOOD

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Introduction

Malignant tumors of the lower urinary tract in children are less common than upper urinary tract tumors. This has been stated in articles and textbooks, and we have also found it to be true in our department. Since 1961, we have seen 19 patients wiith Wilms tumors and 25 with neuroblastomas, but we have had only three cases of sarcomatous lesions of the lower urinary tract in children, one of which is presented here. Since the first case was reported, there have appeared in the literature close to 300 reports of this condition in all ages but less than 40 in children, most of whom were under the age of ten. Most authors report only two to four cases, and the largest series comprises only five cases reported by Williams in 1964.

Case Report

Y. E., about two years old (exact date of birth unknown) was referred to the urology clinic of Hacettepe Children's Hospital on April 5, 1964. At that time he had an indwelling urethral catheter because of urinary retention. According to his father, the child had had some kind of urinary difficulty since birth, such as frequent urination, crying while voiding, etc. About a month prior to referral to our hospital, he had been taken to the nearest State Hospital because of dribbling of urine. A urethral catheter was inserted and left indwelling for 18 days. An intravenous pyelogram was taken by the attending physician. He told the father that the child had a tumor in his bladder and should be taken to Hacettepe Children's Hospital.

Family history revealed that the child's mother had died of breast cancer, and a brother had died at the age of nine months following a five day period of urine retention.

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Physical Examination : The patient appeared to be a well-developed, well-nourished child, with a urethral catheter. First degree burns and tenderness were noted in the suprapubic region. Inguinal lymph nodes were palpable.

Laboratory Examination : Hemoglobin, 11.25 gm; white blood count, 12,200, 62 segmented, one mono, and 37 lymphocytes. Urinalysis: specific gravity, 1:015; microscopic examination revealed many white blood cells. Urine culture: over 100,000 colonies of E. coli were grown, which were resistant to all kinds of antibiotics. N.P.N., 32.5 mg/%; Co_2, 22.3 mEq/L; Na, 143 mEq/L; K, 5.2 mEq/L; Cl, 105 mEq/L. X-rays of the chest were clear. Intravenous pyelogram revealed no radiopaque densities on the flat plate of the abdomen. There was good excretion of the dye from both kidneys in five minutes. Bilateral caliectasis, pelviectasis and ureterectasis, tortuosity of the ureters, and elevation of the base of the bladder were pertinent findings (Figure 1).

Cystoscopic examination was made with the patient under general anesthesia on April 9, 1964. A number 12 F McCarthy infant cystoscope was used, and the examination was confined to the vesical neck because of inability to visualize the bladder. There was an irregular, botryoid appearance of the vesical neck and prostatic urethra. Cystoscopic examination was discontinued and rectal examination was made which revealed a hard, firm, smooth mass in the prostatic region, measuring six by eight centimeters, and semi-fixed. A biopsy specimen was taken with a Vim-Silverman biopsy needle. Our clinical diagnosis at this point was rhabdomyosarcoma of the prostate and the diagnosis was confirmed by the pathologist (Figures 2, 3, 4 and 5).

On April 16, with the patient under endotracheal anesthesia, a total cystectomy, seminal vesiculectomy and prostatectomy with bilateral cutaneous ureterostomy was performed. The patient was given 600 cc of whole blood during surgery and tolerated the operative procedures well. The post-operative course was uneventful and he was discharged on May 9, 1964 in good condition. The father was advised to bring the boy to the hospital once every three months, but we have not seen him since discharge. However, the father writes that the patient is well except for slight stenosis of the ureterostomic stomas, which require dilatation by the local family doctor.

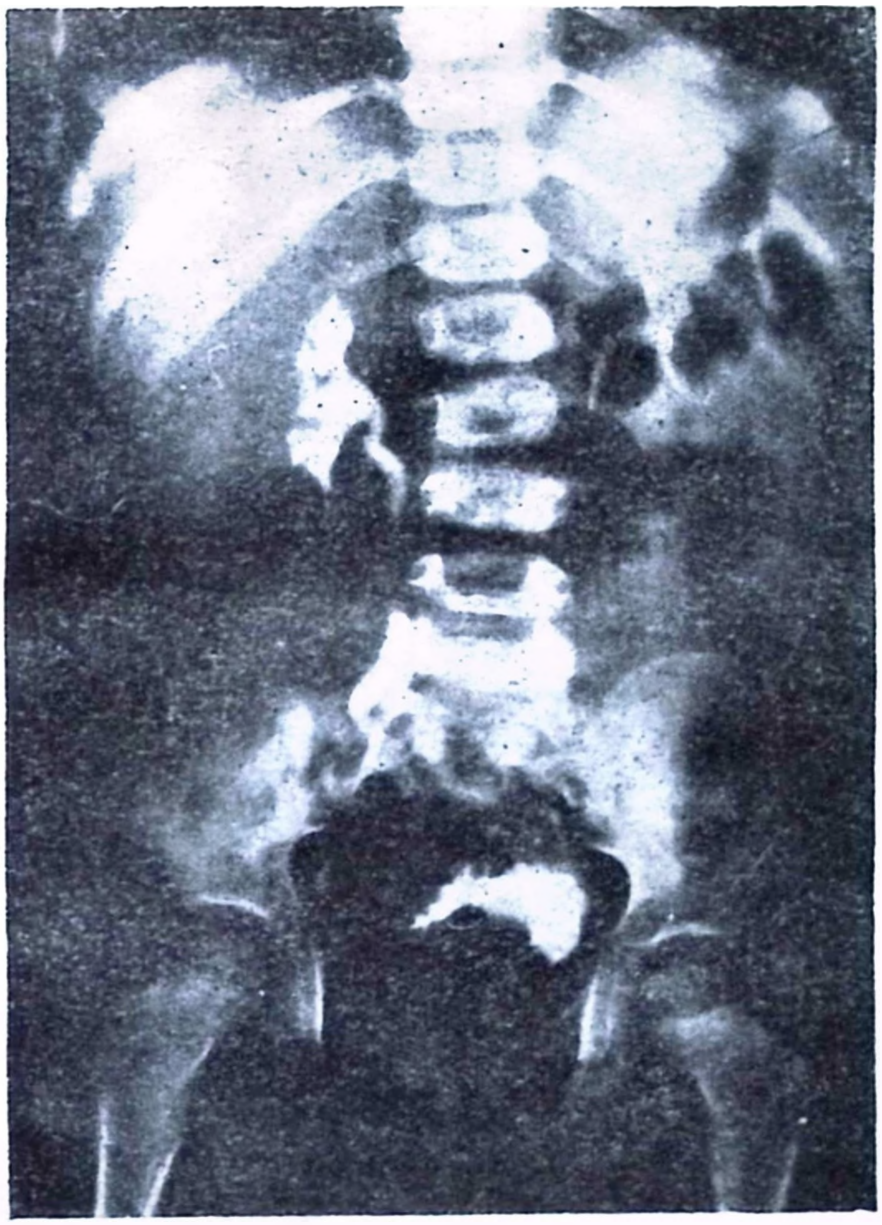

Figure 1. Intravenous pyelogram shows good bilateral excretion of the dye with a large filling defect at the base of the bladder.

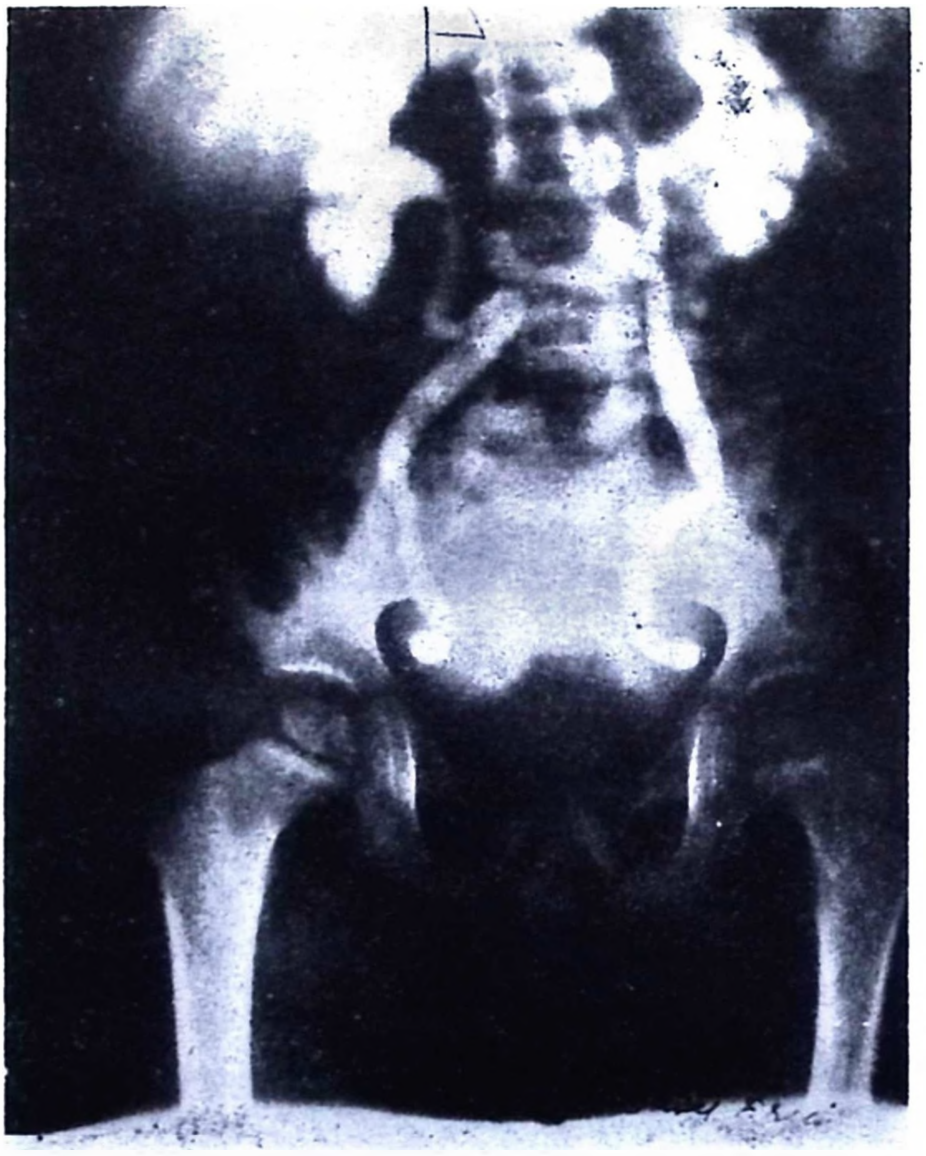

Figure 2. Intravenous pyelogram one month after operation, showing hydronephrosis and hydro-ureters.

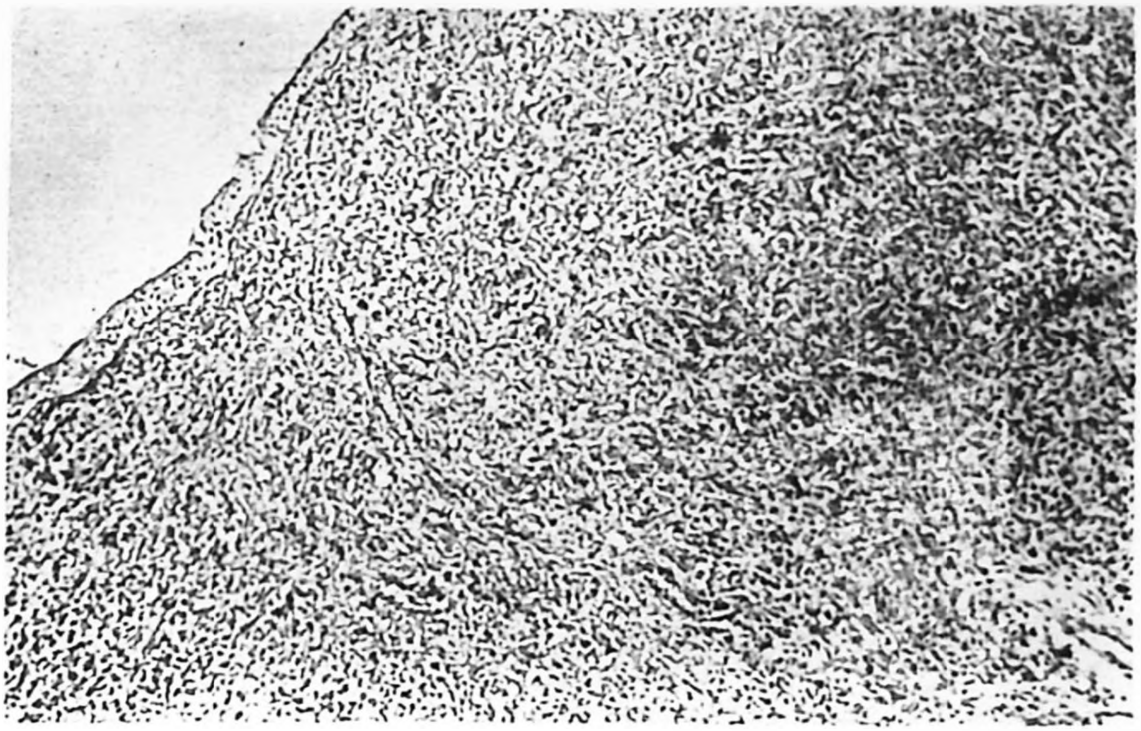

Figure 3. Normal transitional cells appear in the left upper corner; tumoral cells below.

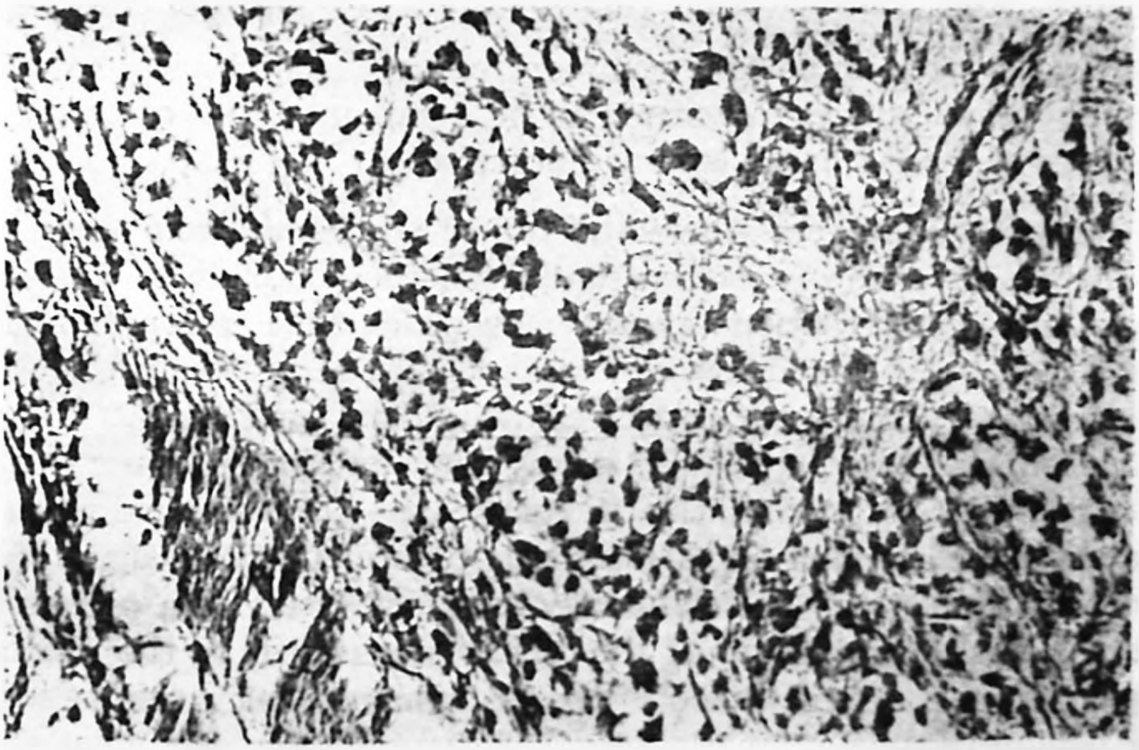

Figure 4. Pleomorphism, giant cells, and loose edematous stroma in the tumoral cells.

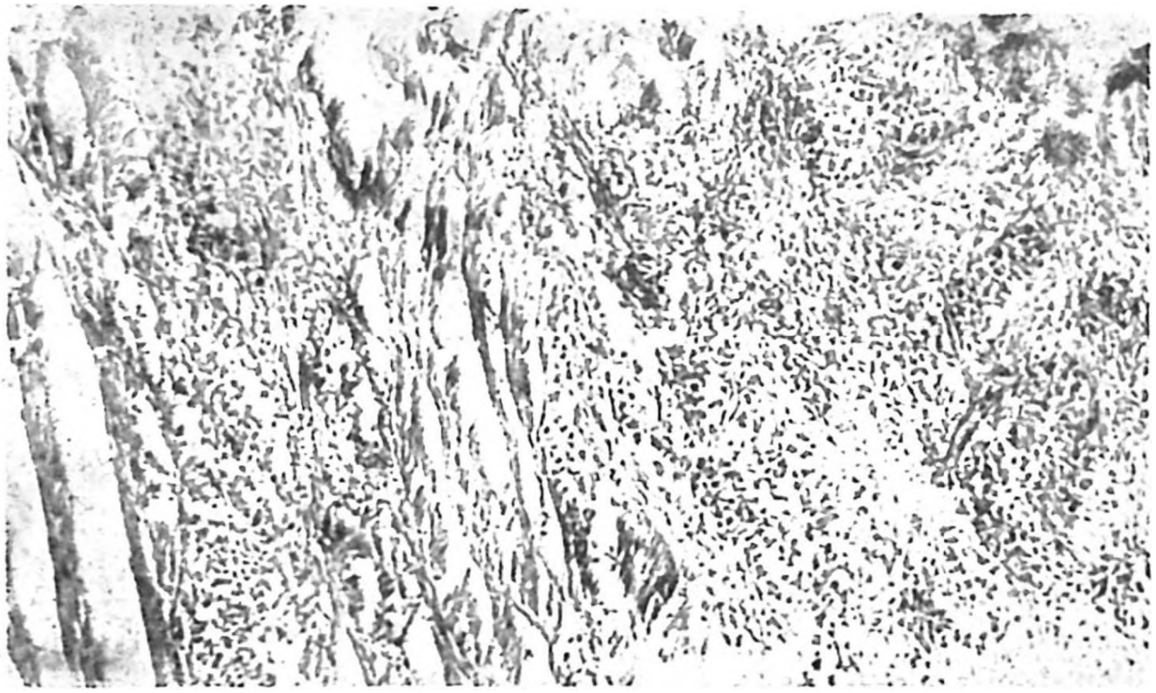

Figure 5. Tumor infiltration through the walls of the bladder.

Discussion

Many different names have been used to describe these sarcomatous lesions: leiomyosarcomas, myxosarcomas, fibrosarcomas, chondrosarcomas, rhabdomyosarcomas, and lymphosarcomas of the prostate. The classification is sometimes perplexing, but all the tumors have one important common feature: histological examination shows spindle cell elements to be dominant in all of them. The etiology of these tumors is as yet unknown.

In those instances where the tumor originates from some part of the bladder, the patient usually has no complaints until the symptoms of obstruction appear, by which time the prognosis is poor. In cases of rhabdomyosarcomas arising at the vesical neck or the prostate, urinary obstructive symptoms, such as difficulty in urination, dribbling, dysuria, stranguria, and acute urinary retention, are the first to be seen. In some cases, an abdominal mass in the suprapubic region or perineal bulging are the first signs of the condition. Infections are commonly seen, but hematuria occurs less often. Rectal examination, if the tumor is confined to the prostatic region, reveals a firm, smooth, tender, symmetrical mass, resembling benign prostatic hypertrophy. Differential diagnosis must eliminate cysts of the Mullerian duct, prostatic cysts, or absesses. Intravenous pyelograms show filling defects or elevation of the base of

the bladder, as in our patient. Hydro-ureter and hydronephrosis are also seen at this stage.

Cystoscopic examination sometimes may be unsatisfactory because of poor visualization of small endoscopic instruments. Irregularities and elongation of the prostatic urethra are pertinent findings. In our case, the typical botryoid appearance of the vesical neck was noted.

To date, there has been no agreement on the proper treatment for this condition. Most authors would agree that, if the tumor is resectable, i.e., not adherent to the pelvic organs and without metastases, the treatment of choice is radical excision of the prostate, bladder and seminal vesicles. Although the tumor is not radiosensitive, radical prostatectomy with radiotherapy is justified by some authors. Radiotherapy is usually given pre- and post-operatively in dosages varying between 2100 R and 7500 R.

Prognosis is generally poor. According to Henry, the morbidity rate is 93 per cent. Most patients die within two to three years, but at least one patient is reported to have survived for ten years.

Summary

The case of a two year old patient with rhabdomyosarcoma of the prostate is presented. In spite of the generally grave prognosis in such cases, this patient underwent radical treatment and is still alive after 22 months, with no indication of recurrence of the disease.

REFERENCES

1. Williams, D. I., and Schistad, G. : Lower urinary tract tumors in children, Brit. J. Urol. 36 : 51, 1964.

2. Danckers, U. F., Phillipsborn, H. F., Baylor, J. E., and Burkhead, H. C. : Sarcomas of the prostate, periprostatic tissue and bladder neck in children, Am. J. Roentgenol. 84 : 555, 1960.

3. McPhail, J. L. : Rhabdomyosarcoma of the prostate, J. Urol. 87 : 617, 1962.

4. Johnson, W. W. : Rhabdomyosarcoma of the prostate with extensive central nervous system metastases: a case report, J. Urol. 84 : 758, 1960.

5. Siegel, J. : Sarcoma of the prostate: a report of four cases and a review of current therapy, J. Urol. 89 : 78, 1963.