Correspondence

Mature renal teratoma and a synchronous malignant neuroepithelial tumour of the ipsilateral adrenal gland

The main primary childhood renal neoplasms are nephroblastoma, mesoblastic nephroma, clear cell sarcoma, and rhabdoid tumour. Other primary renal neoplasms include primitive neuroectodermal tumour (PNET), renal cell carcinoma, and angiomyolipoma. Nephroblastoma is the most common renal tumour in children. It is a complex embryonal tumour of metanephric blastemal derivation, which often contains diverse epithelial and stromal tissues. Diagnostic problems are often encountered when tumours contain a variety of heterologous elements. The term teratoid nephroblastoma has been used to describe a variant of nephroblastoma with a predominance of heterologous tissues. It is this variant that can be confused with a teratoma. Renal teratomas are rare and most have been dismissed as cases of teratoid nephroblastomas or retroperitoneal teratomas secondarily invading the kidney. The differentiation between these two neoplasms in the kidney is often problematic.

Neurogenic tissues in the kidney can be found in primary tumours or as part of metastatic tumours. The primary tumours are nephroblastoma, which may contain ganglion cells, neuroblast, and neuroglial tissue, and PNET. Adrenal and renal neuroblastomas can directly invade the adjacent kidney. We describe the pathology of a right renal mass in a 3 year old child and discuss the differential diagnosis.

A 3 year old girl presented with abdominal pain and diarrhoea. On examination she was found to have signs of pulmonary tuberculosis and was started on antituberculous treatment. Subsequently, a large, firm, tender, right flank mass clearly separate from the kidney was detected and she was referred to the Regional Paediatric Surgical Unit for further investigation and management.

On admission, the child was apyrexial, enaciated, and weighed 13 kg. She had bilateral coarse cracks and a wheeze. The abdomen was distended and a non-tender 3 cm hepatomegaly was palpated. Furthermore, a 10 × 12 cm non-tender, firm, non-pulsatile right flank mass was detected.

Results of routine laboratory tests were as follows: haemoglobin, 90 g/litre (normal, 112–143); white blood cell count, 8.2 × 10⁹/litre (normal, 5.5–15.5); and platelet count, 224 × 10⁹/litre. Urinary catecholamine values were as follows: noradrenaline, 0.279 µM/mM creatinine (CRT) (normal, 0–0.08); adrenaline, 0.023 µM/mM CRT (normal, 0–0.035); dopamine, 0.67 µM/mM CRT (normal, 0.1–1.3); vanillylmandelic acid 9 µM/mM CRT (normal, 0–1.5); and homovanillic acid, 11 µM/mM CRT (normal, 0–1.5). Renal and liver function tests were normal.

Computed tomography (CT) scan of the abdomen revealed a large tumour involving the right side of the abdomen. There were also multiple hepatic lesions consistent with metastases. A fine needle aspiration biopsy of the mass was performed. After a cytological diagnosis of neuroblastoma the child was started on the appropriate chemotherapy protocol consisting of vincristine, actinomycin, cyclophosphamide, and Adriamycin. She suffered seizures while on chemotherapy, which was subsequently decreased to 75% dosage. A CT scan of the brain was normal.

The child died two weeks after commencement of chemotherapy. A necropsy was performed.

Necropsy revealed an enlarged right kidney measuring 16 × 10 × 8 cm and weighing 1200 g. There was a well demarcated tumour mass in the upper pole of the kidney, which measured 7 × 6.5 × 5 cm (fig 1). The upper pole mass was encapsulated (fig 1, arrows), predominantly firm in consistency, and had yellow and white areas. There was a small area of soft, friable tumour present within the mass, close to the junction of the adjacent kidney. On the capsular surface the upper pole mass was clearly demarcated from the remaining kidney. It was not possible to recognise normal renal tissue because the remaining kidney was diffusely swollen, pale, soft and contained focal areas of necrosis (fig 1). The right adrenal gland was not identified despite serial sectioning. There was extension of the tumour into the right renal vein and inferior vena cava. Tumour spread into the ureter. A CT scan of the chest was also performed. There were multiple, greatly enlarged para-aortic lymph nodes containing metastatic tumour. Metastatic tumour deposits were present in the liver, vertebrae, and both lungs. In addition, caseous nodules were identified in the lungs and hilar lymph nodes. The left kidney and adrenal gland were normal. The rest of the postmortem examination was normal.

Histological examination of the well demarcated upper pole mass showed a tumour composed of multiple heterologous tissue elements. Tissues derived from all three germ layers—ectoderm, mesoderm, and endoderm—were present. The yellow areas corresponded to mature adipose tissue microscopically. In addition, striated muscle, smooth muscle, and fibrous tissue were present, as were small islands of neuroglial tissue. The epithelial component was variable and consisted of small tubular structures lined by cuboidal epithelial cells with clear cytoplasm. Also present were larger cystic structures lined by respiratory epithelium. Adjacent to these cysts were small closely packed acini composed of mucin containing epithelium. A prominent feature was the presence of several cysts lined by keratinising stratified squamous epithelium with hair follicles and hair shafts (fig 2). Small round tumour cells diffusely infiltrated the remaining kidney. These were arranged predominantly in solid sheets, but focal neuroectodermal canals and rosettes were also identified (fig 3). In addition, there was microscopic evidence of metastases in the lungs, liver, ovaries, and vertebrae. The metastatic tumour in all sites consisted of the primitive small, round cell component. Sections taken from the region of the right adrenal gland showed a diffuse infiltrate of small round blue cells, but no residual adrenal gland tissue was identified. There were no nephrogenic rests in the kidney.

A panel of immunohistochemical stains was performed on the small cell malignant tumour to detect cytokeratins (AE1/AE3, CAM5.2), synaptophysin, neuron specific enolase (NSE), chromogranin, WT1, desmin, muscle specific actin, S100 protein, glial fibrillary acidic protein, O13, leucocyte common antigen, and epithelial membrane antigen. The small round cells showed immunoreactivity for NSE, synaptophysin, and chromogranin. The cells were non-reactive for the remaining markers.

Histology confirmed tuberculosis of the lungs and lymph nodes. Acid fast bacilli were identified.

To diagnose a renal teratoma, the primary tumour should be unequivocally of renal origin and the tumour should exhibit unequivocal heterotopic organogenesis clearly recognised as evidence of attempts to form organs other than the kidney. It is the second criterion that often presents a problem. The question is: what constitutes unequivocal organogenesis? The presence of bone, cartilage, muscle, fat, neuroglial tissue, and mature epithelium cannot on their own be regarded as...
light, as is evident from the fact that it is difficult to differentiate from PNET. The immunohistochemical profile and cytogenetics also suggest the presence of a well-differentiated PNET component. Therefore, PNET is the most likely component, especially when considering the presence of hair shafts. The presence of hair shafts is not unexpected in a PNET, as it is a neuroectodermal tumour. The presence of hair shafts is also consistent with the histological findings, which show the presence of mature and immature tissues. The presence of immature tissues in a PNET is not uncommon, as it is a neuroectodermal tumour.
the cost effectiveness of a practice for which there are no current grade A recommendations.

The important issue that Dr Murphy raises relates to why thrombophilia tests are performed; are they to explain why an individual develops thrombosis or are they to optimise clinical decisions? In other words, are they for the sake of science or medicine. If testing is performed to explain why an individual develops thrombosis then the discriminate approach suggested by the 1990 guidelines quoted by Dr Murphy will increase the proportion of tested patients who are found to have a laboratory abnormality. However, if testing is performed to optimise clinical decisions then there is no rational basis for such a recommendation given the data that are currently available. There is no evidence that the predictive value of thrombophilia testing is in any way superior in the clinical decisions then there is no rational basis to do them.

The importance of the thrombophilia tests is whether their results influence his clinical decisions. Most textbooks are reference works where one can look up a specific problem. This book is also a guide. The problem with splenic pathology is where to look, what to look for. The first two chapters (“Introduction” and “Normal structure, development and functions of the spleen”) serve as a firm basis on which the approach to the study of splenectomy specimens is built. The last chapter (“Summary: some key points in splenic differential diagnosis”) provides an easy approach for some common situations. These three chapters include about 50 pages of text and illustrations and the evening it will cost studying them is a worthwhile investment. The other chapters deal with the disorders expected in the spleen, such as haematopoietic and infectious diseases, but there is also a chapter on post-traumatic and incidentally removed spleens, illustrating the practical approach of the authors.

Are there no complaints? Of course there are a few points that can be made. Not all of the illustrations are perfect; for instance, splenic marginal zone lymphoma, a difficult to recognise lymphoma is not very well illustrated, especially the important low power figure, which is not very informative. The references are well chosen, run into 1998, but again are erratic; in some areas, such as alterations in glomerular filtration, it tabulates large amounts of raw data, which omitting other important pieces of recent work. As a result, it is unlikely to satisfy researchers in this field.

Although nephrologists might find some useful information in this book, pathologists will find it of limited value.

I ROBERTS

The Principles of Clinical Cytogenetics.

Gersen SL, Keagle MB, eds. ($79.50.) Humana Press, 1999. ISBN 0 89603 414 0.

This book is introduced as being “suitable for cytogenetic technologists and clinicians alike”, and at first glance, appears to provide a comprehensive overview of all aspects of clinical cytogenetics. However, on closer examination, the book is a rather curious mixture of information. The introductory chapters are perhaps the most confusing. The overview of the biochemistry of genetics is brief, to the point that after a rather cursory knowledge of the processes of replication, transcription, and translation to understand the chapter, thereby rendering it largely surplus to requirements. The chapter on chromosome nomenclature, specifically the truncation of the Paris ISCN and because the authors are only too happy to refer the reader to suitable manuals for laboratory methodology, it would seem appropriate to do so for this topic also. There is a general lack of cohesion about the book, almost as though none of the authors had any knowledge of the contents of the other chapters. That said, several of the chapters on clinical cytogenetics provided very good overviews, and several included comprehensive tables of data pooled from various sources to give an easily manageable reference for aetiologies of several classes of chromosome disorder. The chapter on cancer genetics, although thorough in its presentation of tumour specific breakpoints, fails to expand on the specific biochemical and genetic results of the rearrangements specified. I felt that this was particularly remiss in the present climate of molecular genetic discovery, because only a few well selected and well described exceptions would be necessary to wet the reader’s appetite for further discovery. The reader should also be aware that the book suffers from shoddy proofreading. Diagrams are A VARGAS

Illustrated Pathology of the Spleen.


Illustrated Pathology of the Spleen by Bridget Wilkins and Dennis Wright is a beautiful book. It is a pleasure to hold it and browse through it because of its size, layout, and the quality of the print and figures. But, that is only the exterior. At least equally important is the contents. These match the style.

The authors aim with this book to demystify the spleen, putting forward a systematic, analytical approach to the interpretation of splenic pathology. The authors succeed by using numerous illustrations and the book is indeed a combination of an atlas and a textbook.

The book is well written and very readable. It relates to why thrombophilia tests are performed in any way superior in the clinical decisions then there is no rational basis for such a recommendation given the data that are currently available. There is no evidence that the predictive value of thrombophilia testing is in any way superior in the clinical decisions then there is no rational basis to do them.

The preface of the book should be read by all those who occasionally is confronted with splenic pathology. The authors succeed by providing a comprehensive overview of all aspects of splenic pathology, pathogenesis, and mechanisms of progression. The first of these is a brief eight pages. The introduction to light microscopy (LM) strangely includes a half page summary of a study that reported the absence of DNA polymerase-α positive cells in IgA nephropathy. Hoping that the next section, on LM classification of IgA nephropathy, would make sense of what is currently a confused area, I was disappointed to find that only three systems were discussed, all from Japan (Shirai’s, Sakaguchi’s, and Sakai’s), with no attempt to rationalise disparate schema or to differentiate disease activity from chronicity. The chapters on pathogenesis improve on this poor start, but again are erratic; in some areas, such as alterations in glomerular filtration, it tabulates large amounts of raw data, which omitting other important pieces of recent work. As a result, it is unlikely to satisfy researchers in this field.

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Book reviews


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mislabeled, and typographical errors are fre-
quent and often misleading— in the chapter
on chromosome nomenclature several exam-
ples are given using a chromosome band that
does not exist. Overall, I found the book to be
a useful aide memoire for several topics, and
although perhaps an expensive luxury for the
individual, I would recommend the book as an
addition to any laboratory or hospital library.

B HOLLAND

1 Mitelman F, ed. ISCN 1995. An international
system for human cytogenetic nomenclature. Basel:

Rotaviruses, Methods and Protocols.
Methods in Molecular Medicine. Gray J,
Desselberger U, eds. (US$89.50.) Humana

In the UK between January 1989 and
December 1999 there were 164 279 reports
to the Public Health Laboratory Service,
Disease Surveillance Centre of cases of gastroenteritis
caused by rotaviruses—an average of 14 934
each year. Rotaviruses were discovered in
animals in the early 1960s and in humans in
the early 1970s. For several years, electron
microscopy was the only widely available
method used for diagnosing infection. Since
the early 1980s, molecular, serological, and
Cell culture methods have come into use and fruitful
research on these important patho-
gens has resulted. This excellent new book
provides detailed protocols for these meth-
ods.

The central chapters begin with an up to
date review of the relevant field, all of which
are clearly written, informative, and excel-
lently referenced. The editors have provided a
short, but informative, introductory chapter
aptly titled “Basic facts”. In a chapter
contributed by BV Venkataram Prasad and
Mary Estes, on electron cryomicroscopy
and computer image processing techniques, the
structure–function studies of rotaviruses are
beautifully illustrated with computer generated
dimensional reconstructions of
rotavirus particles. A chapter by Mary
Ramsay and David Brown describes the
epidemiology of rotavirus infections and
concentrates on surveilance and the, surprisingly
high, disease burden caused by rotaviruses.
The other chapters, all of which are of the
same high standard, include rotavirus replica-
tion, cell entry, genetics, immunology, animal
models, serotyping, and genotyping.

G BEARDS

CD-ROM review

Stomatology-ENT. Brocheriou C,
Baglin AC, Wassef M, et al. (US$149.00.)
This CD ROM of stomatology-ENT is a
reference image database of the field of ENT.
It is in English and in French. It contains
macroscopic and histopathological pictures
of the main lesions in the field of oral
pathology and ear, nose, and throat pathol-
ogy. Clinical, radiological, cytological,
immunohistochemical, or ultrastructural pic-
tures of some of these lesions are present.
Indeed, this CD ROM contains a large
number of pictures of most lesions in this
area. The images of interest are very easy to
find using a list or keyword driven search.
The quality of the photographs is variable;
most of them are sharp, but others are not
always focused or are very dark. The data
sheet contains all relevant information about
the picture but this information is often
scanty. However, additional information
about different lesions is supplied by separate
text slides.

There are three fields containing a selec-
tion, image, and database window. It is a nui-
sance that the database window stays on the
screen when another program (such as Word
or PowerPoint) is used. The program is easy to
use and it is very easy to copy the pictures to
PowerPoint slides for use in presentations.
In addition, they can be copied to a photo
editor (such as Photoshop) so that the
pictures can be edited.

In conclusion, this CD ROM contains a
wealth of photographic material, which can
be used for diagnostic or educational pur-
poses. The layout, however, could be
improved.

J A KUMMER

Calendar of events

Full details of events to be included should be
sent to Maggie Butler, Technical Editor
JCP; The Cedars, 36 Queen Street, Castle
Hedingham, Essex CO9 3HA, UK;
email: maggiebutler@pilotree.prestel.co.uk

BSCC Northern Spring Tutorial:
Gynaecological Cytology
8 March 2001, Manchester, UK
Further details: BSCC Office, PO Box 352,
Uxbridge UB10 9TX, UK. (Tel +44 01895
274 020; fax +44 01895 274 080; email
lesley.couch@psilink.co.uk)

Urological Surgical Pathology for the
Practising Pathologist
24–26 March 2001, Sanibel Harbour Resort
and Spa, Fort Myers, Florida, USA
Further details: Department of Continuing
Education, Harvard Medical School, 25
Shattuck Street, Boston, MA 02115, USA.
(Tel +1 617 432 1525; fax +1 617 432 1562;
email hms-cme@hms.harvard.edu)

Haematological Morphology and
Leukaemia Classification for
Cytogeneticists
29 March 2001, St Mary’s Hospital, London,
UK
Further details: The Academic Secretary,
Department of Haematology, St Mary’s Hos-
pital Campus of ICSM, Norfolk Place,
London W2 1PG, UK. (Fax +44 020 7262
5418)

6th European Forum on Quality
Improvement in Health Care
29–31 March 2001, Bologna, Italy
Further details: BMA/BMJ Conference Unit,
BMA House, Tavistock Square, London
WC1H 9JR, UK. (Tel +44 020 7383 6409;
fax +44 020 7383 6869; email
Quality@bma.org.uk; website www.quality.b-
mjp.com)

Diagnostic Histopathology of Breast
Disease
23–27 April 2001, Hammersmith Hospital
(Imperial School of Medicine), London, UK
Further details: Wolfson Conference Centre,
Hammersmith Hospital, Du Cane Road,
London W12 ONN, UK. (Tel +44 020 8383
3117/3227/3245; fax +44 020 8383 2428;
email wccc@ic.ac.uk)

Gynecologic and Obstetric Pathology
26–29 April 2001, Fairmont Copley Plaza,
Boston, Massachusetts, USA
Further details: Department of Continuing
Education, Harvard Medical School, 25
Shattuck Street, Boston, MA 02115, USA.
(Tel +1 617 432 1525; fax +1 617 432 1562;
email hms-cme@hms.harvard.edu)

BSCC London Spring Tutorial: Lung
and Pleural Cavity Fluid Cytology
27 April 2001, Guy’s Hospital, London,
UK
Further details: BSCC Office, PO Box 352,
Uxbridge UB10 9TX, UK. (Tel +44 01895
274 020; fax +44 01895 274 080; email
lesley.couch@psilink.co.uk)

International Consultation on the
Diagnosis of Noninvasive Urothelial
Neoplasms
11–12 May 2001, University of Ancona
School of Medicine, Torrette, Ancona, Italy
Further details: R Montironi, Ancona Italy
(email r.montironi@popsci.unian.it),
DG Bostwick, Richmond, VA, USA (email
bostwick@bostwicklaboratories.com),
P-F Bassi, Padua, Italy (email bassif@u01.unipd.it),
M Droller, New York, USA
(email michael_droller@smtlink.mssm.edu),
or D Waters, Seattle, WA, USA
(email waters@vet.vet.purdue.edu)

BSCC Annual Scientific Meeting
9–11 September 2001, Majestic Hotel, Har-
rogate, UK
Further details: BSCC Office, PO Box 352,
Uxbridge UB10 9TX, UK. (Tel +44 01895
274020; fax +44 01895 274080; email
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