TUMOURS OF THE URINARY TRACT IN CHILDREN

RICHARD A. MOGG, *V.R.D.*, M.CH., F.R.C.S., *Consultant Urologist, United Cardiff Hospitals and Welsh Hospital Board*

Tumours of the urinary tract in children, in addition to being interesting and excessively rare, provide a fascinating study. In this paper a personal series of tumours of the urinary tract in children is presented; some of the tumours have been described briefly in a previous paper, but are included to make a comprehensive study and to present progress reports and survival figures. Two of the tumours described, an atypical embryoma of the kidney in a premature infant and an anaplastic carcinoma of the bladder in a child, are to date the youngest recorded cases with survival in the world literature. A detailed analysis of the tumours described in this paper is shown in the table at the foot of the page.

CARCINOMA OF THE ADRENAL

The first tumour of the series is in fact not strictly renal, since it originated in the adrenal gland, invading the kidney secondarily, but the method of presentation, the physical signs and pyelographic appearances are of extreme interest.

Carcinoma of the Adrenal Cortex (Michael, Aged 2 Years)

This small boy was admitted on account of abdominal colic and vomiting with a history of the passage of watery stools of 38 hours' duration. Routine examination of the abdomen revealed a tender mass in the left hypochondrium, thought to be of renal origin, and a provisional diagnosis of an infected left hydronephrosis or a renal tumour into which haemorrhage had occurred was made. The child at this time had a low-grade fever ranging from 99·6°F. to 100·6°F. An intravenous pyelogram showed a poor pelvi-calyceal pattern with distortion of the upper calyces on the left side, the right side being normal, whereas a retrograde pyelogram showed not only distortion, but displacement of the upper group of calyces on the left side (Fig. 1). Exploration of the renal area through a left lateral incision revealed a large haemorrhagic tumour, adherent to the posterior peritoneum, and apparently arising from the upper pole of the kidney. After tedious dissection a left nephrectomy was performed since it was considered on the operative findings that the tumour was a nephroblastoma into which there had been a recent haemorrhage. Macroscopically the specimen appeared to be of adrenal origin, and microscopy confirmed it as a carcinoma of the adrenal cortex (Fig. 2). The recent haemorrhage into the tumour had obviously been responsible for the atypical symptomatology and physical signs. An uneventful convalescence ensued, the child is fit and well 12 years later and has achieved academic distinction in having passed the formidable 11-plus examination in a creditable manner.

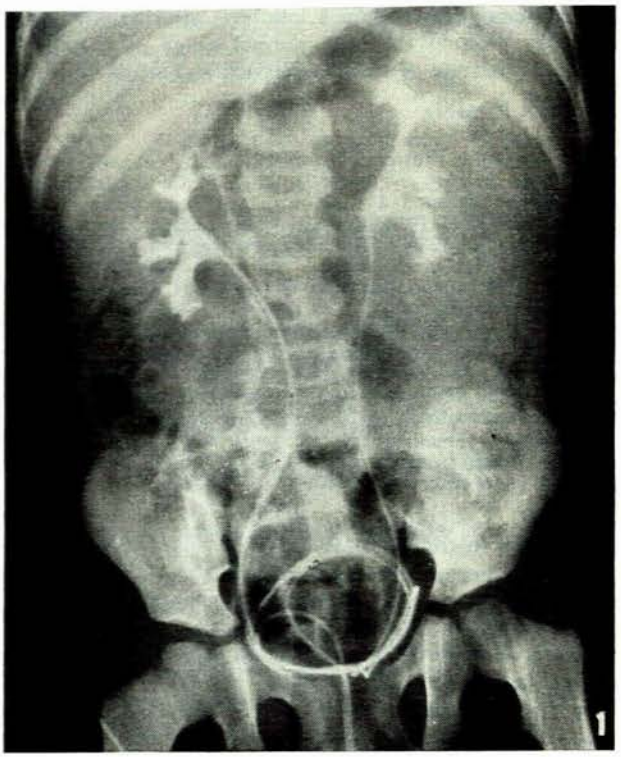

Fig. 1. Retrograde pyelogram illustrating changes in upper calyces on the left side.

Name	*Sex*	*Age (years)*	*Type of tumour*	*Operation*	*Survival*
Michael	M	2	Carcinoma of adrenal invading kidney	Left nephrectomy	12 years, alive and well
Janet	F	Premature, 34 weeks	Embryoma right kidney	Right nephrectomy	11 years, alive and well
Roger	M	10	Carcinoma right kidney	Right nephrectomy	Died 10 months after operation
Patricia	F	15	Renal hamartoma	Right nephrectomy	10 months, alive and well
Terence	M	6½	Anaplastic carcinoma of bladder	Partial cystectomy	3¼ years, alive and well
Andrew	M	3¼	Pedunculated fibromyoma of posterior urethra	Resection	2 years, alive and well
Philip	M	12	Pedunculated fibromyoma of posterior urethra	Resection	6 months, alive and well
Simon	M	7	Rhabdomyosarcoma of prostate	Laparotomy	Died 7 days after operation
Peter	M	1½	Myxosarcoma of prostate	Suprapubic cystotomy	Died 4 months after operation

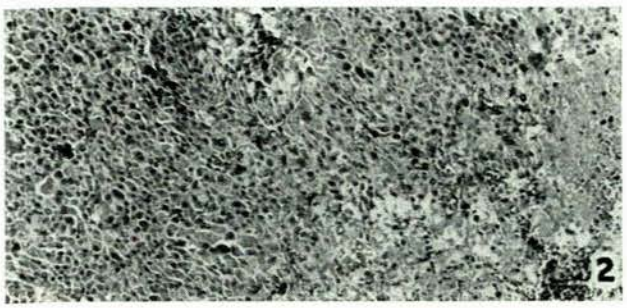

Fig. 2. Photomicrograph of adrenal tumour.

EMBRYOMA OF THE KIDNEY

The next tumour is in fact a highly differentiated embryoma of the kidney which occurred in a premature infant and was removed successfully with a 12-year survival to date. After an exhaustive survey of the world literature, this appears to be the youngest patient with a nephrectomy on record for a renal embryoma who has survived. Renal embryomas have always interested the urologist and pathologist, and the multiplicity of terms applied to these tumours indicates a wide variation in structure and histogenesis. Nephroblastoma, fibrosarcoma and embryosarcoma are some of the descriptive terms formerly used to describe the embryoma. It is now agreed that these terms merely describe a tumour in which varying degrees of tissue differentiation have occurred.

Silver (1947), in an interesting and critical review of 18 cases seen between 1926 and 1944 at the University of California, observed the interesting clinical feature that hypertension occurred in 7 of the 8 recorded cases; the hypertension disappeared on removal of the tumour, but recurred when metastases developed. He postulated that there was some pressor substance in the tumour which caused this change to occur.

Ladd and White (1941), reviewing the voluminous literature in respect of Wilm's tumours up to 1941, found that in a series of 563 cases treated operatively only 38 were alive and well at the end of a 5-year period. Hartenstein (1949) reported a case of a renal embryoma in a full-term child who lived for 2 hours, the tumour having caused obstruction to labour, and he drew attention to the fact that the autopsy specimen showed normal areas of renal tissue throughout the tumour. He postulated that these areas were not islets of normal kidney, but were highly differentiated portions of the embryoma, which had formed normal renal tissue within the tumour. It is universally accepted that the survival rate is poor, but on the other hand, the more differentiated the tumour, the greater the chance of survival. The embryoma described below presented as a most highly differentiated tumour which was first thought to be a fibrosarcoma, and it was discovered during routine examination of a premature infant; I think it testifies to the value of routine examination in all cases.

Embryoma of the Kidney (Janet, Born Prematurely at 34 Weeks)

During routine neonatal examination of this premature child a small swelling in the region of the right kidney was discovered and was thought to be an enlargement of the kidney. An attempt to obtain a pyelogram soon after birth was not completely successful and there were inconclusive findings. A subsequent intravenous pyelogram, however, showed secretion from the left kidney, and the presence of a right renal shadow but without secretion from this side. The child at birth weighed only 3 lb. 4 oz. Four weeks later, at which time it weighed 4 lb. 3 oz., a right nephrectomy was performed. The kidney was surprisingly mobile and was found to contain a tumour which replaced approximately $\frac{2}{3}$ths of the substance. Nephrectomy was performed without difficulty and there was no evidence of extrarenal spread of the tumour. Macroscopically the tumour was pearly white in colour without any evidence of haemorrhage into its substance, and the cut surface showed well-defined, interlacing bands of fibrous tissue (Fig. 3). A preliminary microscopic examination suggested that it was a fibrosarcoma, but more detailed studies of other portions of the kidney revealed an islet of cartilage within its substance, and as a result of this finding it was reclassified as a renal embryoma. The child is still alive and well 11 years later, though showing mental retardation presumably resulting from the effects of a profound toxaemia which her mother developed during pregnancy.

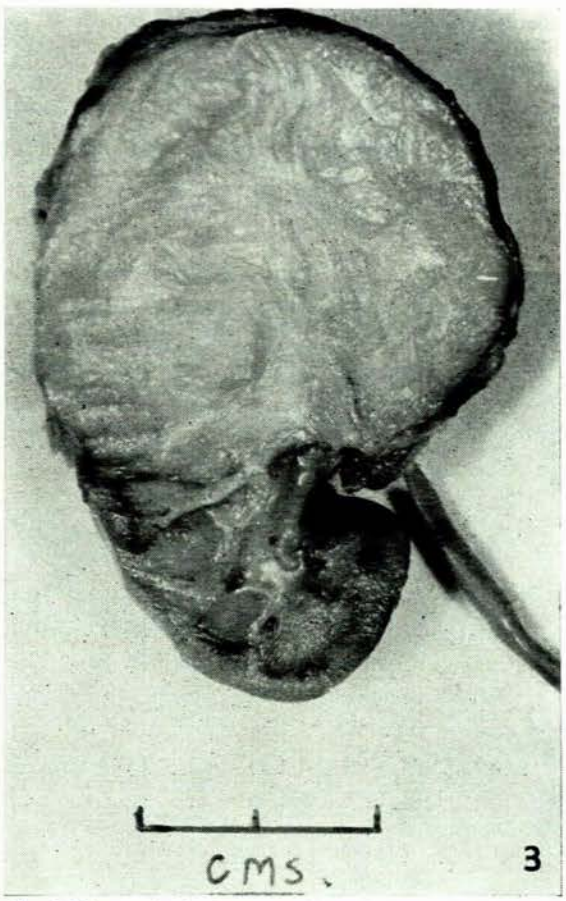

Fig. 3. Photograph showing cut surface of the renal embryoma.

CARCINOMA OF THE RENAL TUBULES

Carcinoma of the renal tubules in children is rare. The case to be described occurred in a boy aged 10. Philip and Salen (1913) reported a similar case in a 19-month-old girl, but on the other hand Meredith Campbell (1951), quoting from an extensive experience, stated that he had only once encountered a carcinoma of the renal tubules,

in a 7-year-old boy, in a series of 52 cases of renal tumours in young children. Ladd (1941) reported a carcinoma of the kidney occurring in a boy of 12. As a complete contrast the American Registry of Tumours reports only 4 proved carcinomata of the kidney in children, the age groups being 6, 12, 13 and 14 years, and only one of these children survived for 6 months. It would appear that this is a more lethal disease in the young than it is in older people.

Carcinoma of Right Kidney (Roger, Aged 10 Years)

This boy presented with a sudden profuse painless haematuria. Unfortunately the child was a 'spastic' and exhibited considerable mental retardation as a result of a birth injury. An intravenous pyelogram showed evidence of a space-occupying lesion in the right kidney, and cystoscopy revealed a marked efflux of blood from the right ureteric orifice. In spite of the age and the mental impairment, a pre-operative diagnosis of carcinoma of the kidney was made, though with some reservation, since the symptoms and findings could have been due to an atypical form of tuberous sclerosis with an associated renal hamartoma. Right nephrectomy was performed without difficulty, and there was no apparent renal vein involvement at operation (Fig. 4). Convalescence was uneventful and he was discharged home. A section of the tumour showed the typical appearances of a carcinoma of the renal tubules (Fig. 5). Unfortunately he died 10 months later from metastases in the para-aortic glands, in the line of the operative incision, and the lungs.

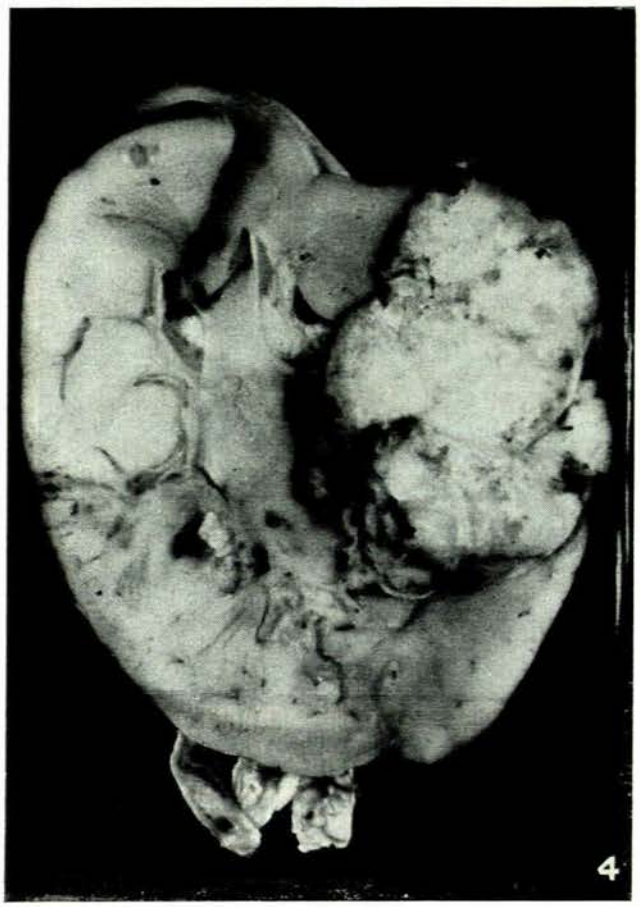

Fig. 4. Nephrectomy specimen showing carcinoma of the renal tubules.

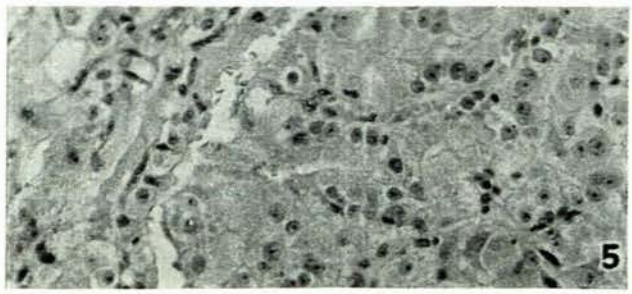

Fig. 5. Photomicrograph of renal tubular carcinoma.

HAMARTOMA

The last tumour of the upper urinary tract to be described is a hamartoma occurring in a child of 15 years. This child presented many interesting features and initially presented a problem in diagnosis.

Renal Hamartoma (Patricia, Aged 15 Years)

This child was first seen in December 1961 with a history of 4 days' pain in the right loin associated with vomiting and an irregular fever varying from 100°F. to 101°F. There was a large tender swelling in the region of the right kidney and an X-ray showed a soft-tissue mass and absence of a psoas shadow. There was in addition a white cell count of 18,400. An intravenous pyelogram showed a normal left kidney, but on the right side there was elongation and displacement of the lowest calyx, below which was a large soft-tissue shadow. In view of the raised white cell count, pyrexia, and the large renal swelling, a provisional diagnosis of renal carbuncle or of rapid necrosis within a renal neoplasm was made. Response was dramatic to penicillin therapy, the mass decreased rapidly in size, the patient became apyrexial and convalesced rapidly. She was next seen in March 1962 because there had been a recurrence of her right renal pain, and it was noted that the renal mass had increased in size. As a result of this dramatic change, exploration of the right kidney was performed and a large soft-tissue swelling was seen to arise from and be continuous with the lower pole of the right kidney. Nephrectomy was performed, there was no evidence of any renal vein involvement by the tumour, and an uneventful convalescence ensued. A section of the excised kidney showed that it was a benign mesodermal hamartoma of the kidney in which smooth muscle and vascular spaces were the predominant components. In retrospect it is considered that the pyrexia was due to haemorrhage into this rather vascular tumour, but this did not explain completely the elevated white cell count. The patient is now fit and well and has returned to her scholastic duties.

ANAPLASTIC CARCINOMA OF THE BLADDER

Tumours of the lower urinary tract in children, though extremely rare, have on occasion been reported in the literature. The majority are usually mesodermal in origin, and the most commonly encountered variety is the sarcoma. The prognosis is always grave, though D. Innes Williams in a personal series has reported survival following cystectomy in several cases of sarcoma of the bladder. Epithelial tumours in the lower urinary tract in children, on the other hand, are the most rare lesions; in fact Beer and Hyman in their book state that 'lower urinary tract tumours are so rare that an extensive discussion on their symptomatology and prognosis is hardly necessary'. In their series of 500 cases of neoplasm of the urinary tract, an epithelial tumour of the lower urinary tract was not encountered. Deming reported 64 tumours of the urinary tract occurring in the 1st decade, and the predominant tumour encountered was sarcoma; only one

epithelial tumour, a papilloma of the bladder, was described. Vesical carcinoma has not been encountered in the young, and there is to my knowledge no authentic case recorded in the literature.

Formerly considerable confusion has occurred regarding the exact origin of the reported bladder tumours in children since, very often, when first seen the lesion was so far advanced that it was impossible to ascertain the site of origin of the tumours. It is now recognized that a large proportion of these tumours were primarily prostatic and of mesodermal origin, invading the bladder secondarily. Deming pointed out that the maximum incidence of the tumour was at 2 years and that males were more affected than females, and the mortality in his series was approximately 90%. Albarran, in a series of 252 tumours of the urinary tract in children, had only encountered 6 cases before the age of 10. The case here described is an anaplastic carcinoma of the bladder which occurred in a boy of 6½ years who survived, this being the only reported carcinoma of the bladder encountered and treated in a child.

Carcinoma of the Bladder (Terence, Aged 6½ Years)

This child was first seen in May 1959 on account of a transient painless haematuria associated with some frequency of micturition. Routine examination revealed a very healthy child with no clinical abnormalities. An intravenous pyelogram showed a normally functioning upper urinary tract, but in the cystogram film there was a filling defect on the right side of the bladder suggestive of a vesical tumour (Fig. 6). On

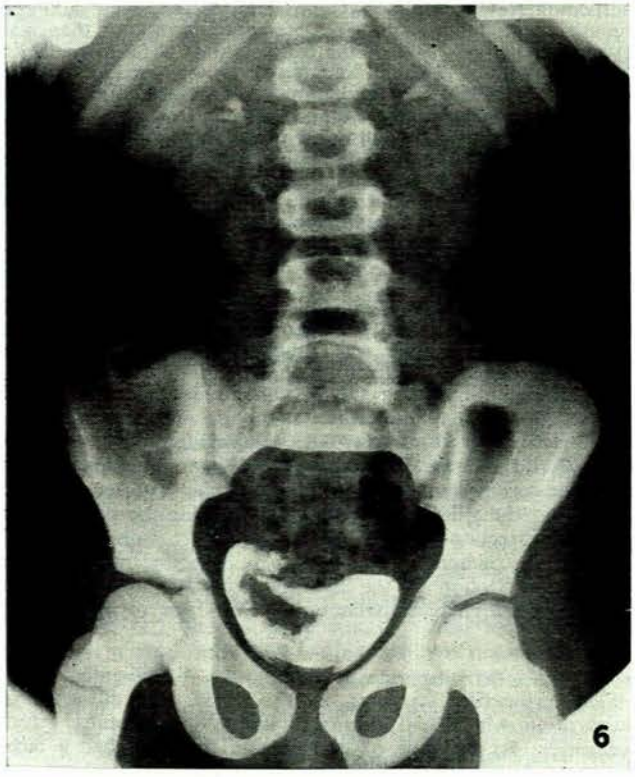

Fig. 6. Cystogram showing filling defect on right side of bladder.

cystoscopy a papillary solid carcinoma of the bladder of the adult type, surrounding but not obstructing the ureter, was demonstrated. Bimanual examination revealed a slight thickening in the area of the tumour, but no evidence of any extravesical spread. In July 1959, using a left paramedian incision, a transvesical segmental resection of the tumour was performed, combined with re-implantation of the right ureter into the bladder. The tumour measured 3 × 2·5 cm. and 1·5 cm. thick. It completely surrounded but did not obstruct the right ureter (Fig. 7). Convalescence was entirely uneventful. A section of

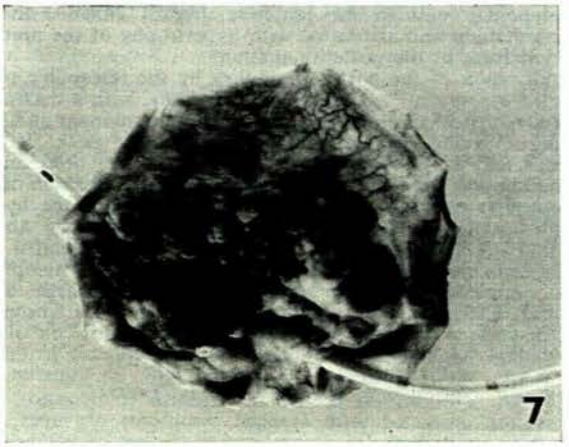

Fig. 7. Photograph of excised carcinoma of the bladder.

the tumour showed an anaplastic transitional-cell carcinoma of the bladder. The child is still alive and well 3¼ years after operation, and cystoscopies have not revealed any evidence of recurrence. An intravenous pyelogram taken after operation now shows excellent function in the right kidney, complete absence of hydro-ureter, and a normal cystogram.

FIBROMYOMA OF THE POSTERIOR URETHRA

Tumours of the posterior urethra in children causing urinary obstruction and retention are rare, but in this series 2 pedunculated fibromyomata arising from the verumontanum are recorded. Polypoidal tumours of the posterior urethra have been classified as (*a*) congenital, and (*b*) acquired.

The acquired type is more common and it is the sequel of bacterial infection, the causative factor being a chronic prostatitis. These polypi have a typical structure of fibrovascular tissue covered by transitional epithelium, sometimes containing glandular tissue and frequently showing infection in the stroma. As a complete contrast the congenital type is rare, and Stevens describes 2 cases occurring in children of 5½ and 3½ years causing obstructive symptoms in the lower urinary tract. During the act of micturition these pedunculated polypoidal tumours become inverted and firmly wedged in the membranous urethra, thereby occluding the lumen. The 2 cases are now described in detail since they present interesting symptoms and radiological appearances.

Pedunculated Fibromyoma (Andrew, Aged 3¼ Years)

This child was seen in August 1960 on account of difficult micturition which apparently occurred only in the morning on waking. At this time the act of micturition was intermittent, and only small quantities of urine were passed with some difficulty and pain. The parents had observed that the urinary flow would suddenly cease, to recommence after an interval of seconds, and once the bladder was emptied micturition returned to normal for the remainder of the day. Two attacks of retention had occurred, one 4 weeks before admission, which

required relief by catheterization, the other resolving spontaneously with expectant treatment. There was no obvious cause for these symptoms and it was considered that congenital posterior urethral valves may have been the aetiological factor. Intravenous pyelogram revealed a normal upper urinary tract, but a micturating cystogram showed a definite filling defect in the posterior urethra which was considered to be either a cyst of the posterior urethra of prostatic origin or an atypical formation of posterior urethral valves. Urethroscopy showed a polypoidal mass in the posterior urethra attached to the verumontanum and associated with hypertrophy of the urethral mucosal folds of the posterior urethra.

Exploration of the posterior urethra by the retropubic route revealed a pedunculated tumour 1·5 cm. long, with a maximum diameter of 0·75 cm., arising from the verumontanum and protruding into the bladder through the bladder neck. It was obvious that a ball-valve type of obstruction had been caused by the tumour, producing the symptomatology. Resection of the tumour was carried out with a wedge excision of the hypertrophied bladder neck, the bladder being drained by an indwelling urethral catheter. Convalescence was uneventful and he has remained fit and well and free of any urinary symptoms with completely normal micturition since discharge from hospital. The tumour on section was shown to be a fibromyoma, the surface of which was covered by squamous epithelium, this metaplasia had probably resulted from chronic irritation.

Polypoidal Fibromyoma (Philip, Aged 12 Years)

This boy presented with atypical symptoms of suprapubic pain and pain in both loins unrelated to the act of micturition. There was a vague history of haematuria on one occasion. A preliminary intravenous pyelogram showed a normal upper urinary tract, but a voiding cystourethrogram revealed a definite filling defect in the upper portion of the prostatic urethra; the appearances suggested a pedunculated polyp arising from the posterior urethra. Retropubic exploration of the prostatic urethra revealed a polypoidal tumour, 2 cm. long, arising from the verumontanum and extending upwards to and through the bladder neck. The tumour was resected together with a portion of the hypertrophied bladder neck, and an uneventful convalescence ensued. There was complete relief of symptoms and on discharge from hospital complete urinary control had returned. Microscopy showed that the polyp was a fibromyoma arising from the posterior urethra.

SARCOMA OF THE PROSTATE

Finally, two cases of sarcoma of the prostate are presented. Tumours of the prostate are notoriously highly malignant and extremely rare; the predominant tumour is the sarcoma though a few benign tumours, notably leiomyomata, have been described. Sarcoma of the prostate is a disease of childhood, but cases have been reported in early adult life, though it is rare to find the condition after the age of 18 years. A total of 209 cases have been recorded in the literature, and it is interesting to note that 50% of these tumours were encountered in the first decade. The tumours are mesoblastic in origin and vary markedly in histological structure. The characteristic feature of the tumour is that it has a rapid growth, often presenting as a large soft tumour mass which rapidly fills the pelvis, displacing the bladder and posterior urethra at an early stage in its development and thereby causing obstructive urinary symptoms. Cases have been reported in which the tumour had extended inferiorly to involve the perineum, buttock and rectum. Rhabdomyosarcoma of the prostate is also a rare tumour and it is considered to arise in the anterior commissure of the prostate where connective tissue and striated muscle fibres have been demonstrated. The remaining prostatic tumours that have been described in the literature are derived from the normal stroma of the prostate gland.

The presenting symptoms are predominantly urinary — frequency or difficulty of micturition, and acute or chronic retention being some of the more common manifestations. Cachexia, uraemia, anaemia, urosepsis as a result of an obstructive uropathy, are late manifestations. Pyrexia and painful frequency of micturition have also been recorded as presenting symptoms. The typical history combined with routine abdominal and rectal examination establishes the diagnosis without doubt. Occasionally a large tumour presenting as a palpable mass above the symphysis has been mistaken for a chronically distended bladder. Confusion in the past has occurred and rendered diagnosis difficult since the soft-tissue mass felt on rectal examination in the prostatic region had inadvertently been mistaken for a prostatic abscess. Excretion pyelography combined with cystourethrography establishes the diagnosis; some of the common findings are varying degrees of obstructive uropathy affecting the upper urinary tract, with gross deformity and displacement of the bladder and elongation and distortion of the prostatic urethra.

When first seen the majority of cases are so far advanced and inoperable that treatment is practically useless except as directed to the palliative relief of symptoms. Radical cystoprostatectomy has been performed in isolated cases, but without lasting success.

The first of the tumours to be described is a rhabdomyosarcoma of the prostate.

Rhabdomyosarcoma (Simon, Aged 7 Years)

This child was admitted with retention of urine with a most surprising history, as he was apparently fit and well until one month before admission. The presenting symptom then was pain in the upper part of the left thigh, as a result of which he had acquired a limp and walking was painful. Attacks of pyrexia with lower abdominal discomfort together with difficulty of micturition developed for the first time one week before admission, culminating in retention of urine. The child was ill and distressed, complaining of pain in the lower abdomen, and there was a very large, hard, tense mass in the lower abdomen with superimposed distended superficial veins of the abdominal wall. Intravenous pyelography showed bilateral hydronephrosis, and the cystogram film demonstrated gross displacement of the bladder with elongation and distortion of the posterior urethra. Laparotomy revealed a large inoperable tumour mass occupying the pelvis and extending 12 cm. above the symphysis, the tumour being adherent to the rectum, and the bladder was displaced in an upward anterior direction rendering any form of operative treatment impossible. A rapid deterioration in the general condition occurred, and he died 7 days after operation. Biopsy showed an undifferentiated sarcoma. At autopsy the clinical findings were confirmed, and a large pelvic tumour, 14 cm. long, 10 cm. in anteroposterior diameter, and 11 cm. wide, was demonstrated occupying the whole of the pelvis, displacing the urethra in a posterior direction and the bladder superiorly, causing bilateral ureteric obstruction. In addition there was thrombosis of the vena cava. Section of the autopsy material showed that the tumour was a rhabdomyosarcoma of prostatic origin.

Myxosarcoma (Peter, Aged 1½ Years)

Finally a myxosarcoma of the prostate is presented.

The presenting symptom in this child was difficulty of micturition which had occurred gradually 2 months before admission. At the time of examination the child had a large tumour of the prostate which obstructed and displaced the urethra, causing urinary retention. A biopsy proved it to be a myxosarcoma arising from the prostatic urethra. The tumour increased in size slowly, and finally ulcerated into the rectum and right buttock, death occurring 4 months after admission.

SUMMARY

An attempt has been made to describe and illustrate some of the more interesting and rare tumours which have been encountered in children, affecting both the upper and lower urinary tracts. The importance of a complete urological investigation in any child complaining of urinary tract symptoms cannot be overemphasized, and if success is to be achieved in the future, and a cure is to be afforded to these unfortunate children, then early diagnosis is essential and radical surgery should be performed.