

Eberhard Löhr Lutz-Dietrich Leder (Eds.)

Renal and Adrenal Tumors

Pathology, Radiology, Ultrasonography,
Magnetic Resonance (MRI), Therapy, Immunology

With Contributions by

R. Ackermann, K.-D. Bachmann, H. Behrendt, P. E. Billimoria,
H. Chr. Dominick, M. D. Gross, R. Hartung, W. Havers,
R. Heckemann, J. V. Kaude, R. E. Kinard, E. K. Lang, L.-D. Leder,
E. Löhr, A. A. Moss, R.-D. Müller, H. J. Richter, E. Scherer,
M. Serdarevic, B. Shapiro, W. P. Shuman, J. L. Williams, C. Wirtz

Second, completely revised Edition



Springer-Verlag

Eberhard Löhr · Lutz-Dietrich Leder (Eds.)

Renal and Adrenal Tumors

Pathology, Radiology, Ultrasonography,
Magnetic Resonance (MRI), Therapy, Immunology

With Contributions by

R. Ackermann, K.-D. Bachmann, H. Behrendt, P.E. Billimoria,
H.Chr. Dominick, M.D. Gross, R. Hartung, W. Havers, R. Heckemann,
J.V. Kaude, R.E. Kinard, E.K. Lang, L.-D. Leder, E. Löhr, A.A. Moss,
R.-D. Müller, H.J. Richter, E. Scherer, M. Serdarevic, B. Shapiro,
W.P. Shuman, J.L. Williams, C. Wirtz

Second, completely revised Edition

With 229 Figures (17 in Colour) in 406 Separate Illustrations

Springer-Verlag Berlin Heidelberg New York
London Paris Tokyo

Professor Dr. EBERHARD LÖHR
Röntgendiagnostisches Zentralinstitut
Klinikum der Universität Essen
Hufelandstraße 55
D-4300 Essen

Professor Dr. LUTZ-DIETRICH LEDER
Institut für Pathologie
Klinikum der Universität Essen
Hufelandstraße 55
D-4300 Essen

ISBN 3-540-09192-0 1. Auflage Springer-Verlag Berlin Heidelberg New York
ISBN 0-387-09192-0 1st Edition Springer-Verlag New York Heidelberg Berlin

ISBN 3-540-16554-1 Springer-Verlag Berlin Heidelberg New York
ISBN 0-387-16554-1 Springer-Verlag New York Berlin Heidelberg

Library of Congress Cataloging in Publication Data

Renal and adrenal tumors. Bibliography: p. Includes index. 1. Kidneys – Tumors. 2. Adrenal glands – Tumors. I. Ackermann, R. (Rolf) . II. Löhr, E. (Eberhard) . III. Leder, Lutz-Dietrich. [DNLM: 1. Adrenal Cortex Neoplasms. 2. Kidney Neoplasms. WJ 358 R393] RC280.K5R44 1987 616.99'261 86-31555

ISBN 0-387-16554-1 (U.S.)

This work is subject to copyright. All rights are reserved, whether the whole or part of the material is concerned, specifically the rights of translation, reprinting, re-use of illustrations, recitation, broadcasting, reproduction on microfilms or in other ways, and storage in data banks. Duplication of this publication or parts thereof is only permitted under the provisions of the German Copyright Law of September 9, 1965, in its version of June 24, 1985, and a copyright fee must always be paid. Violations fall under the prosecution act of the German Copyright Law.

© Springer-Verlag Berlin Heidelberg 1979, 1987
Printed in Germany

The use of registered names, trademarks, etc. in this publication does not imply, even in the absence of a specific statement, that such names are exempt from the relevant protective laws and regulations and therefore free for general use.

Typesetting, printing and bookbinding: Universitätsdruckerei H. Stürtz AG, Würzburg
2127/3145-543210

Preface

After the positive response which followed the first edition of this book 6 years ago, the editors were encouraged to prepare a completely reworked second edition that includes the modern advances in this field. There has been a complete change of diagnostic procedure in the detection of renal tumors, which is now based on sonography, computed tomography, and nuclear magnetic resonance imaging, pushing intravenous urography and angiography completely into the background. Also, new methods of treatment with radionuclides using embolisation are incorporated.

The description of morphological structures forms the basis for understanding and recognising pathology of the kidneys and adrenals. The contents of pathological morphology could be extended, as we are of the opinion that the detection and therapy of renal and adrenal tumors are derived from different areas of diagnostic science.

We are indebted to Springer-Verlag for the excellent book production. On behalf of all the authors, we would like to thank our colleagues and associates for their cooperation in the realisation of this project.

The editors hope that this volume will be of interest to radiologists, pathologists, urologists, pediatricists, and also radiotherapists.

Essen, April 1987

EBERHARD LÖHR
LUTZ-DIETRICH LEDER

List of Contributors

ACKERMANN, R., Medizinische Einrichtungen der Universität Düsseldorf,
Urologische Klinik, Moorenstraße 5, D-4000 Düsseldorf 1

BACHMANN, K.-D., Universitätskinderklinik Münster, Vredenweg 19,
D-4400 Münster

BEHRENDT, H., Urologische Klinik, Universitätsklinikum der Gesamthoch-
schule Essen, Hufelandstraße 55, D-4300 Essen

BILLIMORIA, P.E., Loma Linda University, Medical Center, Department of
Radiology, Loma Linda, CA 92354, USA

DOMINICK, H.CHR., St. Anna-Stift, Kinderklinik, Karolina-Burger-Straße
51, D-6700 Ludwigshafen

GROSS, M.D., Division of Nuclear Medicine, University of Michigan, Medi-
cal Center, Veterans' Administration Hospital, 2215 Fuller Road, Ann
Arbor, MI 48105, USA

HARTUNG, R., Urologische Universitäts-Klinik Rechts der Isar, D-8000
München

HAVERS, W., Klinik und Poliklinik für Kinder- und Jugendmedizin, Uni-
versitätsklinikum Essen, Hufelandstraße 55, D-4300 Essen

HECKEMANN, R., Augusta Krankenanstalt Bochum, Abteilung Radiologie,
Bergstraße 26, D-4630 Bochum

KAUDE, J.V., Department of Radiology, University of Florida, College of
Medicine, Box J-374, JHMHC, Gainesville, FL 32610, USA

KINARD, R.E., Department of Radiology, University of Florida, College
of Medicine, Box J-374, JHMHC, Gainesville, FL 32610, USA

LANG, E.K., University of New Orleans, Medical Center, Department of
Radiology, 1542 Tulane Avenue, New Orleans, La., USA

LEDER, L.-D., Institut für Pathologie, Universitätsklinikum Essen, Hufe-
landstraße 55, D-4300 Essen 1

LÖHR, E., Röntgendiagnostisches Zentralinstitut, Universitätsklinikum Es-
sen, Hufelandstraße 55, D-4300 Essen 1

XVIII List of Contributors

MOSS, A.A., University of Washington, School of Medicine, Department of Radiology SB-05, Seattle, Wash. 98195, USA

MÜLLER, R.-D., Radiologisches Zentrum, Universitätsklinikum Essen, Hufelandstraße 55, D-4300 Essen 1

RICHTER, H.J., Pathologisches Institut, Universitätsklinikum Essen, Hufelandstraße 55, D-4300 Essen 1

SCHERER, E., Strahlenklinik, Universitätsklinikum Essen, Hufelandstraße 55, D-4300 Essen 1

SERDAREVIC, M., Röntgendiagnostisches Zentralinstitut, Universitätsklinikum Essen, Hufelandstraße 55, D-4300 Essen 1

SHAPIRO, B., Division of Nuclear Medicine, University of Michigan, Medical Center, Veterans' Administration Hospital, 2215 Fuller Road, Ann Arbor, MI 48105, USA

SHUMAN, W.P., University of Washington, School of Medicine, Department of Radiology SB-05, Seattle, Wash. 98195, USA

WILLIAMS, J.L., Department of Radiology, University of Florida, College of Medicine, Box J-374, JHMHC, Gainesville, FL 32610, USA

WIRTZ, C., Strahlenklinik, Universitätsklinikum Essen, Hufelandstraße 55, D-4300 Essen 1

Table of Contents

Pathology of Renal and Adrenal Neoplasms

LEDER, L.-D., RICHTER, H.J.

1	Tumors and Tumorlike Lesions of the Kidney in the Adult . . .	1
1.1	General Remarks	1
1.2	Heterotopic Tissue	1
1.2.1	Adrenal Tissue	1
1.2.2	Endometriosis	1
1.2.3	Cartilaginous Islands	1
1.3	Benign Mesenchymal Tumors	1
1.3.1	Leiomyoma and Lipoma	1
1.3.2	Angiomyolipoma	2
1.3.3	Hemangioma	3
1.3.4	Juxtaglomerular Tumor	4
1.3.5	Lymphangioma	4
1.3.6	Fibroma (Renomedullary Interstitial Cell Tumor)	5
1.3.7	Neurogenic Tumors	5
1.4	Malignant Mesenchymal Tumors	5
1.4.1	Leiomyosarcoma	5
1.4.2	Rhabdomyosarcoma	6
1.4.3	Liposarcoma	6
1.4.4	Angiosarcoma (Hemangioendothelioma)	6
1.4.5	Hemangiopericytoma	7
1.4.6	Fibrosarcoma	7
1.4.7	Fibroxanthosarcoma and Malignant Fibrous Histiocytoma	8
1.4.8	Osteogenic Sarcoma and Chondrosarcoma	9
1.4.9	Hemoblastoses	9
1.5	Epithelial Tumors	11
1.5.1	Renal "Adenoma"	11
1.5.2	Renal Cell Oncocytoma	11
1.5.3	Renal Carcinoid	13
1.5.4	Renal Adenocarcinoma	13
2	Renal Neoplasias of Infancy and Childhood	27
2.1	Nephroblastoma	27
2.1.1	General Remarks	27
2.1.2	Epidemiology	27
2.1.3	Cause	27
2.1.4	Gross Pathology	27
2.1.5	Histopathology	29
2.1.6	Special Histologic Variants	32
2.1.7	Grading	35

VIII Table of Contents

2.1.8	Staging	35
2.1.9	Spread and Biologic Behavior	35
2.2	Nodular Renal Blastema and Nephroblastomatosis	36
3	Tumors of the Renal Pelvis	36
3.1	General Remarks	36
3.2	Papilloma	36
3.3	Inverted Papilloma	36
3.4	Fibroepithelial Polyp	36
3.5	Carcinoma	36
3.5.1	Introduction	36
3.5.2	Causes	37
3.5.3	Epidemiology	37
3.5.4	Transitional Cell Carcinoma	38
3.5.5	Squamous Epithelial Carcinoma	40
3.5.6	Adenocarcinoma	40
3.5.7	Undifferentiated Carcinoma and Carcinosarcoma	41
3.5.8	Grading	41
3.5.9	Staging	42
3.5.10	Spread	42
3.5.11	Prognosis	42
4	Tumors of the Adrenal	42
4.1	Introduction	42
4.2	Tumors of the Adrenal Cortex	43
4.2.1	General Remarks	43
4.2.2	Adenomas	44
4.2.3	Primary Carcinoma	46
4.2.4	Secondary Carcinoma	49
4.3	Tumors of the Adrenal Medulla	49
4.3.1	General Remarks	49
4.3.2	Neuroblastoma and Ganglioneuroblastoma	49
4.3.3	Ganglioneuroma	53
4.3.4	Pheochromocytoma	54
4.4	Myelolipoma and Cysts	58
4.4.1	Myelolipoma	58
4.4.2	Cysts	58

Ultrasound Diagnosis of Renal and Pararenal Tumors

HECKEMANN, R.

1	Introduction	69
2	Method	69
2.1	Apparatus	69
2.2	Technique of Examination	69
3	The Abnormal Kidney	69
3.1	Cysts	69
3.1.1	Simple Cysts	69
3.1.2	Atypical Cysts	70
3.1.3	Parapelvic Cysts	71
3.1.4	Adult Polycystic Disease	72

3.1.5	Infantile and Juvenile Polycystic Disease	73
3.2	Solid Tumors	73
3.2.1	Renal Cell Carcinoma	73
3.2.2	"Adenomas" and Oncocytomas	75
3.2.3	Angiomyolipoma	76
3.2.4	Metastases	76
3.2.5	Wilms' Tumor	77
3.2.6	Transitional Cell Carcinoma of the Kidney	77
3.3	Perirenal Lesions	77
3.3.1	Hematomas	77
3.3.2	Abscesses	77
3.3.3	Lymphoceles	78
3.3.4	Pancreatic Pseudocysts	78
4	Extrarenal Lesions	78
5	Diagnostic Procedure in Renal Tumors	79

Computed-Body-Tomography of Renal Carcinoma and Perirenal Masses

LÖHR, E., BILLIMORIA, P.E., SERDAREVIC, M.

1	Introduction	81
2	Anatomy	81
3	Methods	83
3.1	General Considerations	83
3.2	Positioning of the Patient	83
3.3	Contrast	84
3.4	CT Enhancement Values	85
3.5	Levels of Confidence	86
4	Renal Carcinoma	90
4.1	Staging	90
4.2	CT Findings	90
4.3	Tumor Spread	93
4.4	Tumor Invading Vena Cava	93
5	Clear Cell Carcinoma	94
6	Metastases Simulating Primary Renal Carcinoma	94
7	Malignant Tumors of Different Histologic Pattern	94
8	Benign Tumors	95
8.1	Lipomas	95
8.2	Angiomyolipomas	95
8.3	Angiomas, Aneurysms	98
9	Tumors of the Collecting System, Transitional Cell Carcinoma	98
10	Retroperitoneal Lymphomas	100
11	Neuroblastoma and Nephroblastoma	100
12	Differential Diagnostic Considerations:	
	Pseudotumorous Mass Lesions vs Renal Tumor	103
12.1	Peri- and Paranephritic Abscesses	103

12.2	Xanthogranulomatous Nephritis	103
12.3	Hyperdense Cysts	106
12.4	Pseudotumors of Different Origin	106

Magnetic Resonance Imaging of Renal Mass Lesions

KAUDE, J.V., KINARD, R.E.

1	Introduction	115
2	Equipment	115
3	Technique of Examination	115
3.1	Normal Kidney	116
4	Benign Lesions	116
4.1	Renal Edema	116
4.2	Hydronephrosis	117
4.3	Cysts	117
4.4	Renal Abscess	118
4.5	Renal Hemorrhage	118
4.6	Benign Renal Tumors	122
4.7	Renal Sinus Fibrolipomatosis	122
5	Renal Malignancies	122
5.1	Renal Cell Carcinoma	122
5.1.1	The Primary Tumor	122
5.1.2	Staging and Metastases	123
5.2	Transitional Cell Carcinoma	126
5.3	Renal Sarcoma	127
5.4	Renal Lymphoma	127
5.5	Metastatic Renal Tumors	127
6	Differential Diagnosis – Malignant vs Benign Lesions	129
7	Conclusion	130

¹²⁵I Embolotherapy of Renal Tumors

LANG, E.K.

1	Introduction	133
2	Concepts Governing Transcatheter Interstitial Radiation Therapy	133
3	Technique	134
4	Clinical Results	136

Adrenal Mass Lesions in Infants and Children

WILLIAMS, J.L., KAUDE, J.V.

1	Introduction	141
2	Imaging Techniques	141
2.1	Abdominal Radiography	141
2.2	Excretory Urography	141

2.3	Ultrasound	141
2.4	Computed Tomography	142
2.5	Radionuclide Imaging	142
2.6	Angiography	142
2.7	Magnetic Resonance Imaging	143
3	Normal Adrenal Glands	143
4	Pathological Lesions	144
4.1	Congenital Adrenal Hyperplasia	144
4.2	Adrenal Hemorrhage	144
4.3	Wolman's Disease	146
4.4	Neuroblastoma	147
4.5	Adrenocortical Tumors	148
4.6	Pheochromocytoma	151

Computed Tomography of the Adrenal Glands

MOSS, A.A., SHUMAN, W.P.

1	Introduction	153
2	Techniques of Examination	153
3	Anatomy	153
4	Location	156
4.1	Right Adrenal Gland	156
4.2	Left Adrenal Gland	156
5	Morphology	157
6	Size	157
7	Pathology	157
7.1	Cortical Abnormalities	157
7.1.1	Cushing's Syndrome	157
7.1.2	Primary Aldosteronism (Conn's Syndrome)	161
7.1.3	Nonfunctioning Adrenal Adenoma and Carcinoma	164
7.1.4	Myelolipoma and Adenolipoma	164
7.2	Medullary Abnormalities	165
7.2.1	Pheochromocytoma	165
7.2.2	Neuroblastoma	166
7.3	Secondary Neoplasms	168
7.4	Unusual Primary Tumors	169
7.5	Adrenal Cysts	170
7.6	Miscellaneous Abnormalities	170
7.6.1	Infection	170
7.6.2	Hypoadrenalism	171
7.6.3	Acromegaly	171

Scintigraphic Studies of Renal and Adrenal Function

GROSS, M.D., SHAPIRO, B.

1	Introduction	173
2	Radiopharmaceuticals	173

XII Table of Contents

2.1	Renal Imaging Agents	174
2.2	Adrenal Imaging Agents	174
2.2.1	Adrenal Cortical Imaging Agents	174
2.2.2	Adrenal Medulla Imaging Agents	175
3	Functional Scintigraphy of the Kidneys	176
3.1	Measurement of Glomerular Filtration	176
3.2	Renal Tubular Function Studies	177
3.3	Dynamic Renography	178
3.4	Radionuclide Renal Localization	180
4	Adrenal Cortical Scintigraphy	181
4.1	Normal Adrenal Imaging	181
4.2	Quantification of Adrenal Gland Radiocholesterol Uptake	181
4.3	Adrenal Cortical Tumor Imaging	183
4.3.1	Lesions Producing Cushing's Syndrome	183
4.3.2	Adrenal Lesions Producing Aldosteronism	184
4.3.3	Adrenal Lesions Producing Hyperandrogenism	185
4.3.4	Scintigraphic Evaluation of Clinically Silent Adrenal Masses	186
5	Adrenal Medulla Scintigraphy	186
5.1	Normal mIBG Scintigraphy	186
5.2	Scintigraphic Localization of Lesions Resulting in Catecholamine Excess	186
5.2.1	Localization of Pheochromocytomas	187
5.2.2	Localization of Neuroblastomas	187
5.3	Scintigraphic Localization of Lesions Other than Adrenergic Tumors	188
5.3.1	Localization of Carcinoid Tumors	189
5.3.2	Localization of Medullary Carcinoma of the Thyroid	189
5.3.3	Other Tumors Imaged with mIBG	190

Surgical Management of Renal Cell Carcinoma

BEHRENDT, H.

1	Radical Nephrectomy	193
2	Preoperative Embolization	193
3	Regional and Radical Lymphadenectomy	195
4	Tumor Extension into the Inferior Vena Cava	195
5	Partial Nephrectomy in Renal Cell Carcinoma	196
6	Surgery for Disseminated Disease	197
7	Adjunctive Nephrectomy	197
8	Future Prospects	198

Operative Therapy of Nephroblastoma

HARTUNG, R.

1	Introduction	201
1.1	Incidence, Age, and Sex Distribution	201

1.2	Pathology	201
1.3	Presentation and Clinical Symptoms	201
1.4	Tumor Stages	202
1.5	Diagnostic Procedures	202
2	Treatment	204
2.1	Preoperative Treatment	204
2.2	Operative Technique	204
2.2.1	Tumor Enucleation	205
2.2.2	Extrarenal Wilms' Tumors	206
2.2.3	Surgery of Metastases	206
2.2.4	Intraoperative Complications	206
2.2.5	Postoperative Complications	206
2.3	Postoperative Treatment	207
2.3.1	Follow-up	207
2.3.2	Recurrent Disease	207

Nonoperative Treatment of Renal Cell Carcinoma

MÜLLER, R.-D., SCHERER, E., WIRTZ, C.

1	Introduction	209
2	Radiotherapy	211
3	Chemotherapy	215
4	Hormone Therapy	216

Prenatal Wilms' Tumor

DOMINICK, H.CHR., BACHMANN, K.-D.

1	Introduction	223
2	Development of Prenatal Wilms' Tumor	224
3	Diagnosis	225
3.1	Case 1	225
3.2	Case 2	225
3.3	Case 3	226
4	Related Disorders	226
5	Therapy and Complications	226
6	Prognosis	227

Congenital Neuroblastoma

BACHMANN, K.-D., DOMINICK, H.CHR.

1	Introduction	229
2	Case Studies	231

XIV Table of Contents

2.1	Case 1	231
2.2	Case 2	231
2.3	Case 3	231
2.4	Case 4	232
2.5	Case 5	232
2.6	Case 6	232
2.7	Cases in the Literature	232

Neuroblastoma

DOMINICK, H.CHR., BACHMANN, K.-D.

1	Introduction	241
2	Etiology	241
3	Pathologic Anatomy	242
4	Immunology	243
5	Biochemistry	243
6	Clinical Aspects	244
7	Metastases	245
8	X-ray Findings	245
9	Differential Diagnosis	245
10	Diagnosis	246
11	Therapy	246
12	Prognosis	247

Nonsurgical Management of Wilms' Tumor

HAVERS, W.

1	Introduction	251
2	Prognostic Factors Influencing the Treatment Plan	251
3	Radiation Therapy	252
4	Chemotherapy	253
5	Planning Patient Treatment	254
6	Treatment of Bilateral Wilms' Tumor	255
7	Treatment of Metastases at Diagnosis	255
8	Treatment of Recurrent Disease	256
9	Outlook for Further Development in the Treatment of Wilms' Tumor	256

Immunologic Aspects of Malignant Renal Disease

ACKERMANN, R.

1	General Immunologic Considerations	259
2	Humoral Immunity in Renal Cell Carcinoma	260
3	Cell-Mediated Immunity in Renal Cell Carcinoma	261
4	Renal Carcinoma Antigens Defined by Monoclonal Antibodies	263
5	Immunotherapy	264
Subject Index		269

Pathology of Renal and Adrenal Neoplasms

L.-D. LEDER and H.J. RICHTER

1 Tumors and Tumorlike Lesions of the Kidney in the Adult

1.1 General Remarks

To understand and correctly interpret the morphology of tumors of the kidneys, and in the case of malignancy the morphology of their metastases, it is important to know that the kidney of man is a true metanephros. This means it is of mesodermal origin [165]. On the other hand, there is no doubt of the epithelial character of the tubular cells, which are of mesodermal origin. Thus it is understandable that in renal neoplasms both epithelial and nonepithelial, e.g., mesenchymal, elements may be found, for instance in Wilms' tumor and adenocarcinoma of the kidney.

1.2 Heterotopic Tissue

1.2.1 Adrenal Tissue

It is very seldom that one finds heterotopic tissue in bioptic material. Such observations are usually confined to autopsy studies. APITZ [12] found accessory adrenal tissue in 261 of 4309 individual autopsies. There are no differences between sexes [338]. In most of the cases, the adrenal tissue is located near the upper pole of the kidney.

Macroscopically, one sees a subcapsular nodular plaque with a roundish shape and a yellow-orange color. Some of these nodules may reach a size of up to 2 cm in diameter. Up to now ectopic adrenal tissue has never been found in the medulla.

Microscopically, the lesions look very much like normal adrenal cortical tissue, sometimes mimicking the normal zones [338].

1.2.2 Endometriosis

There are rare and occasional reports describing involvement of the urinary tract by endometriosis [1, 24]. Histologically, typical endometrial glands surrounded by endometrial stroma are found. Since endometrial glands follow the normal cycle, mitoses may occur during the proliferation phase. This must be kept in mind in order to avoid misinterpreting the condition as primary or metastatic adenocarcinoma of the kidney.

1.2.3 Cartilaginous Islands

Cartilaginous islands are designated by some as true benign tumors, namely chondromas [238]. However, cartilage is found within renal parenchyma, usually together with renal dysplasia [37]. Also, there are histogenetic relationships among renal dysplasia, nodular renal blastema, mesoblastic nephroma, and nephroblastoma, which are discussed in Sect. 2. Macroscopically and histologically the lesions present the typical appearance of mature cartilaginous tissue. It should be stressed here that neither heterotopic adrenal tissue in the kidney nor endometrial tissue (if confined to the kidney) nor cartilaginous islands appear to produce any symptoms during life.

1.3 Benign Mesenchymal Tumors

1.3.1 Leiomyoma and Lipoma

Leiomyomas and lipomas are discussed together here because it is known that both kinds of tumors may be found side by side in the same kidney and, furthermore, adipose tissue may be mixed up with smooth muscle cells to form leiomyolipomas [12, 333].

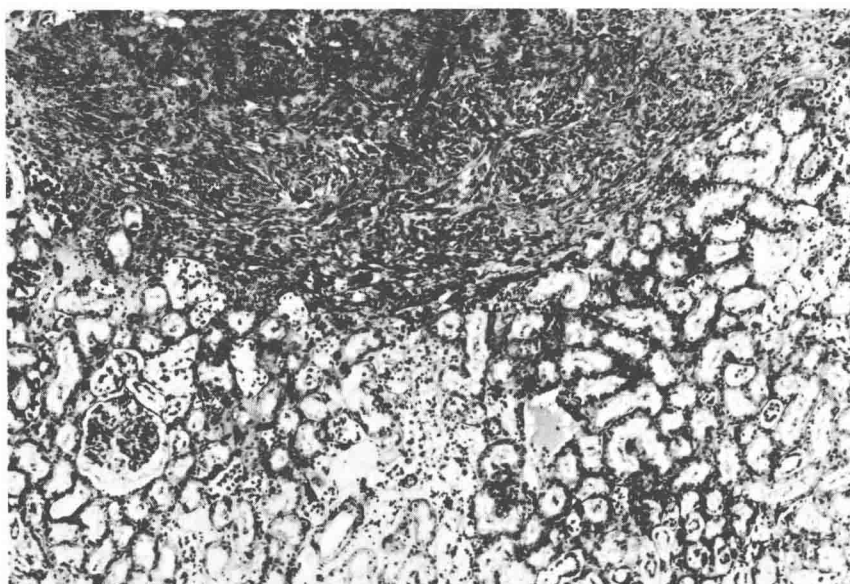


Fig. 1. Renal cortical nodule (leiomyoma) consisting predominantly of smooth muscle cells. H & E, $\times 56$

Benign mesenchymal tumors are much more common in women than in men. This holds true for both leiomyomas and lipomas as well as for myolipomas. The lesions are very rare and most of them are found in the elderly. Usually, they are detected incidentally at autopsy, since they are in general too small to cause clinical symptoms. Their origin is not quite clear. If tumor-terms are used, then a neoplastic origin is implied; however, this has not yet been proved. A hamartomatous origin may also be taken into consideration [161]. BENNINGTON and BECKWITH [31] think that the term choristoma, which denotes tumorlike formations of displaced tissues, is much more appropriate.

Generally, the lesions have little diagnostic importance in comparison to primary malignant neoplasms of the renal parenchyma. BENNINGTON and BECKWITH [31], for instance, mention that before 1975 fewer than 50 leiomyomas and lipomas had been described that were large enough to produce clinical symptoms during life, and DINEEN et al. [100] found only 17 cases of renal lipomas in the literature that were large enough to be of surgical significance. It may be mentioned in this connection that leiomyomas and lipomas are found relatively often in patients with tuberous sclerosis [198], which was first described by FISCHER [122].

Macroscopically, most of the lesions represent only small nests of fatty tissue and/or smooth

muscle and are usually found in the cortex. Many range from 0.1–1.0 cm in diameter. They are relatively sharply circumscribed.

Microscopically, they consist of mature adipose tissue or smooth muscle cells (Fig. 1) or both.

1.3.2 Angiomyolipoma

This lesion should also be regarded as a choristoma [31]. It is known that about 80% of patients with tuberous sclerosis bear angiomyolipomas [79, 186, 108, 116]. On the other hand, a considerable number of cases is not related to tuberous sclerosis.

Angiomyolipomas are not extremely rare. Whereas FARROW et al. [116] reported only 32 cases out of 2409 surgically excised tumorous kidneys observed over a period of 50 years, MA and CHAN [260] noted 57 cases including five personal observations, and BUSCH et al. [57] found in the literature as many as 200 cases not associated with tuberous sclerosis.

According to FARROW et al. [116], the lesions are usually solitary and unilateral. Size and clinical symptoms vary considerably from case to case. Fever or hypertension may be the first and only clinical symptoms [135, 61]. Interestingly enough, YUM et al. [432] identified juxtaglomerular cells with typical rhomboid and spheric granules within an angiomyolipoma.