GROSS LUNG PATHOLOGY
A COLOR ATLAS

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Prelude

It is with great pleasure that I browsed through the final draft of the “Gross Lung Pathology: A Color Atlas” edited by Drs. Ritu Kulshrestha, P. Vaideeswar, G. Amonkar, U.G. Moesenbacher and H. Popper. The atlas no doubt, is based on painstaking collection and study of a rich and varied pathological material covering a whole range of pulmonary pathology. The collaborative efforts between three institutions from India and two from Austria has obviously provided for specimens of diseases prevalent in the tropics as well as in the temperate climate. The inclusion of radiological features along with the respective lung pathology adds to the diagnostic repertoire. I am sure that the atlas will be very useful for the students and others dealing with pulmonary diseases. I wonder if addition of histological features would not have enhanced its utility. Probably this has been reserved for a future venture.

I take this opportunity to congratulate the editors and the other contributors for their commendable efforts to bring out this well documented and produced atlas on a subject of great relevance to health professionals.

Prof. P.N. Tandon
Chairman Governing Body
Vallabhbhai Patel Chest Institute
University of Delhi
Foreword

The science of Pulmonary Pathology and Pulmonary Medicine is the correlation of the histopathological and gross features with clinical and radiological appearances in a careful and systematic manner. Examination of the gross lung pathology is a crucial and essential step in this process. However due to a reduction in the number of autopsies and open lung biopsies, the focus on gross pathology is getting reduced by the day. In this color atlas, Dr Ritu Kulshrestha has fostered an outstanding collaboration among experts from India and Europe and made an attempt to present the common gross lung pathologies in a practical and concise manner for medical students, practicing pathologists and clinicians. I hope this edition will serve as a useful guide in understanding the complex and challenging pathologies, particularly the gross appearances of various pulmonary diseases.

Prof. S. N. Gaur
Director,
V.P.C.I
Preface

Pulmonary pathology is a medical discipline where knowledge of the etiology, function (pathophysiology), anatomical and clinical manifestations of the disease are essential for final diagnosis by the pulmonary pathologist. This 'Color Atlas of Gross Lung Pathology' is intended to provide students with a clear and concise presentation of the acute and chronic lung disease pathology seen on gross examination of the specimen. Inspite of the rapid geometric expansion in the fields of radiology, the diagnosis of human disease is incomplete without a clear understanding of the ultimate expression of disease as anatomical changes in tissues and organs. Therefore this atlas is prepared to provide readily understandable representations of common human diseases, concentrating on pathologic anatomy and relating the anatomical changes to the functional, radiological and clinical manifestations of disease. The atlas is meant to be an adjunct when studying gross pathology specimens in the pathology museums. It is hoped that this atlas will serve as a useful learning aid for students involved in their first human pulmonary pathology course and as a review for medical students, physicians and other health care professionals at subsequent stages in their careers.

Editors
I gratefully acknowledge the contribution of the National and International faculty colleagues who have participated in this venture and provided the best of their inputs. I pay my sincerest appreciation to students and staff of the department of Pathology at V. P. Chest Institute for their untiring assistance. I also owe a great deal of gratitude to my family who have borne with my being busy and helping me sort out difficult matters. Nothing could have been accomplished without the efficient editing and publishing by M/s Vidyanilyam Publishers, Delhi, who have done an excellent job. Ms D. Soundarya and Ms Jyoti Singhal deserve my special thanks for coordinating the process of publication.
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Chapter 1

Normal Lung Anatomy

Ritu Kulshrestha

A. Right Lung
B. Left Lung
C. Airways
D. Bronchopulmonary Segments
E. Blood Supply
The lungs are paired, roughly conical asymmetric organs. Their normal combined weight averages 850 gm in men and 750 gm in women (Whimster WF, 1974). The lung has three surfaces, a convex costal surface abutting the rib cage, a concave mediastinal surface and a concave diaphragmatic surface. The **costal surface of the lung (external or thoracic surface)** is smooth, convex, of considerable extent, and corresponds to the form of the cavity of the chest, being deeper behind than in front. It is in contact with the costal pleura, and presents, in specimens which have been hardened in situ, with slight grooves corresponding with the overlying ribs.

Towards the posterior portion of the mediastinal surface lies the hilum of the lung, where the structures which form the root of the lung enter and leave the viscus. The root is formed by the bronchus, the pulmonary artery, the pulmonary veins, the bronchial arteries and veins, the pulmonary plexuses of nerves, lymphatic vessels, bronchial lymph glands, and areolar tissue, all of which are enclosed by a reflection of the pleura. The chief structures composing the root of each lung are arranged in a similar manner from before backward on both sides, viz., the upper of the two pulmonary veins in front; the pulmonary artery in the middle; and the bronchus, together with the bronchial vessels, behind. From above downward, on the two sides, their arrangement differs, thus: On the right side their position is eparterial bronchus, pulmonary artery, hyparterial bronchus, pulmonary veins, but on the left side their position is pulmonary artery, bronchus, pulmonary veins. The lower of the two pulmonary veins, is situated below the bronchus, at the apex or lowest part of the hilus (West J et al, 1964). The pulmonary ligament droops down from the hilum of the lung and terminates in a free, or falciform, edge. The free surfaces of the lung are covered by a serous membrane, the visceral pleura, which is reflected over the hilum to cover the mediastinum, chest wall and diaphragm as the parietal pleura.

The **apex of the lung** is rounded, and extends into the root of the neck, reaching from 2.5 to 4 cm above the level of the sternal end of the first rib. A sulcus is produced by the subclavian artery as it curves in front of the pleura immediately below the apex. The **base of the lung** is broad, concave, and rests upon the convex surface of the diaphragm, which separates the right lung from the right lobe of the liver, and the left lung from the left lobe of the liver, the stomach, and the spleen. Since the diaphragm extends higher on the right than on the left side, the concavity on the base of the right lung is deeper than that on the left. The ventilation/perfusion ratio is higher in the apex of lung than it is in the base of lung (Permutt S et al, 1962).

**A. The right lung** is divided into three lobes, superior, middle and inferior by two interlobular fissures. The major fissure follows an oblique course from a level above the hilum dorsally to the base of the lung anteriorly, dividing the inferior lobe from the remainder of the lung. The minor fissure is nearly horizontal and separates the superior from the middle lobe.

The middle lobe, the smallest lobe of the right lung, is wedge-shaped, and includes the lower part of the anterior border and the anterior part of the base of the lung. Certain anatomical characteristics make the right middle lobe susceptible to transient obstruction as a result of inflammation or edema. These include the narrow diameter of the lobar bronchus and acute take-off angle which create poor conditions for its drainage. Relative anatomical isolation of the middle lobe and poor collateral ventilation decrease the chance of reinflation once atelectasis occurs. Right middle lobe syndrome (RMLS) refers to atelectasis in the right middle lobe of the lung which is caused by various etiologies. It is characterized by a wedge-shaped density that extends anteriorly and inferiorly from the hilum of the lung, which is best visualized using lateral chest radiography. The superior and inferior lobes are similar to their counterparts on the left lung. The right lung is slightly larger than the left because of the space required to accommodate the heart on the left side of the mediastinum however the vertical (cephalocaudal) dimension of the right lung is less than that of the left lung because of the higher position of the right hemidiaphragm.
Chapter 2

Principles, techniques and guidelines for fixation and grossing of the lung

Ritu Kulshrestha

A. Receiving the specimen
B. Examination of the unfixed lung specimen
C. Fixation and fixatives
D. Guidelines for the gross examination of lung
E. Grossing of neoplastic lesions
Specimen processing in the histopathology laboratory is a complex and labor-intensive process which needs thorough quality control at each and every step inorder to ensure the quality of the pathology reports.

A. Receiving the specimen

On receipt of the specimen in a histology laboratory, it should first be assigned a unique specimen number and a worksheet should be created for use by the histopathology technicians as they move the specimen through its subsequent steps. It is during this process that demographics are entered for the patient, and it is here that most mistakes are discovered and should be timely corrected. These mistakes commonly include:

1. Incomplete specimen requisitions
2. Missing, nonspecific or incorrect specimen site information
3. Specimen and requisition information do not match
4. No registration number
5. Specimens are incompletely labeled.
6. Improper preservative
7. Wrong patient information

B. Examination of the unfixed lung specimen

On examination of the specimen, the type of lung biopsy submitted should be noted as per table I. The worksheet should indicate whether the specimen was received fresh or fixed, intact or open. The first step is to orient the specimen and determine the anatomic location of the lesion. This is especially important if the lesion is suspected malignancy. The clinician would usually provide some means of identifying the superior, inferior, medial and lateral borders (Attaching sutures of different lengths or types to the specimen). These should be identified as per the requisition form provided and marked by indelible ink of different colors at the Grossing room inking station. The weight of the whole specimen should be recorded. The specimen should then be described in a logical sequential fashion with a clear description of gross abnormalities, their size, color and location.

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<th>Procedure</th>
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<td>Removal of an entire lung</td>
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<tr>
<td>Lobectomy</td>
<td>Removal of one lobe of the lung</td>
</tr>
<tr>
<td>Sleeve Lobectomy</td>
<td>Removal of a cancerous lobe of the lung along with part of the bronchus that attaches to it</td>
</tr>
<tr>
<td>Wedge Resection</td>
<td>Removal of a small, wedge-shaped portion of the lung by VATS.</td>
</tr>
<tr>
<td>Segmentectomy</td>
<td>Removes a larger portion of the lung lobe than a wedge resection, but not the whole lobe.</td>
</tr>
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C. Fixation and fixatives

The ideal fixative to be used on the specimen and its optimal duration depends upon the pathological techniques which are intended to be carried out and therefore vary depending upon the suspected clinical etiology of the case. One block of tissue should be processed for frozen section where required.

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<td>Conventional histology and/or immunohistochemistry</td>
<td>4% Neutral buffered formalin (NBF) (in phosphate buffer)</td>
<td>24 hours</td>
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<tr>
<td>Immunofluorescence</td>
<td>None if tissue is flash frozen. 95% ethanol or acetone for touch preparations</td>
<td>30 seconds-1 minute</td>
</tr>
<tr>
<td>Electron Microscopy</td>
<td>4% buffered paraformaldehyde-glutaraldehyde fixative</td>
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<tr>
<td>In situ Hybridisation</td>
<td>NBF for DNA studies. Frozen tissue preferred for RNA studies</td>
<td>6-12 hours at 4°C</td>
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Chapter 3

Radiological diagnosis of lung pathology

Balakrishnan Menon, Ritu Kulshrestha

1. Infectious lung diseases
2. Hemodynamic disorders involving lung
3. Interstitial lung diseases
4. Neoplastic lung disease
The chest radiograph and the computed tomography (CT scan) techniques are the prime imaging investigations in respiratory medicine. In the radiological examination of the lung it is recommended to systematically examine the following areas for abnormalities: (1) Tracheobronchial tree (2) Pulmonary vessels (3) Pulmonary hilum (4) Lung parenchyma and Pleura.

**Tracheobronchial tree:** The lumen of the trachea and main bronchi is visualized for any narrowing and/or dilatation of the air column. This may be caused by any space occupying lesion or an abnormal vessel or intrinsic disease of the tracheobronchial wall. In bronchiectasis, prominent ‘Tramline shadows' may be caused by thickened bronchial walls. Ectatic bronchi filled with mucus may mimic pulmonary vessels and lead to an impression of accentuated bronchovascular bundles and a ‘Dirty Chest' radiograph. When the ectasia with mucus retention is confined to only one lobar or segmental bronchus and the proximal segments of its ramifications produce an image of gloved fingers or gloved hand and this is called as 'Gloved fingers/hand appearance'. This often denotes presence of a long standing bronchial obstruction. In the **Air Bronchogram sign**, the air containing bronchi stand out in a consolidated segment of the lung resulting in a configuration known as air bronchogram. This sign reliably indicates whether the opacity is intrapulmonary and not pleural or mediastinal in location. The most common causes of an air bronchogram are pneumonia and pulmonary edema. Atelectasis caused by pleural effusion, pneumothorax or bronchiectasis and less commonly bronchioloalveolar carcinoma and lymphoma growing around airways without compressing them can give rise to air bronchogram (Kuriyama K, 1987).

**Pulmonary Hila:** The complex shadow of the pulmonary hilum is composed mainly of pulmonary arteries and veins and to a lesser extent bronchi. When the hilar shadow appears enlarged the lateral view helps to analyze the enlarged nodes in the hilar and subcarinal region which increase the opacity in this area. To distinguish hilar/lymph node mass from enlarged vessel a CT scan with contrast may be required. The administration of intravenous contrast material can facilitate the distinction on a CT scan image of enhancing pulmonary vessels from nonenhancing hilar lymph nodes.

**Pulmonary Vasculature:** The normal ratio of upper to lower lung perfusion is, 1:3. Therefore the normal pulmonary vascular markings appear more prominent in the medial lower lungs. The pulmonary arteries follow the course of the bronchi and run vertically. The pulmonary veins are variable in their size and course. They empty lower and course horizontally. Increase in vascular prominences can represent engorgement of arteries, veins, lymphatics, or all three. In pulmonary artery hypertension a central widening of all pulmonary arteries with narrowing of the peripheral arteries is indicative of the diagnosis. In venous prominence due to regurgitation of blood from left ventricle to lungs (mitral stenosis, congestive heart failure(CHF), myocardial infarction, mitral regurgitation), an upper lobe engorgement is seen on Xray Chest with presence of horizontal linear densities which are called Kerley's lines.

**Pulmonary Parenchyma:** The normal lung parenchyma does not produce a separate shadow on chest radiograph. Loss of lung parenchyma is seen as **Hyperlucency**. Increase in density due to diseases involving pulmonary parenchyma are often "patchy" and called infiltrates. **Infiltrates** are “patchy” density associated with infections, neoplasms, and hemorrhage in the lung parenchyma. For further localization; the silhouette sign, mediastinal shift, infiltrate identification (air bronchogram sign) are used. **Silhouette sign:** This occurs because of loss of density contrast borders due to increased density of another tissue. If the lung tissue is replaced by blood, pus, or tumor, the radiographic outlines of the mediastinal or diaphragmatic organs superimposed on these infiltrated areas will disappear (Felson B, 1950). The lesions are classified on the basis of location into upper/lower lobe and central/peripheral. Then the different patterns associated with the various diseases in the differential are used to further narrow the diagnostic considerations.

Tram track appearance- Bronchiectasis showing dilated bronchi giving appearance of tram track.
1. **Infectious lung diseases**: These can present with a vast variety of radiological patterns: Bacterial pneumonia can present as (i) lobar consolidation with expansion of the lobe and air bronchogram (lobar pneumonia). (ii) Tree-in-bud pattern (alveolar opacities in bronchopneumonia, small airway disease caused by aspiration and panbronchiolitis): This pattern is seen on CT scan and consists of centrilobular nodules in association with branching centrilobular linear opacities, which resemble a budding tree branch. (iii) Nodular opacities (Ground glass appearance) for eg. pneumocystis carinii pneumonia which presents with multiple Ground glass opacities. The most frequent complications of pneumonia are pleural effusion and cavitation and these show up as lucent patches in the density. Tuberculosis presents with a vast variety of radiological patterns ranging from multiple parenchymal opacities which may be lobular, multilobular or subsegmental. The airway and pleura are also involved. Acute interstitial/viral pneumonias give a pattern of finely reticulated densities.

Fungal infections of the lung present as 'Mycetoma' (a mass of matted hyphae in a ball) usually in a preexisting cavity. A **cavity** is defined by the Fleischner society as a gas filled space within a zone of pulmonary consolidation, a mass or a nodule. There may or may not be an accompanying fluid level (Tuddenham WJ, 1984). The morphology of the cavity is also helpful. Lung abscesses and benign lesions have thinner, smoother walls than cavitating malignant neoplasms. The air within the cavity forms a peripheral halo or crescent between the intracavitary mass and the cavity wall, giving rise to the 'Air crescent sign'. Invasive fungal infection is seen as bilateral multiple patchy infiltrates that do not follow segmental distribution. The term 'CT halo sign' refers to ground glass attenuation surrounding a nodule on CT scan. The most common cause of a CT halo sign is infection, notably invasive aspergillosis (Primack SL, 1994).

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**Xray showing non homogenous opacity involving right upper, middle and lower zones. Sputum : positive for Nocardia.**
Infections of the lung are classified as bacterial pneumonias, viral pneumonias, fungal and parasitic pneumonias. Most bacterial and viral pneumonias initially are acute inflammatory diseases which may resolve completely with adequate treatment. However pneumonias caused by mycobacteria, parasites or fungi run a protracted course entailing an immune response and incomplete resolution. They heal with focal or diffuse scarring and the risk of chronic restrictive pulmonary disease. Infectious lung diseases can thus present with a vast variety of pathologic and radiological patterns.

A. Pulmonary Tuberculosis

Tuberculosis has been traditionally classified into:

- **Primary tuberculosis** - (absence of preexisting immunity and hypersensitivity
  - Exogenous first infection (never infected)
  - Exogenous reinfection (loss of immunity after death of TB Bacilli)
- **Progressive primary tuberculosis** - inadequate acquired immunity, genetic factors, age - infant, adolescent, aged
- **Secondary or Post primary tuberculosis** - reinfection, adult
  - Endogenous reactivation of latent Tuberculosis
  - Exogenous infection in BCG vaccinated
  - Exogenous superinfection on previous infection or inactive disease

**Primary lung tuberculosis:** The initial focus of infection is a small subpleural granuloma accompanied by granulomatous hilar lymph node infection and is called as Ghons complex or Primary complex. Usually the granulomas resolve and there is no further spread of the infection. Rarely a fibrocalcific lesion of peribronchial lymph node and thickened, scarred lymphatic vessel may develop from the Ghon complex and this combination of late fibrocalcific lesions of the lung and lymph node is referred to as the Ranke complex.

This occurs as a result of bronchial spread leading to extensive, confluent parenchymal granulomatous lung lesions or secondary to bloodstream spread via the pulmonary vein with miliary dissemination of organism into lung or via the pulmonary artery with subsequent systemic dissemination. It can present as number of gross pathologies:

1. **Tuberculoma:** It is a well-circumscribed, usually single, round, firm, nodular lesion which is located immediately beneath a white or slightly yellowish pleura, and can present as a "coin lesion" on chest X-ray.

2. **Tuberculous bronchopneumonia:** This is characterized by an intense alveolar infiltrate of polymorphonuclear cells, macrophages, necrotic debris and multinucleated cells. The hilar and mediastinal lymph nodes are seen to enlarge bilaterally in almost every patient of progressive pulmonary Tuberculosis (this feature is not commonly seen in secondary tuberculosis). These lymph nodes may enlarge sufficiently to rupture into a bronchus leading to tuberculous bronchopneumonia.

3. **Military Tuberculosis:** When resistance to infection is particularly poor, a "miliary" pattern of spread occurs in which there are a myriad of small millet seed (1-3 mm) sized granulomas, either in the lung or in other organs.

**Secondary/ Chronie/ Reactivation tuberculosis:** Secondary tuberculosis is mostly seen in adults as a reactivation of previous infection (or reinfection), particularly when health status declines. The granulomatous inflammation is much more florid and widespread. Typically, the upper lung lobes are most affected. Grossly most of the tissue consists of inflamed, fibrotic and otherwise nonfunctioning lung parenchyma. Bronchial involvement leads to stricture formation with distal bronchiectasis, atelectasis and superimposed infection. Peribronchial lymph nodes are enlarged, show caseation and/or calcification. A number of patterns can occur:

1. **Cavitary tuberculosis:** Extensive necrosis due to high bacterial antigen load, usually occurring in the upper lung or apex, is a characteristic feature of "secondary" or "adult type" tuberculosis. Cavities form when necrosis involves the wall of an airway and the necrotic material is released into the bronchial tree. Communication of the centers of the tuberculous lesions with the airway exposes the bacteria to a high concentration of oxygen and promotes their proliferation. The risk of spread of infection to non-infected persons from individuals with cavitary tuberculosis is very high. Complications which can arise after cavitation include: intracavitary hemorrhage, intracavitary aspergilloma and tuberculous bronchopneumonia.

2. **Tuberculous bronchopneumonia:** This develops in secondary tuberculosis when the infected material is aspirated and may seed other parts of the lung via the airways to produce a tuberculous bronchopneumonia. It presents radiologically as multiple, often bilateral, patchy, "cotton-wool" densities.

3. **Residual Tubercular changes:** The usual residual changes include
   - Walled-off, cavitous necrosis
   - Thin- or thick-walled cavities
   - Emphysema
   - Endobronchial stenosis followed by bronchiectasis
   - Obliterative bronchiolitis
   - Focal pleural and parenchymal scars
   - Focal calcifications
**Slices of right pneumonectomy**

**Case history**
- 19 years female with clinical diagnosis of multi-drug resistant tuberculosis. Persistent sputum acid-fast bacilli positive and repeated anti-tuberculous therapy.

**Gross examination**
- Moderate to marked pleural thickening especially over the upper lobe
- Large, irregular but well-demarcated cavity, occupying most of the upper lobe; Cavity lined by shaggy grayish-white caseous material with focal thin fibrous wall
- Areas of caseation necroses also seen in the middle lobe (arrow)
- Focal hemorrhage in middle lobe (peri-operative trauma) and congested appearance of lower lobe

**Diagnosis:** Secondary Tuberculosis with Cavitation

**Cut surface of right lung**

**Case History**
- 10 years; Male

**Gross examination**
- Mild, patchy pleural thickening with multiple creamy small & large nodules
- The cut surface shows similar nodules with few subpleural wedge-shaped foci of reddish consolidation in upper and lower lobes (arrows)

**Diagnosis:** Primary Progressive Pulmonary Tuberculosis
tonsillectomies and other ear, nose and throat operations. Presently, most abscesses follow the aspiration of foreign material or represent secondary infections of lung carcinomas. Embolism from distant sources is a cause for multiple bilateral abscesses. The most common locations are the right lower lobe, the right upper lobe (especially the subapical segment) and the left lower lobe (Hagan JL, 1983). Complications of untreated lung abscesses include overgrowth of fungi in the cavity, spread to other portions of the lung, massive hemorrhage, bronchopleural fistula with empyema and brain abscess. Actinomyces sp and Nocardia sp may also manifest in this pattern.
Pediatric and developmental lung disease

Ulrike Gruber Moesenbacher, Ritu Kulshrestha, S. Sandhyamani, Helmut Popper

A. Bronchogenic cyst
B. Pulmonary sequestration
C. CCAM (types 0-4)
D. Pulmonary arteriovenous malformation, AV Angioma
E. Pulmonary capillary angiodysplasia
F. Aneurysms
Congenital malformations result from developmental defects that may occur at any stage of the complex processes leading to the formation of mature lung. They include both maturation arrests (localized or generalized) and abnormal proliferation of vascular and/or bronchial buds. They range from total absence of bronchus and lung (agenesis) to hypoplasia (Unilateral of bilateral).

**Phases of lung development**

<table>
<thead>
<tr>
<th>Phase</th>
<th>Weeks of Gestation</th>
<th>Developmental milestones</th>
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<tbody>
<tr>
<td>Embryonic</td>
<td>Upto 6 wks</td>
<td>Lung buds and segmental airways appear. The lungs acquire a pseudoglandular appearance</td>
</tr>
<tr>
<td>Pseudoglandular</td>
<td>6-16 wks</td>
<td>Airway branching is completed, from the conductive airways to the terminal bronchioles. The differentiation of pulmonary vascular bed occurs.</td>
</tr>
<tr>
<td>Canalicular</td>
<td>16-28 wks</td>
<td>The appearance of airspaces which have an attenuated lung epithelium. The differentiation into type I and type II alveolar epithelial cells starts.</td>
</tr>
<tr>
<td>Saccular</td>
<td>28-36 wks</td>
<td>Subdivision of saccules with formation of terminal sacs and development of true alveoli is completed by 36 weeks.</td>
</tr>
<tr>
<td>Alveolar</td>
<td>36 weeks to Birth and 1st year of life</td>
<td>Alveoli development continues with remodeling of airways.</td>
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</table>

The gross examination of the pediatric lung biopsies and resections can many a times give important clues to its correct diagnosis, even before the biopsy slides have been reviewed. The location of the mass, the presence of solid or cystic areas, the vascular and bronchial supply and the onset and severity of symptoms should be noted in each and every case.

**A. Bronchogenic cysts:** They are of variable size, often uniloculated, usually situated in the mediastinum close to the carina (51%) but rarely they may be intrapulmonary. They usually present in adult life as an incidental X-ray finding or with complications such as infection, hemorrhage and rarely neoplastic transformation. Microscopically they show a lining resembling bronchial mucosa with presence of cartilage and mucus glands in the wall. Their differential diagnosis includes, Type I CCAM, which shows polypoidal infoldings, mucus cell hyperplasia and disordered parenchymal growth with smaller cysts resembling bronchioles.

**B. Pulmonary Sequestration:** It is a condition in which a portion of the lung does not communicate with the tracheobronchial tree and does not derive its blood supply from the pulmonary artery. Two types have been described; intralobar and extralobar sequestrations. The extralobar sequestration has its own covering of visceral pleura and is generally detected in infancy because of associated malformations. Intralobar sequestration is embedded in an otherwise normal lung and typically located in the posterior basal segment of the left lower lobe. The sequestered areas often appear cystic. The cysts are lined by columnar or cuboidal epithelium, or may resemble bronchioles surrounded by alveolar ducts and alveoli histologically resembling type 2 CCAM. Radiographically, sequestrations appear solid due to mucus filling of the multiple interconnecting spaces.

**C. Congenital cystic adenomatoid malformation (CCAM)** is a rare abnormality of lung development that stems from abnormal embryogenesis. The earlier the block in pulmonary development, the more serious are the structural abnormalities. If maturation arrest occurs early, it leads to severe generalised hypoplasia and the architectural abnormalities are striking; the lung consists of irregular cystic structures located mostly near the apex. The bronchi show an abnormal course, fewer than normal branches and apical cystic dilatation. Cases due to later disturbances in organogenesis are characterised by the presence of uniformly distributed cysts of similar average size which give the lungs a spongy honeycomb like appearance. The bronchial tree is relatively well developed and the alveoli are present.

**Types of CCAM:** The term congenital adenomatoid malformation was introduced in 1949 (Ch'in KY, Tang MY) They were initially numbered I-III, but have now been renamed as congenital pulmonary airway malformation (CPAM) with five types (0-4), type 0 being tracheobronchial and type 4, alveolar (Stocker JT, 2002).

**Type 0 CCAM:** (congenital acinar dysplasia) This rare type (1-3%) is seen in neonates and causes death in the first few hours of life, due to absence of alveoli. The lungs are small, firm and solid in appearance. On histology bronchial- type airways with cartilage, smooth muscle and glands separated by abundant mesenchymal tissue, are seen.

**Type I CCAM:** (Cystic type of CCAM): This is the most common type. It is a localized lesion which typically affects only a part of a lobe. The boundary between the lesion and the adjacent normal lobe is sharply delineated and so the lesion can be surgically resected. It has the best prognosis of all the types of CCAM. On gross examination, the cysts range from 1-10 cm in diameter. The bronchus is usually atretic. Microscopically the cysts are lined by pseudostratified ciliated columnar epithelium, interspersed with rows of mucus cells, which can undergo malignant transformation into mucinous bronchoalveolar carcinoma.
Type IV CPAM: (Cystic type of CCAM): Radiologically large air filled cysts are seen. Cysts are peripheral and thin walled. Clinically the patient presents as repeated pneumonia in childhood. Microscopically the cysts are lined by flattened alveolar or bronchiolar epithelial cells resting upon loose mesenchymal tissue. If this undergoes malignant transformation, it results in formation of pleuropulmonary blastoma.

Development of Pulmonary vasculature:

The development of the pulmonary vasculature occurs between the 5th and 10th week of intrauterine life. During this time a continuous differentiation of the pulmonary vascular bed occurs, resulting in creation of the separate arterial and venous channels, interconnected by capillaries. The normal stages of vascular development according to Woolard: are a) primitive mesenchyma; b) differentiation in plexiform structures; c) disappearance of the primitive elements and differentiation into vascular structures; d) inadequate development during the embryogenesis of the vascular system with persistence of the primitive vascular communications or arteriovenous malformations. A halt or error in these stages results in vascular malformations at different anatomical sites and with variable morphology (Woolard HH, 1992).

D. Pulmonary arteriovenous malformations: This lesion is a vascular hamartoma resulting from persistence of anastomotic fetal capillaries which presents in older children and adults. This results in abnormal communication between pulmonary arteries and veins bypassing

Case History: 8 years old boy

Gross examination: the cystic structures can be seen. These have thin walls and measure 5-6 cm in diameter

Final Diagnosis: Congenital Cystic Adenomatoid Malformation type 1

Type II CPAM: (Intermediate type of CCAM): This is the second most frequent type and presents in the first month of life with respiratory distress. On gross examination; the lesion is sponge like and consists of multiple small cysts as well as solid areas. Microscopically the cysts are lined by bronchiolar epithelium and separated by poorly developed alveoli or alveolar ducts.

Type III CPAM: (Solid type of CCAM): It is an uncommon subtype which presents in infancy, almost exclusively in male babies. These are large bulky lesions that typically involve and expand the entire lobe of lung, displacing the mediastinum and resultant venous compression which causes hypoplasia of the remaining pulmonary tissue and generalized anasarca. Microscopically there are numerous bronchiolar structures which are separated by airspaces that are small and have a cuboidal lining and resemble fetal lung.
References


A. Chronic venous congestion of lung
B. Pulmonary hemosiderosis
C. Pulmonary thromboembolism
D. Pulmonary Infarct
E. Pulmonary vasculitis syndromes
The hemodynamic disorders involving lung include pulmonary hypertension, pulmonary vascular occlusion with or without infarction, pulmonary vasculitis (which is often a part of a systemic disorder) and rare entities such as pulmonary veno-occlusive disease.

Pulmonary chronic passive congestion: Chronic passive congestion of the lungs occurs secondary to regurgitation of blood from left ventricle to lungs (mitral stenosis, Congestive cardiac failure, myocardial infarction, mitral regurgitation). This results in congestion of the pulmonary veins and development of pulmonary hypertension (Pietra GG et al, 1989). In this slow and progressive process, the lungs are heavy, cyanotic, congested and firm. The gross appearance is characteristic with spontaneous outpouring of pink frothy fluid from cut surfaces. The alveoli contain macrophages stuffed with yellow brown hemosiderin pigment (heart failure cells).

Pulmonary hemosiderosis: Pulmonary hemosiderosis (PH) is characterized by repeated episodes of intra-alveolar bleeding that lead to abnormal accumulation of iron as hemosiderin in alveolar macrophages and subsequent development of pulmonary fibrosis and severe anemia. Pulmonary hemosiderosis can occur as a primary disease of the lungs or can be secondary to cardiovascular or systemic disease. In 1975, Thomas and Irwin divided pulmonary hemosiderosis into 3 categories: (1) pulmonary hemosiderosis associated with antibody to the basement membrane of the lung and kidney (ie, Goodpasture syndrome), (2) pulmonary hemosiderosis associated with hypersensitivity/ immune complex disease, and (3) idiopathic pulmonary hemosiderosis (IPH). A triad of hemoptysis, iron deficiency anemia, and diffuse pulmonary infiltrates characterizes pulmonary hemosiderosis (PH).

Pulmonary embolism: Most pulmonary emboli derive from a free-floating thrombus. Many materials and substances may form emboli and move to the pulmonary circulation; these include fat, tumor, septic emboli, air, amniotic fluid, and injected foreign material (Yakel DL, 1995). The septic emboli are usually secondary to thrombophlebitis or open fractures and may give rise to infected infarcts with suppuration and abscess formation. Macroscopically, emboli are sausage shaped and straddle vascular bifurcations. The size of a pulmonary embolism determines at which points in the pulmonary vasculature it lodges. If the embolism does not cause instant death, pulmonary infarction may follow.

Pulmonary infarcts: These are usually hemorrhagic and often multiple. They tend to be wedge shaped with the base resting near the pleura and the apex pointing towards the hilum. Fibrinous pleuritis can be observed over the infarcted area.

Pulmonary vasculitis: Pulmonary vasculitis describes a number of distinct disorders that are pathologically characterized by the destruction of blood vessels. Lung involvement is more commonly seen with the primary, idiopathic, small vessel or antineutrophil cytoplasmic antibody associated vasculitis; Wegeners granulomatosis, microscopic polyangiitis and Churg Strauss syndrome (Brown KK 2006).

### Classification of Pulmonary vasculitis (Brown KK 2006)

<table>
<thead>
<tr>
<th>Primary idiopathic vasculitis</th>
<th>Secondary vasculitis</th>
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<tbody>
<tr>
<td>1. Small vessel vasculitis</td>
<td>1. Autoimmune disease</td>
</tr>
<tr>
<td>Wegeners granulomatosis</td>
<td>SLE</td>
</tr>
<tr>
<td>Microscopic polyangiitis</td>
<td>Rheumatoid arthritis</td>
</tr>
<tr>
<td>Churg Strauss syndrome</td>
<td>Polyposis/Dermatomyositis</td>
</tr>
<tr>
<td></td>
<td>Scleroderma</td>
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<tr>
<td>2. Medium vessel vasculitis</td>
<td>2. Drug induced</td>
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<tr>
<td>Polyarteritis nodosa</td>
<td></td>
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<tr>
<td>Kawasaki disease</td>
<td></td>
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<tr>
<td>3. Large vessel vasculitis</td>
<td>3. Paraneoplastic</td>
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<tr>
<td>Giant cell arteritis</td>
<td></td>
</tr>
<tr>
<td>Takayasu arteritis</td>
<td></td>
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<tr>
<td>4. Primary immune complex</td>
<td>4. Infection</td>
</tr>
<tr>
<td>mediated vasculitis</td>
<td></td>
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</tbody>
</table>

Wegeners granulomatosis: Wegener granulomatosis is a disease of unknown etiology that is characterized by necrotizing granulomatous vasculitis of the upper and lower respiratory tract, glomerulonephritis, and small-vessel vasculitis of variable degree (classic Wegener granulomatosis) (Cordier JF, 2008). A limited form has also been described, in which the disease is primarily confined to the lung. In this form, involvement of the kidney, skin, and tracheobronchial tree is distinctly unusual (Woywodt A et al, 2006). Pulmonary nodules are the most common chest radiographic manifestation of Wegener granulomatosis; they occur in 40-70% of the cases. Nodules may be solitary or multiple; they are cavitated in as many as 50% of patients with nodules. Both thick- and thin-walled cavities may be present. Their size varies, ranging from 1.5-10 cm, and the nodules may wax and wane over time (Agarwal R et al, 2007). Together with microscopic polyangiitis and Churg-Strauss Syndrome, Wegener's granulomatosis forms part of a group of disorders known as anti-neutrophil cytoplasmic antibody (ANCA) associated systemic vasculitis (AASVs) (Jose RJ et al 2010). On gross examination, multiple, bilateral, solid nodular zones of consolidation with areas of punctuate or geographic necrosis are seen. When acute lung hemorrhage is prominent, the cut surface of the lung is bloody and dark red.
Case History

22 years; Female. Presentation with exertional dyspnea and episodic hemoptysis. History of blood transfusion in the past for “refractory” anemia

Gross Examination

- Mild diffuse pleural thickening
- Firm consistency of the lung with a diffuse rusty brown cut surface

Diagnosis: Idiopathic Pulmonary Hemosiderosis
A. Acute lung injury: Diffuse alveolar damage
B. Pneumoconiosis
C. Diffuse lung diseases with granulomas
D. Idiopathic interstitial pneumonitis
E. Other causes
The diffuse parenchymal lung disease (DPLD) or Interstitial lung disease (ILD), constitute a group of diseases affecting the interstitium in which the lungs have a limited potential to expand and the lung compliance is reduced. They consist of disorders of known causes (collagen vascular disease, environmental or drug related) as well as disorders of unknown cause. The latter include idiopathic interstitial pneumonias (IIPs), granulomatous lung disorders (e.g., sarcoidosis), and other forms of interstitial lung disease (ILD) including lymphangioleiomyomatosis (LAM), pulmonary Langerhans'cell histiocytosis/histiocytosis X (HX), and eosinophilic pneumonia. The most important distinction among the idiopathic interstitial pneumonias is that between idiopathic pulmonary fibrosis and the other interstitial pneumonias (IIPs), which include nonspecific interstitial pneumonia (a provisional term), desquamative interstitial pneumonia, respiratory bronchiolitis-associated interstitial lung disease, acute interstitial pneumonia, cryptogenic organizing pneumonia, and lymphocytic interstitial pneumonia (ATS/ERS, 2002).

DPLD's occur in acute and chronic forms. The classic examples of acute DPLD are the adult respiratory distress syndrome and acute hypersensitivity pneumonitis. Chronic forms include pathogenetically diverse entities such as chronic interstitial pneumonitis in collagen vascular disease, pneumoconiosis, sarcoidosis and Idiopathic. Only patients in early stages of acute ILD may recover completely, later stages and especially the chronic forms of DPLD remit to scarring or progress to extensive interstitial pulmonary fibrosis with honeycombing, pulmonary artery hypertension and development of cor pulmonale. Grossly in ILD, the lungs are firm, stiff and often bosselated. The cut surface shows areas of fibrosis separating air spaces of variable shapes and sizes, called honeycomb lung. Emphysematous areas may be associated and intercalated with fibrotic areas. Recurrent superimposed infections further complicate the microscopic and gross lung pathology. DPLD may be classified according to the etiology as follows:

1. Inhaled substances
   - Inorganic: Silicosis, Asbestosis, Berylliosis, Organic, Hypersensitivity pneumonitis
2. Drug induced
   - Antibiotics, Chemotherapeutic drugs, Antiarrhythmic agents, Statins
3. Connective tissue disease
   - Systemic sclerosis, Polymyositis, Dermatomyositis, Systemic lupus erythematosus, Rheumatoid arthritis

4. Infection
   - Atypical pneumonia, Pneumocystis pneumonia (PCP), Tuberculosis, Chlamydia trachomatis, Respiratory Syncytial Virus

5. Idiopathic
   - Sarcoidosis, Idiopathic pulmonary fibrosis, Hamman-Rich syndrome, Antisynthetase Syndrome

A. Acute Lung Injury: Diffuse alveolar damage DAD represents a nonspecific pattern of acute alveolar injury caused by a variety of noxious agents and radiologically characterised by bilateral generalised ground glass opacification of the lungs more pronounced in lower lobes.

Causes of DAD:
These include: Infection, Drugs, Collagen Vascular disease, Pulmonary hemorrhage syndrome and vasculitis, Others: Shock, sepsis, massive aspiration, toxic fumes

On gross examination the appearance depends upon the phase of ARDS (Poletti V, 2010):

a) In the exudative phase; the lungs are heavy, often weighing over 1 kg each, dark and airless. On slicing of the lung, the cut surfaces exude blood or heavily blood stained watery fluid. Microscopically (congestive atelectasis) with widespread collapse, intense capillary congestion, interstitial edema and distension of lymphatics is seen. Therefore early terms used for acute lung injury have been, 'shock lung' or 'congestive atelectasis' (Tomashefski J. Jr, 2000).

b) In the repair phase; the lower lobe shows diffuse fibrosis by the end of the 2<sup>nd</sup> week. The lungs are contracted and firm with a fine sponge like pattern on their cut surface, this represents bronchiectasis and irregular microcystic distortion of alveolar architecture. It resembles the end stage of any fibrotic process but is reached remarkably quickly. The fibrosis differs by being more cellular and less collagenous. There is resemblance to bronchopulmonary dysplasia in late stages of infantile respiratory distress syndrome.
**Case History**

- 24 years, Male. Presentation as acute febrile illness with jaundice and thrombocytopenia

**Gross Examination**

- Mild, patchy pleural opacification with flakes of fibrin, especially towards the fissural aspects (arrows)
- The lung is firm to feel with a diffuse hemorrhagic cut surface

**Diagnosis:** Adult respiratory Distress Syndrome (Diffuse Alveolar Damage, Leptospirosis Serology Positive)

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**Case History:**

- 42 years old female, admitted to intensive care unit with diagnosis of acute lung injury, and survived. After 2 months on treatment, residual radiological changes were interpreted to be suspicious for tumor.

**Gross Examination:** Cut surface shows features of DAD in organization, with a fine diffuse sponge dike pattern.

- The more solid light brown areas represent fibrosis (organizing pneumonia) whereas the darker areas still contain florid inflammatory foci.

**Diagnosis:** Diffuse alveolar damage in organization.
Lung Neoplasms

Ritu Kulshrestha, Pradeep Vaideeswar, Helmut Popper

A. Benign tumours of lung
B. Malignant epithelial tumours
C. Soft tissue tumours
D. Mesothelial tumours
E. Miscellaneous tumours
F. Metastatic tumours
G. Tumour-like Lesions
The benign and malignant tumours of the tracheobronchial tree and the lung parenchyma have been classified by the WHO classification according to their cell of origin and degree of differentiation into benign, preinvasive and malignant lesions.

**Benign Tumours of Lung**

- **Papillomas:** Squamous cell papilloma / Glandular / Mixed papilloma
- **Adenomas:** Alveolar / Papillary / Bronchial gland adenoma / Adenomas of salivary-gland type / Pleomorphic adenoma / Mucinous cystadenoma

**Papilloma:** A papilloma is a benign epithelial tumour of the trachea and bronchus which is made up of well differentiated mature squamous epithelial cells (squamous papilloma). Squamous papillomas usually arise from the large bronchi. They may be single (mostly in adults) or multiple (papillomatosis, mostly in children). Rarely, mixed squamous and glandular papillomas (mixed papilloma) have been reported to present as an endobronchial polypoid tumour (Inamura K 2011). Macroscopically, it appears as a pedunculated tan white polypoid excrescence in the mucosa of the airway with a smooth or verrucoid surface.

**Alveolar adenoma:** These are rare benign tumours of the lung which present as a peripheral solitary lesion in asymptomatic older patients. Histologically they show benign proliferations of alveolar epithelium and septal mesenchyme (Glaab R 2009).

**Bronchial gland adenoma:** The term bronchial adenoma describes a diverse group of tumors which arise arising from mucous glands and ducts of the trachea or bronchi. It includes the mucous gland adenomas, and other mixed seromucinous tumors arising from mucous glands, adenoid cystic carcinomas, mucoepidermoid carcinomas (England DM 1995). The bronchial mucus gland adenoma is a rare, solitary, benign, well circumscribed, multicystic, predominantly exophytic bronchial tumour. The cut surface is shiny, mucoid, cystic and usually firm. Complete removal of the tumour is curative.

**Pleomorphic adenoma or mixed tumour:** This uncommon primary tumour of the lung can present as a central polypoidal endobronchial lesion or peripheral well circumscribed tumour attached to the bronchi. The cut surface is blue gray translucent and chondroid in appearance and differential diagnosis from hamartoma/chondroma is difficult on gross examination (Sakamoto H, 1991).

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Case History:

10 years old girl, central lung mass identified on radiology. The surgeon noted a tumor that had almost occluded the lobar bronchus.

**Gross Examination**
Solitary well circumscribed lesion which shows shiny mucoid and firm surface.

**Diagnosis:** Bronchial gland adenoma
Adenocarcinoma: Adenocarcinomas are more common in women. It is often peripheral, since the tumour tends to arise from the smaller bronchi. Grossly adenocarcinomas present as poorly circumscribed gray-white-yellow lesions, with irregularly lobulated cut surface and presence of satellite nodules around the main tumour mass. They may be single or multiple. They can have a mucoid glairy appearance if they secrete abundant mucin. Cavitation is extremely unusual. Frequently adenocarcinomas are associated with a lung scar. Pleural spread of carcinoma with fibrosis and puckering is common in 2/3 cases. Very rarely it can present as a large endobronchial polypoidal mass (Kodama T et al 1984).

Bronchioloalveolar carcinoma (BAC): BAC can present in various forms: single peripheral nodule, multiple nodules, diffuse pneumatic like infiltrate and may involve several lobes or even be bilateral (Barsky SH et al 1994). Grossly it may be difficult to recognize, since it may be centered around bronchioles or may mimic a pneumatic consolidation. The architecture of the lung is maintained, however the mucinous type can have a glistening appearance on gross. The BAC can present as bilateral reticulonodular or consolidative lesions and is sometimes associated with a central scar.

Large cell undifferentiated carcinoma: It is almost always centrally located. Cut surface is gray-white, may be lobulated and resembles fish flesh appearance. Hemorrhage and necrosis may be prominent. Differentiation from sarcoma, lymphoma may be difficult on small biopsy.

Small cell carcinoma lung: They typically arise from the central portions of the lung but occasionally can be peripherally located. Grossly it is white-tan, soft, friable, and extensively necrotic lesion centered around a large bronchus. Most patients are males and smokers.

Carcinoid tumours: These are predominantly central but may occur in a peripheral location. Central tumours form a polypoidal growth into the bronchial lumen. The cut surface is grayish yellow, sometimes divided by fibrous septa and very well vascularised. Peripheral carcinoids, tend to be subpleural, multiple and presents grossly as a nonencapsulated gray to tan nodule not bearing any anatomic relationship with a bronchus.

Adenosquamous carcinomas: Most are located peripherally and often are associated with a scar (Scar carcinoma). They exhibit simultaneous squamous and glandular differentiation in the same mass. They can be differentiated on the basis of location from the salivary gland analogue tumors of the lung which tend to arise in the large central airways.

Carcinosarcomas: Rare lesions which may be central or peripheral.

Pulmonary blastoma: They are usually peripherally located, solid and well circumscribed large lesions.