HYPERNEPHROMA IN A CHILD OF FIVE YEARS

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This rarity, hitherto regarded as practically unknown, has cropped up here and there in the literature of late years.

The adult type of renal parenchymal carcinoma was regarded by Bell[1] as so rare as to cause doubt if it ever occurred at all. Nearly all renal growths occurring in the 1st decade are embryomas (Wilm's tumours), and growths of any sort occur very seldom in the 2nd and 3rd decades. Bell quotes Hellstrom[2] as publishing a case, but criticizes the absence of microscopic illustrations. Gross,[3] over a period of 30 years, found no hypernephromas in a series which included 96 embryomas. He says, 'The extreme rarity of hypernephroma in childhood makes it rather superfluous to consider the lesion in a differential diagnosis'.

Willis[4] in 1,060 necropsis for carcinoma found 27 cases of renal carcinoma, none of which were in children. His youngest cases were 29, 30 and 32 years old. The mean age at death was 58 years.

Priestley[5] in a series of 642 renal tumours found 502 cases of adenocarcinoma. Of these only one patient, aged 23 years, was below the age of 30. The mean age for adenocarcinoma of the parenchyma was found to be 52·8 years, and that for Wilm's tumour 6·5 years.

Riches, Griffiths and Thackray[6] in a magnificent study of 2,314 cases of renal and ureteric growths, found that 75% of these were cases of adenocarcinoma, of which 80% were in the 5th, 6th or 7th decade. The youngest was 11 years old, and the eldest 86. These authors refer to a case, mentioned by Roche,[7] of adenocarcinoma in a girl of 6, and add, 'The known variations in the histology of Wilm's tumour make it possible that this was the more correct diagnosis'.

Nicholls,[8] however, has reported a case of clear-celled papillary adenocarcinoma in a child of 22 months, and Hempstead *et al.*[9] report 2 more, stating that before their 2 cases only 4 cases had been registered in the American Tumour Registry; these 2 cases were aged 8 and 14 years.

Finally Beattie[10] reports a case in which he performed nephrectomy in April 1951. This patient was 7 years old when first detected, and was alive and well 2 years after the operation. Beattie regards the prognosis as encouraging. In the case to be recorded below the patient was well and apparently free from metastasis 3 years after the operation, but died of metastases 8 months later.

Hempstead and his associates attribute the rarity of these tumours in children to the fact that, as is believed, they arise in adenomata, whose development is slow and related to aging processes.

CASE REPORT

M.T., a Cape Coloured girl just under 6 years old, was admitted to Groote Schuur Hospital, Cape Town, in July 1948. A year before she had had a painless haematuria of 4 months' duration, and now had a large right renal mass.

Pyelography showed a normal left kidney and a non-functioning right kidney, whose position was occupied by a mass (Fig. 1).

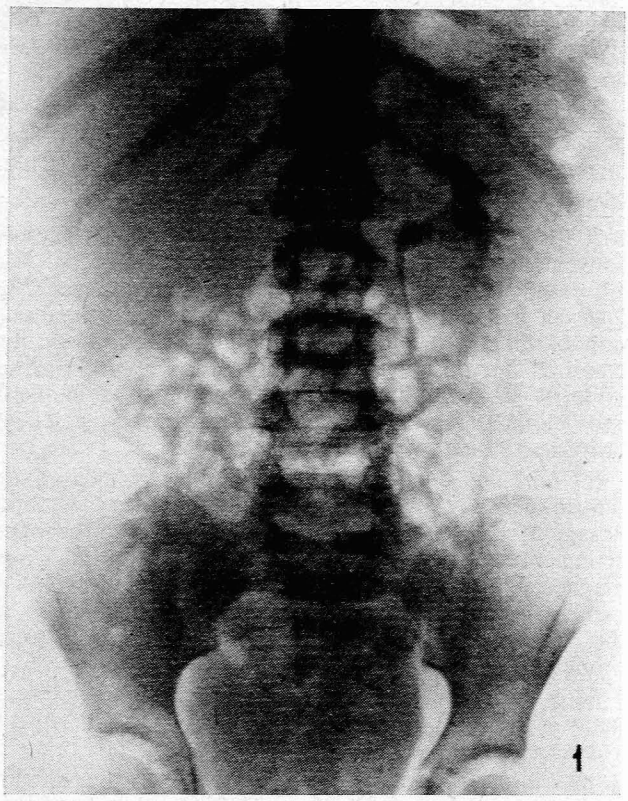

Fig. 1. Pyelogram.

The liver was palpable 2 fingers' breadth below the costal margin. Its edge was firm and smooth. At cystoscopy a normal bladder was seen and the right ureter was catheterized. A retrograde pyelogram (Fig. 2) revealed advanced distortion caused by the growth and extensive invasion of the ureter by polypoid extensions of the tumour, which caused numerous filling defects in the shadow of the upper portion of the ureter.

On 4 August 1948 a right lumbar nephrectomy was performed. The kidney was easily delivered. Growth was felt in the pelvis extending well down the ureter, which was divided well below the extension of the tumour at the lowest possible level. The kidney having been removed, the resultant cavity was explored for glands No enlarged glands were felt and the liver appeared to be normal.

The appearance of the tumour is shown in Fig. 3, which does not, however, illustrate the length of ureter removed, owing to the shrinkage caused by formalin.

The pathologist, Dr. G. Selzer, reported as follows: 'The cut surface shews a cellular tumour divided into lobules by dense bands of fibrous tissue. The tumour has extended into the pelvis and upper part of the ureter. The histology is that of a papillary carcinoma or so-called hypernephroma (Fig. 4). There are numerous small foci of calcification, and one focus of bone formation was encountered. The appearance is not that of a nephroblastoma'.

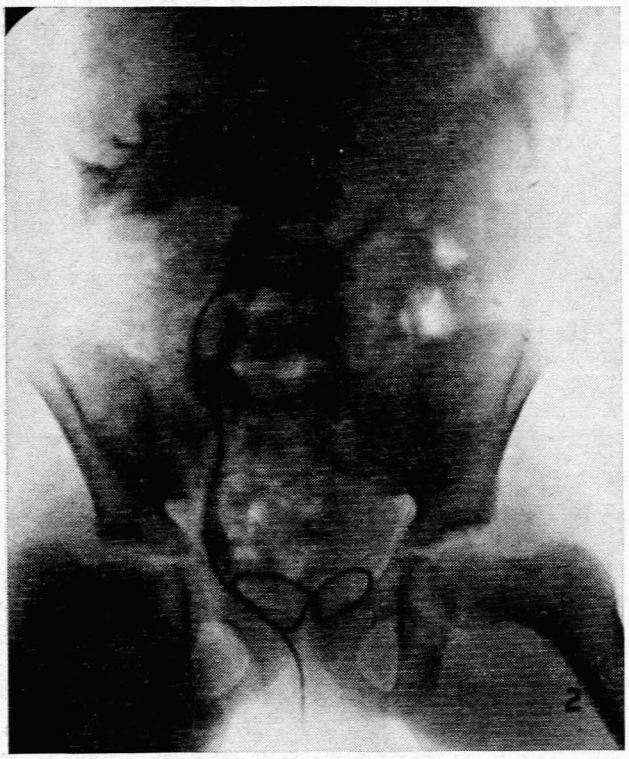

Fig. 2. Retrograde pyelogram.

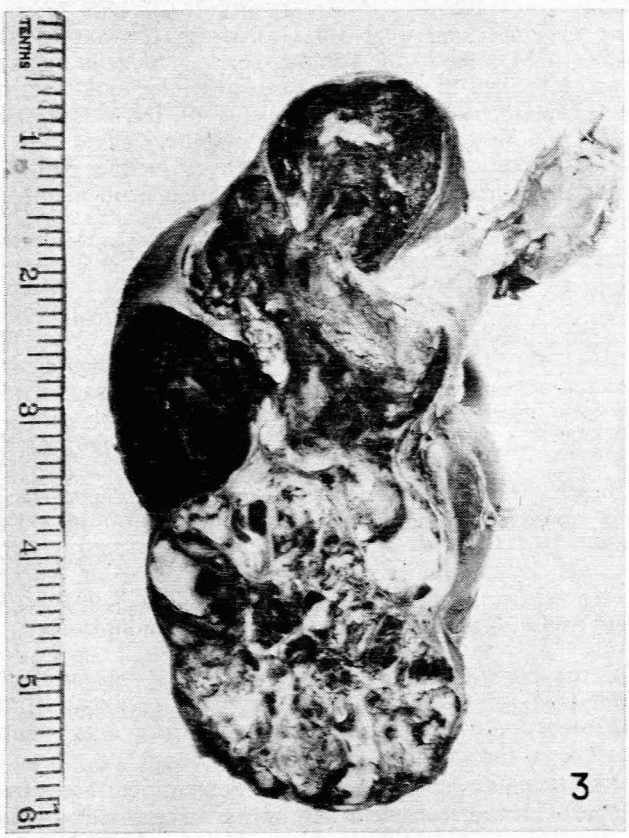

Fig. 3. The tumour after removal.

The patient made an uninterrupted recovery, and after a course of post-operative X-ray therapy returned home in good health. Apart from one or two minor setbacks, in one case due to measles, and in another to a bicycle accident, she remained well and lively for 3 years. It was then found that she was failing to gain weight, and in September 1951 a lump was detected in the left side of the upper abdomen. The left kidney was hydronephrotic and was displaced by the mass. The diagnosis of glandular metastases was made and further irradiation was given. Deterioration in the child's health continued and she died on 12 March 1952, 3 years and 7 months after her operation and over 4½ years after the onset of symptoms. Consent for a post-mortem examination was not obtained.

COMMENT

This case when first seen was diagnosed as a Wilm's tumour. The long survival after operation was so

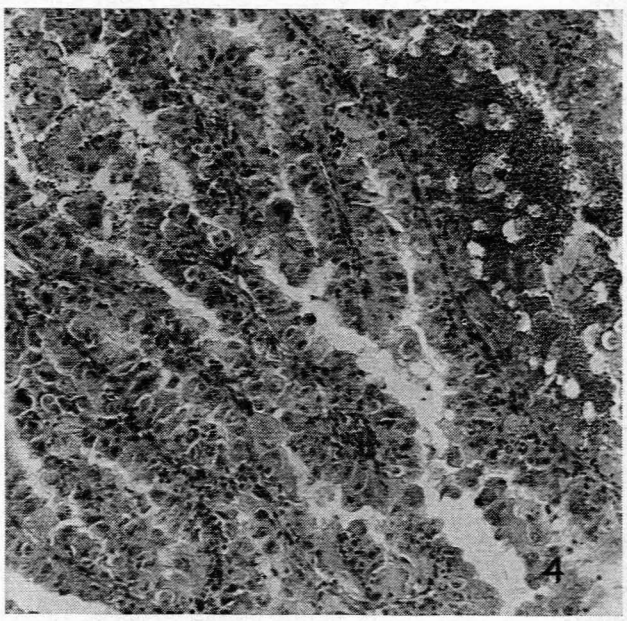

Fig. 4. Photomicrograph of tumour.

encouraging that for some time the patient was a show case, and she was nearly reported as a case of probable long-term survival.

The diagnosis of hypernephroma is based primarily on the histology, which is that of a clear-celled papillary adenocarcinoma. Foci of calcification are not uncommon in hypernephromata and it is probable that metaplasia in one of these was responsible for the one focus of bone formation encountered. The macroscopic appearance of the growth and the subsequent long survival in spite of the very advanced nature of the case are further points which support the diagnosis.

Nearly all authorities are clearly in favour of the abdominal approach for renal tumours in children. In this case it is felt that probably an abdominal nephrectomy would have been better. At the time it was assessed clinically as a tumour which would be particularly easily removed by a lumbar nephrectomy, and this assessment proved correct.

I am indebted to Mr. S. Scher, the Head of the Department of Urology, and to the Medical Superintendent of Groote Schuur

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