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Review

Primary renal tumours in the first year of life

A population based review

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Summary. Of 30 patients presenting with primary renal tumours in the first year of life, there were 23 Wilms' tumours (15 classical, six epithelial and two rhabdomyomatous), three rhabdoid neoplasms and four mesoblastic nephromas. Criteria for the diagnosis of rhabdoid tumours and mesoblastic nephromas are discussed with reference to histological difficulties. Although Wilms' tumour was the commonest neoplasm, mesoblastic nephroma predominated in the first three months of life. The clinical behaviour of the cases is reviewed, and rhabdoid tumours, although relatively few in number, accounted for a significant part of the overall mortality.

Key words: Renal tumours – Infancy histopathology – Classification – Behaviour

In the North West of England, 15.8% of Wilms' tumours occur in children under one year of age. Other primary renal neoplasms are also encountered in this age range.

The opportunity to study primary renal tumours in the first year of life, based on a comprehensive population-based collection, has been provided by the Manchester University Children's Tumour Registry (MCTR). From an assessment of this material, it is possible to obtain the relative incidence of various tumour types and to review histopathological criteria for individual diagnoses.

Materials and methods

Between 1954 and 1982, 162 primary renal tumours from children under 15 years of age have been included in the MCTR, and of these, 30 presented in the first year of life. Detailed

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records of all cases were available, with clinical histories, operation findings, gross descriptions of resected specimens and follow up. At least three haematoxylin and eosin stained sections from each tumour were available for retrospective study.

Results

The 30 cases were classified as shown in Table 1.

Table 1

Classical Wilms' Tumour	15
Epithelial Wilms' Tumour	6
Rhabdomyomatous Wilms' Tumour	2
Rhabdoid Tumour	3
Typical Mesoblastic Nephroma	1
Atypical Mesoblastic Nephroma	3
Total	30

Classical wilms' tumours. These consisted of metanephric blastema with varying degrees of epithelial and mesenchymal differentiation, although striated muscle was present only in limited amounts. Cases with 0, + and + + tubular status were included in this group (Lawler et al. 1975 and 1977).

Epithelial wilms' tumours. The greater part showed multilayered tubules with variable cyst formation (Chatten 1976). These neoplasms corresponded to the + + + tubular status group of Lawler et al. (1975 and 1977).

Rhabdomyomatous wilms' tumours. More than 50% was striated muscle; other mesenchymal components, including smooth muscle and fat, were also present (Wigger 1976).

Rhabdoid tumours. Only one of these three neoplasms had the typical rhabdoid appearance as described by Beckwith and Palmer (1978) (Fig. 1). The other two were made up of similar polygonal cells, but the diagnostic cytoplasmic features were not so clearly defined (Fig. 2).

Typical mesoblastic nephroma. This showed the characteristic histological picture of this neoplasm, with uniform spindle cells (Bolande et al. 1967; Bolande 1973) (Fig. 3).

Atypical mesoblastic nephromas. These were more cellular than the typical mesoblastic nephroma, with increased mitotic figures and nuclear atypia; polygonal-celled areas were present in addition to the spindle-cell component (Figs. 4 and 5). Pre-cartilage was noted in one case, and this tumour extended beyond the kidney margin into the adjacent fat (Fig. 6) and adrenal capsule (Fig. 7).

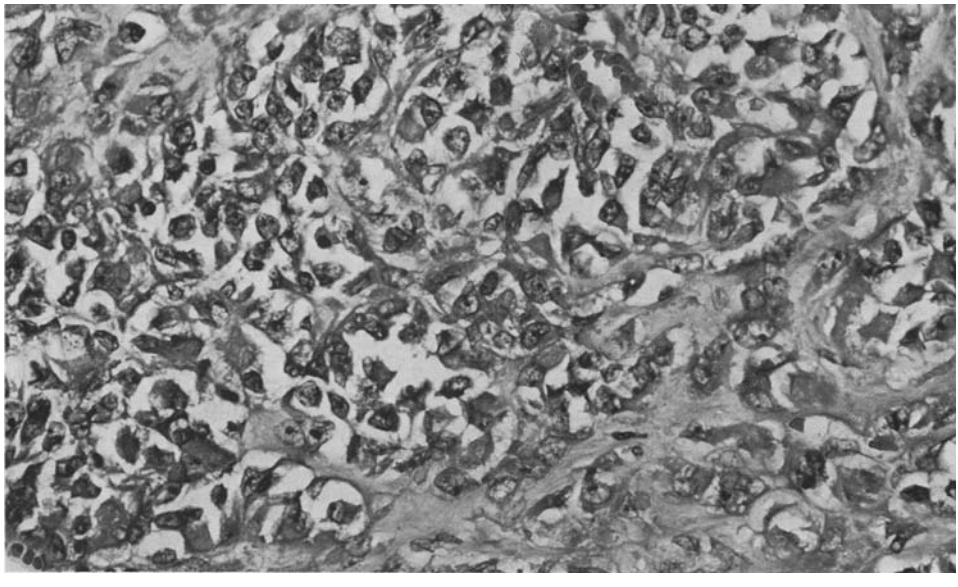

Fig. 1. Typical rhabdoid tumour (case 26) showing cells with abundant cytoplasm and occasional eosinophilic inclusions. Some sclerosis is present. H & E ×630

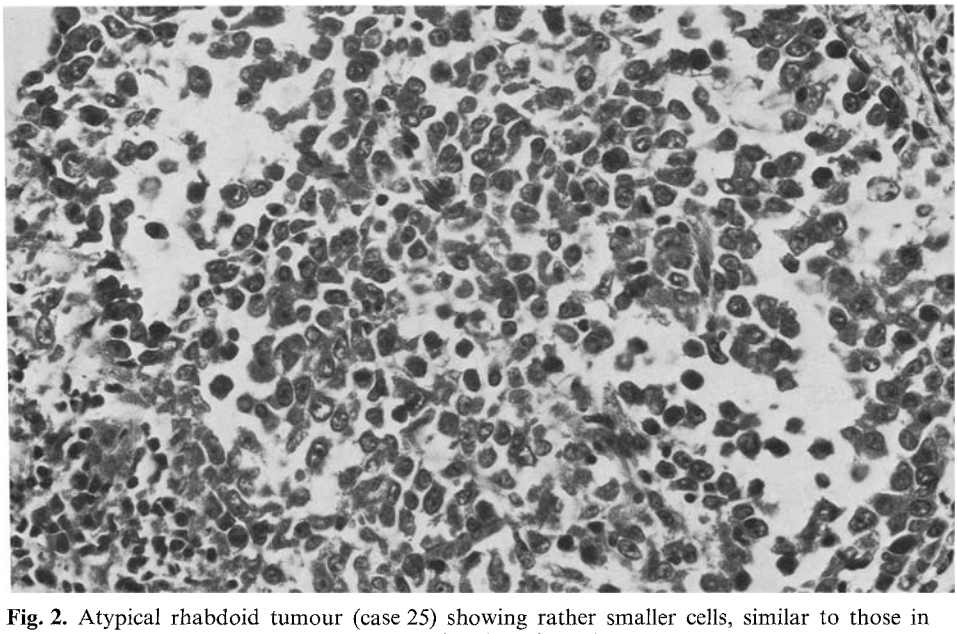

Fig. 2. Atypical rhabdoid tumour (case 25) showing rather smaller cells, similar to those in Fig. 1. but with less abundant cytoplasm and without inclusions. H & E ×630

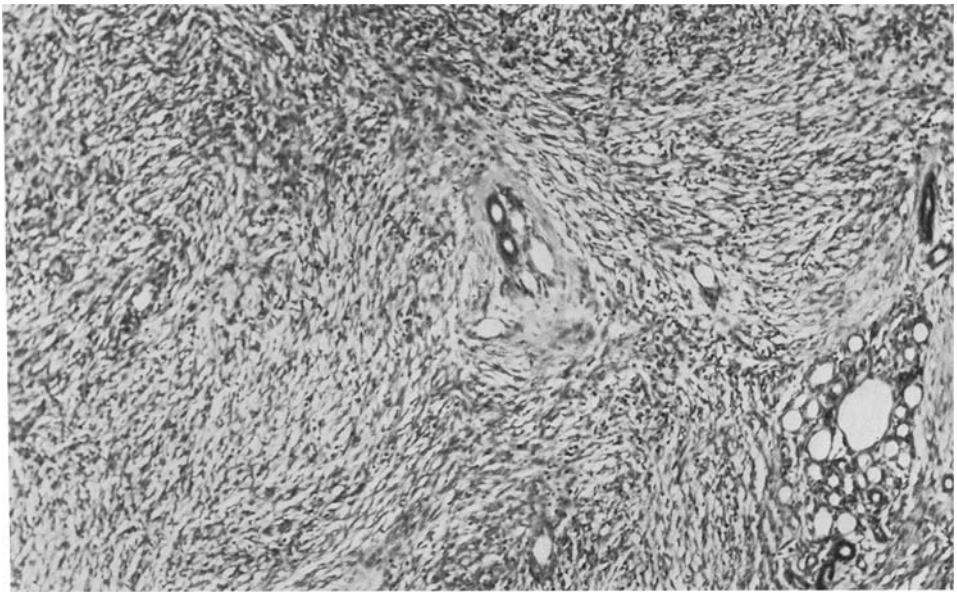

Fig. 3. Typical mesoblastic nephroma (case 27) with uniform spindle cells and embedded renal tubules. H & E ×150

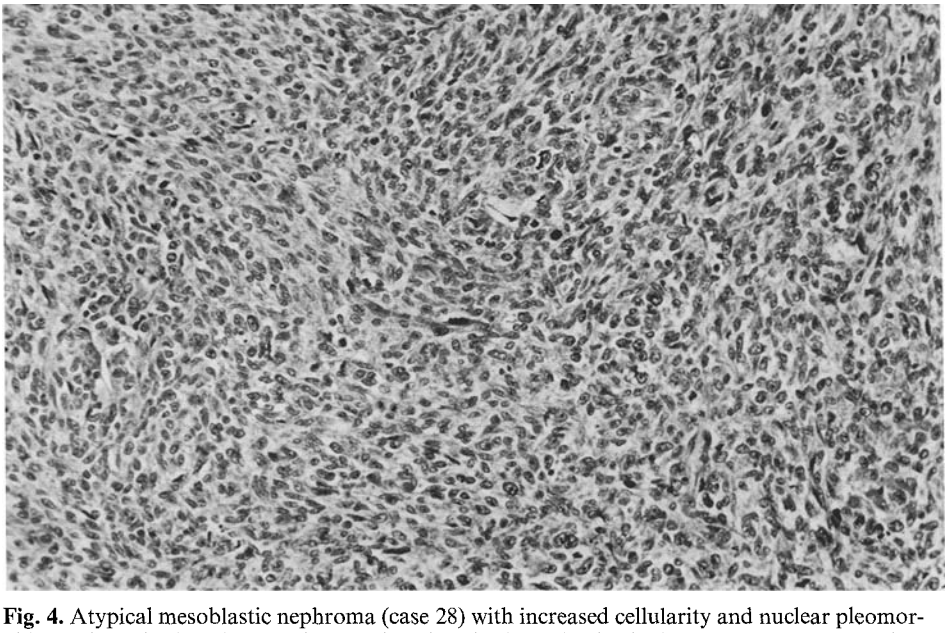

Fig. 4. Atypical mesoblastic nephroma (case 28) with increased cellularity and nuclear pleomorphism; the spindle cells are shorter than in Fig. 3, and mitotic figures are present. H & E ×350

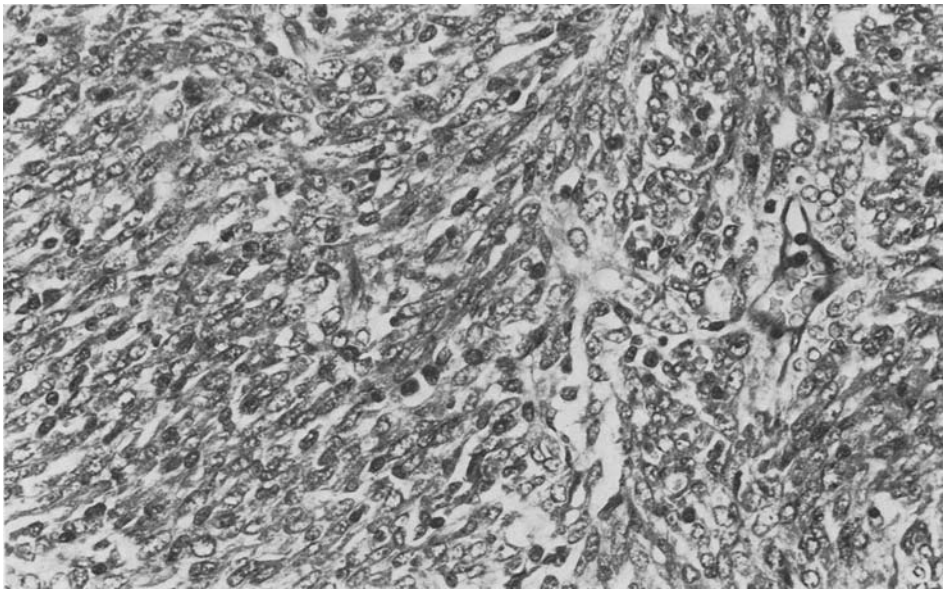

Fig. 5. Atypical mesoblastic nephroma (case 30) showing greater cellular detail than Fig. 4. H & E ×630

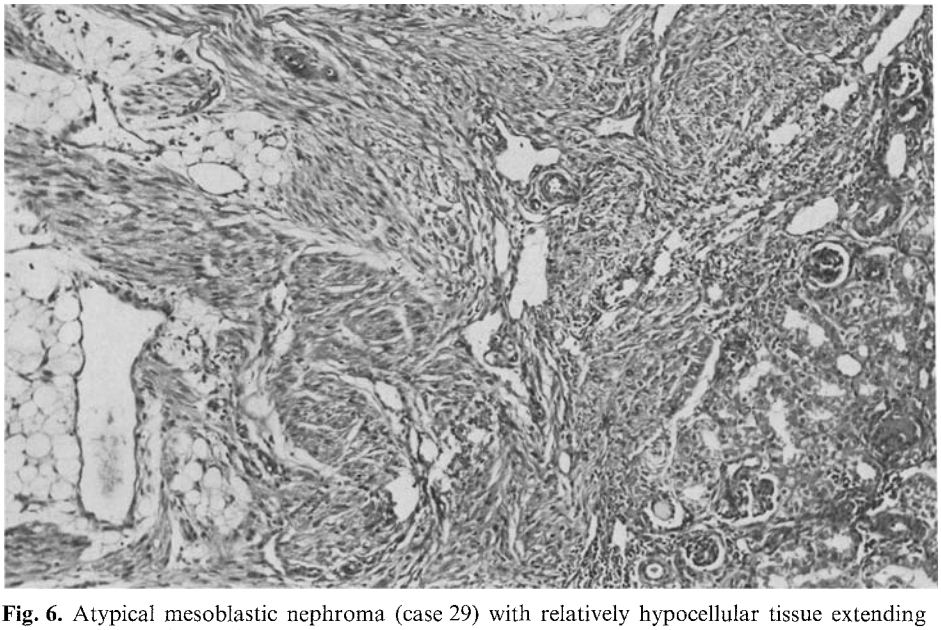

Fig. 6. Atypical mesoblastic nephroma (case 29) with relatively hypocellular tissue extending through the renal capsule into the perirenal fat. H & E ×150

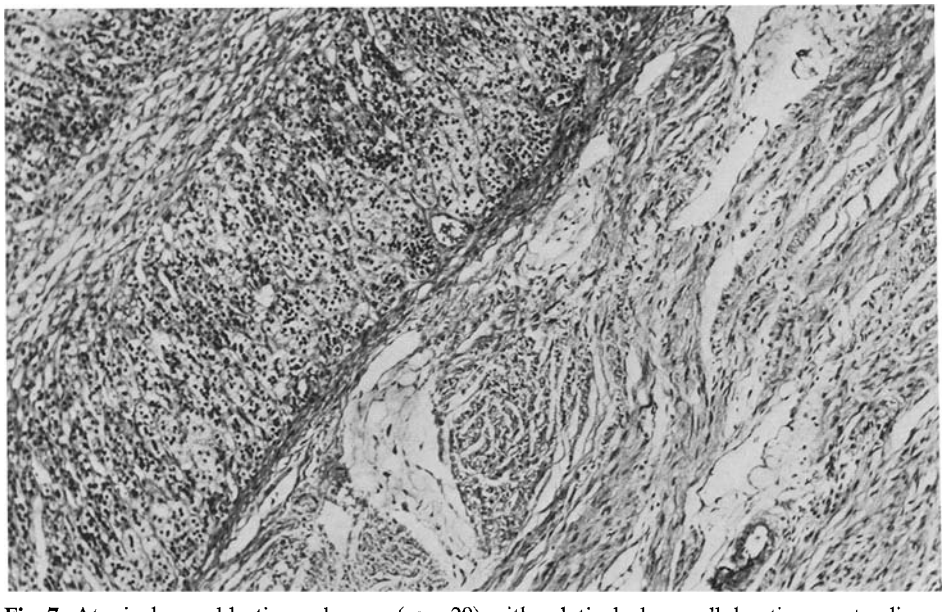

Fig. 7. Atypical mesoblastic nephroma (case 29) with relatively hypocellular tissue extending into the adrenal capsule. H & E ×150

Clinical data. Relevant clinical features of the 30 cases in Table 1 are summarized in Table 2.

Discussion

Most tumours in this series were seen in children aged six months or more, and relatively few were noted in early infancy. The only neonatal neoplasm was the typical mesoblastic nephroma, followed by a more cellular variant and two Wilms' tumours at the age of two months. A third mesoblastic nephroma was diagnosed at three months of age, supporting the view that this is the commonest primary renal neoplasm in early infancy (Bolande 1974).

Twenty three (77%) of the 30 cases in this series were classified as Wilms' tumours, the majority (65%) being typical, with 26% epithelial and 9% rhabdomyomatous. Both rhabdomyomatous neoplasms were bilateral, and Wigger (1976), from an assessment of the literature, regards the incidence of bilaterality to be 30%. The only other bilateral tumours in the present series were one classical and one epithelial Wilms' tumour. Microscopic cysts were recorded in three classical, one epithelial and both rhabdomyomatous nephroblastomas, but in none was cyst formation sufficiently prominent to justify a separate subgroup of cystic partially differentiated nephroblastoma (Joshi 1979). Nephroblastomatosis was seen in three epithelial Wilms' tumours; although it was not found in any other case, this may reflect inadequacy of available material, particularly sections of kidney adjacent to the tumour. Metastatic disease was seen in only five Wilms' tumours

Table 2. Clinical data of the 30 MCTR cases

Series No.	Name	C.T.R. No.	Age at Presentation (months)	Sex	Side	Outcome
Classical Wilms' Tumour						
1	D.P.	72/55	11	F	L	D after 9 months. Nodal and lung metastases
2	L.B.	82/58	4	M	R & L	D after 4 days; left ventricular hypertrophy, no metastases
3	A.B.	18/63	9	M	L	D after 1 week. Nodal and hepatic metastases
4	S.T.	118/64	11	M	R	A after 18 years
5	S.C.	77/65	2	M	L	A after 15 years
6	T.S.	58/72	11	F	L	A after 10 years. Lung metastasis after 8 months
7	P.W.	2/73	9	M	L	A after 9 years. Lung metastasis after 12 months
8	S.W.	102/74	11	F	R	A after 8 years
9	M.R.	39/75	4	M	R	A after 7 years
10	S.P.	65/75	8	F	L	A after 6 years
11	G.W.	3/76	9	F	L	A after 6 years
12	M.L.	31/78	7	M	L	A after 4 years
13	S.P.	24/80	10	F	R	A after 1 year
14	S.R.	3/82	6	F	L	A after 6 months. Three separate tumours
15	C.L.T.	13/82	8	F	R	A after 6 months. Nodal metastases
Epithelial Wilms' Tumour						
16	S.M.	9/58	2	M	R	A after 24 years
17	S.D.	3/71	7	M	L	A after 11 years
18	A.McN.	19/73	6	M	R	A after 9 years
19	C.V.E.	91/77	8	F	L	A after 5 years
20	J.R.	98/77	8	M	R & L	A after 5 years
21	N.J.	18/82	9	F	R	A after 6 months
Rhabdomyomatous Wilms' Tumour						
22	P.T.	49/57	9	F	L & R	D after 6 weeks. Bronchopneumonia; no metastases
23	P.L.	76/72	11	M	L & R	A after 10 years
Rhabdoid Tumour						
24	M.A	58/63	9	M	R	A after 19 years
25	D.W.	17/67	10	M	R	D after 1 day. Inoperable
26	A.K.B.	110/74	5	F	L	D after 5 weeks. Multiple bone metastases
Typical Mesoblastic Nephroma						
27	S.M.	2/64	0	M	L	D after 7 days. Septicaemia; no residual tumour
Atypical Mesoblastic Nephroma						
28	J.O.	34/61	3	F	L	A after 21 years
29	R.H.	66/70	6	F	L	A after 12 years
30	F.W.	45/71	2	M	R	A after 11 years

(all classical), and spread was almost exclusively to lymph node and lung. All epithelial Wilms' tumours survive, and only four patients from the other two Wilms' tumour subgroups died. One of the rhabdomyomatous neoplasms (case 22) was early in the series (1957), and death was from bronchopneumonia without residual tumour. It is noteworthy that all Wilms' tumours diagnosed after 1963, including two who developed lung metastases, have survived.

Only one of the three rhabdoid tumours (case 26) was regarded as typical, with abundant cytoplasm showing eosinophilic inclusions (Haas et al. 1981) (Fig. 1). The other two were made up of cells having a similar appearance but with less abundant cytoplasm and without inclusions (Fig. 2). None of these rhabdoid tumours showed features to support a diagnosis of Wilms' tumour (namely blastema, metanephric tissue, epithelial or mesenchymal differentiation). The proportion of typical diagnostic cells in rhabdoid tumours is variable, and may be limited to occasional focal areas. In this context, it is relevant that the typical rhabdoid tumour in the present series was the only one of the group where the complete operation specimen was available for retrospective study, and it is possible that further sections from the other two neoplasms would have revealed typical rhabdoid areas. Multiple bone metastases were recorded from the typical rhabdoid tumour, but no example of the Bone Metastasising Renal Tumour of Childhood (BMRTC) (Marsden and Lawler 1980) was encountered. Delicate polygonal cells are an important but variable feature of the BMRTC, and these neoplasms have been called the clear celled sarcomatous variant of Wilms' tumour (Beckwith and Palmer 1978). Ugarte et al. (1981) reported finding such a neoplasm in a neonate, although in the present authors' series of 46 cases, all have been over one year of age at presentation.

The only congenital tumour was a typical mesoblastic nephroma (Fig. 3). The other mesenchymal neoplasms showed more varied features and were classified as atypical mesoblastic nephromas (Figs. 4–7), possibly fitting into the intermediate group of Gonzalez-Crussi et al. (1981) or the cellular variant of Snyder et al. (1981). Relatively hypocellular areas may be encountered in atypical mesenchymal tumours, and the only neoplasm in this group which extended beyond the renal capsule (case 29) showed such areas in the perinephric fat (Figs. 6 and 7).

In conclusion, a population-based study has shown that Wilms' tumour is the commonest primary renal neoplasm in the first year of life, although mesenchymal tumours predominate under four months of age. Rhabdoid tumours comprise only 10% of the series, but account for an appreciable part of the overall mortality. Deaths from true nephroblastoma were mainly in the early years of the study, and all Wilms' tumours presenting since 1964 have survived.

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