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>
<thead>
<tr>
<th>Class</th>
<th>Count</th>
</tr>
</thead>
<tbody>
<tr>
<td>Classical Wilms’ Tumour</td>
<td>15</td>
</tr>
<tr>
<td>Epithelial Wilms’ Tumour</td>
<td>6</td>
</tr>
<tr>
<td>Rhabdomyomatous Wilms’ Tumour</td>
<td>2</td>
</tr>
<tr>
<td>Rhabdoid Tumour</td>
<td>3</td>
</tr>
<tr>
<td>Typical Mesoblastic Nephroma</td>
<td>1</td>
</tr>
<tr>
<td>Atypical Mesoblastic Nephroma</td>
<td>3</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td><strong>30</strong></td>
</tr>
</tbody>
</table>

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).