Pathology of the Lung

Edited by
W. Timens and H.H. Popper
Pathology of the Lung
This book is one in a series of European Respiratory Monographs. Each individual issue provides a comprehensive overview of one specific clinical area of respiratory health, communicating information about the most advanced techniques and systems needed to investigate it. It provides factual and useful scientific detail, drawing on specific case studies and looking into the diagnosis and management of individual patients. Previously published titles in this series are listed at the back of this book with details of how they can be purchased.
Pathology of the Lung

Edited by
W. Timens and H.H. Popper
The European Respiratory Monograph

Number 39

March 2007

CONTENTS

The Guest Editors

Preface

Introduction

1. Cystic lesions of the lung in children: classification and controversies
   J.T. Stocker, A.N. Husain

2. Progress in the pathology of diffuse lung disease in infancy: changing concepts and diagnostic challenges
   M.K. Dishop, C. Langston

3. The differential diagnosis of pulmonary pre-invasive lesions
   K.M. Kerr, H.H. Popper

4. New concepts in pulmonary oncology
   R. Pirker, H.H. Popper

5. What is a rare tumour and how should it be dealt with clinically?
   N. Girard, M. Barbarescoi, J.-F. Cordier, B. Murer

6. Smoking-induced lung disease
   H.H. Popper, W. Timens

7. What is new in chronic obstructive pulmonary disease?
   J.L. Wright, H.A.M. Kerstjens, W. Timens

8. Idiopathic interstitial pneumonia and connective tissue disorder-related interstitial lung disease
   O.A. Harari, P.S. Hasleton

9. Chronic hypersensitivity pneumonitis and its differential diagnosis
   A. Churg, N.L. Müller, J. Flint, J.L. Wright

10. Pathology of pulmonary involvement in inflammatory bowel disease
    T.V. Colby, P. Camus
Wim Timens studied Medicine at the University of Groningen (Groningen, the Netherlands) and received his MD in 1983. After a short period as a research fellow, he followed the residency training programme at the Dept of Pathology, University Hospital Groningen, and was registered as a pathologist in 1990. In 1988, he received a PhD with his thesis on “Structure and function of the human spleen”. In 1990, Wim worked as a post-doc at the Cross-Cancer Institute (Edmonton, Canada), and later that year joined the faculty of the Dept of Pathology, University Hospital Groningen. In 1992, he was appointed as an Associate Professor in this department, and in 1994 as a full Professor in Pathology.

Although he started his career in haematopathology and immunopathology, areas in which he also started his research interests, Wim soon became very interested in and challenged by pulmonary pathology. Over the last 16 years, he has dedicated his diagnostic pathology work to the whole area of pulmonary diseases. He has worked on different research themes involving lung pathology, and has been involved in and leads many research projects, most with external funding and almost invariably in close collaboration with the Dept of Pulmonology and several other disciplines. Within the research themes, there is a strong focus on the pathogenesis of obstructive lung diseases, which is a main research area in the University Medical Center Groningen. He is a Co-Chair of the Groningen Research Institute on Asthma and COPD (GRIAC) and has been a member of the Scientific Board of the Netherlands Asthma Foundation for many years.

Helmut H. Popper studied Medicine at the University of Graz, Medical Faculty (Graz, Austria) and received his MD in 1973. From 1973, he worked as an Assistant at the Institute of Pharmacology, University of Graz, but moved to the Pathology Dept in 1975. In 1984, Helmut finished his training with board certification in Pathology and became Associate Prof. of Pathology with residency at the Institute of Pathology. In 1986, he was appointed Head of the Laboratory of Environmental Pathology and in 1991 was certified for Cytopathology, becoming Prof. of Pathology in 1992. In 1999, Helmut received further certification for Human Genetics and was appointed as Head of the Laboratories for Molecular Cytogenetics and Environmental & Respiratory Tract Pathology in 2000. In 1988 and 1989, he spent some time at the Lovelace Inhalation Toxicology Research Institute, Albuquerque (NM, USA), to learn methods in inhalation toxicology and research. His main focus is genetics in lung diseases with a special interest in lung cancer and sarcoidosis.

From 1999–2003 he served as Chair of the Pulmonary Pathology Working Group of the European Society of Pathology, and again from 2005–2007. Since 2001, Helmut has stood as Chair of an EU project on rare pulmonary diseases, and organised the establishment of the European case collection on rare pulmonary diseases. He is currently Co-Chair for the Latin American–European Pulmonary Pathology Working Group. He is also a member of the Editorial Board of Virchows Archiv, and an Associate Editor of Archives of Pathology & Laboratory Medicine.
Preface

In order to better understand clinical issues in pulmonary medicine, it has become obvious that knowledge about structural changes in the lungs has to be improved. The lungs constitute the largest contact area of the human body with the environment and surroundings. In addition, the lungs serve as a capillary filter of venous blood, involving heavy exposure to the external and internal environment. This poses a unique situation in which both exposure to external agents and "endogenous" changes have to be considered when evaluating pathological changes of the lungs. Therefore, a close collaboration between pathologists and clinicians is of great importance in diagnosing and treating all disorders displaying alteration of lung structure. It is, therefore, a pleasure to introduce the current issue of the *European Respiratory Monograph*, which is dedicated to lung pathology. The Monograph covers the pathology of neoplastic diseases, infections, obstructive and interstitial lung diseases, and pulmonary manifestations of systemic diseases, with all the chapters written by distinguished experts. It is my firm belief that this Monograph will serve as a tool for clinicians to better understand lung pathology. It will also be useful to clinicians who meet patients with pulmonary diseases. As it is a comprehensive update of the pathological field I am assured that pathologists and scientists within the field, will also find this Monograph helpful.

K. Larsson

Editor in Chief
INTRODUCTION

W. Timens*, H.H. Popper#

*Dept of Pathology, University Medical Center Groningen, University of Groningen, Groningen, the Netherlands. Fax: 31 503632510; E-mail: w.timens@path.azg.nl  #Dept of Pathology, Laboratories for Molecular Cytogenetics, Environmental and Respiratory Pathology, Medical University of Graz, Austria. Fax: 43 31638583646; E-mail: Helmut.popper@meduni-graz.at

Pathology is a discipline that, in interaction with clinicians active in pulmonary medicine, plays an important role in the diagnosis and, in recent years, also in the evaluation of therapy. No less important, the core research in pathology has always been aimed at understanding disease mechanisms. With respect to understanding and diagnosing diseases of the lung, an important challenge and difficulty is that this unique organ has an open connection with the outside world. This results in a large variation in disease presentation, with a mix of hallmarks of underlying disease with environmental effects, such as from smoke, pollution or occupational exposures. With respect to pulmonary diseases with the resulting complex presentation, pathology has its own role, which, in a multidisciplinary approach, can contribute to a better understanding of pathogenesis.

The present Monograph is aimed at clinicians active in pulmonary medicine and at pathologists with an active interest in pulmonary pathology. This Monograph does not cover a truly thematic subject, but rather new trends and advances in a discipline with an active interaction with pulmonary medicine. The Guest Editors have chosen different areas of pulmonary pathology to focus on and have asked experts within those fields to contribute, adding to new views and perspectives. Therefore, several chapters have been co-written by a pathologist and a physician. It was the Guest Editors’ aim to obtain an interesting blend of state-of-the-art work, which is relevant for both pulmonary physicians and pathologists based in daily clinical work, as well as colleagues with an interest in recent research advances in pulmonology.

As this Monograph exemplifies the long-standing fruitful collaboration between pathology and different areas of pulmonary medicine, it is hoped that it may also serve in sustaining this interaction and collaboration.