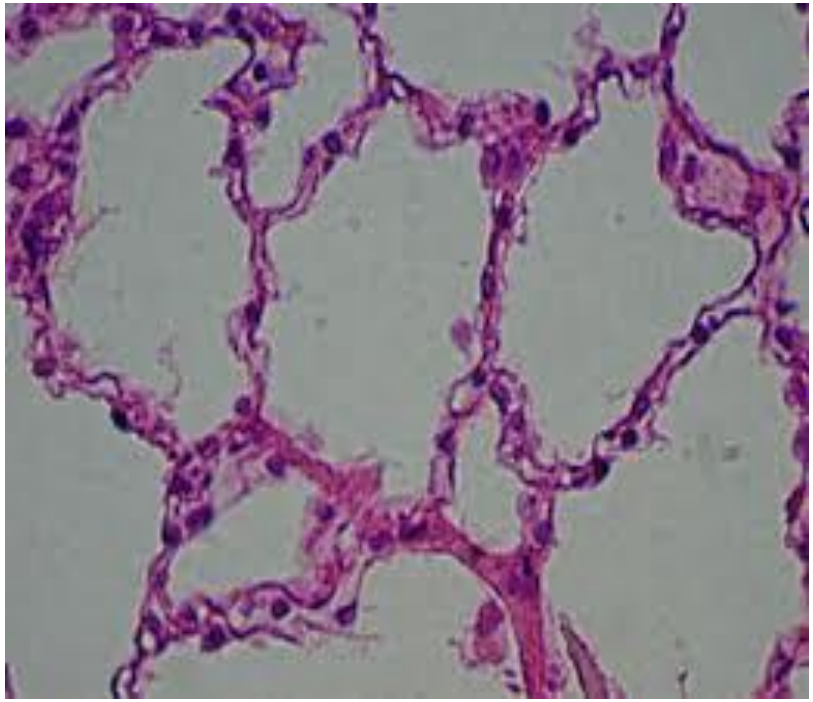
DISEASES OF THE RESPIRATORY SYSTEM LECTURE 5

DR HEYAM AWAD

FRCPATH

RESTRICTIVE, INTERSTITIAL LUNG DISESAES.

- FIROSING DISESES.
- GRANULOMATOUS DISEASES.
- EOSINOPHILIC.
- SMOKING RELATED.



FIBROSING DISEASES

- IDIOPATHIC PULMONARY FIBROSIS
- NONSPECIFIC INTERSTITIAL PNEUMONIA
- CRYPTOGENIC INTERSTITIAL PNEUMONIA
- PNEUMOCONIOSIS

IDIOPATHIC PULMONARY FIBROSIS

- = CRYPTOGENIC FIBROSING ALVEOLITIS.
- IDIOPATHIC, PROGRESSIVE, BILATERAL PULMONARY FIBROSIS.

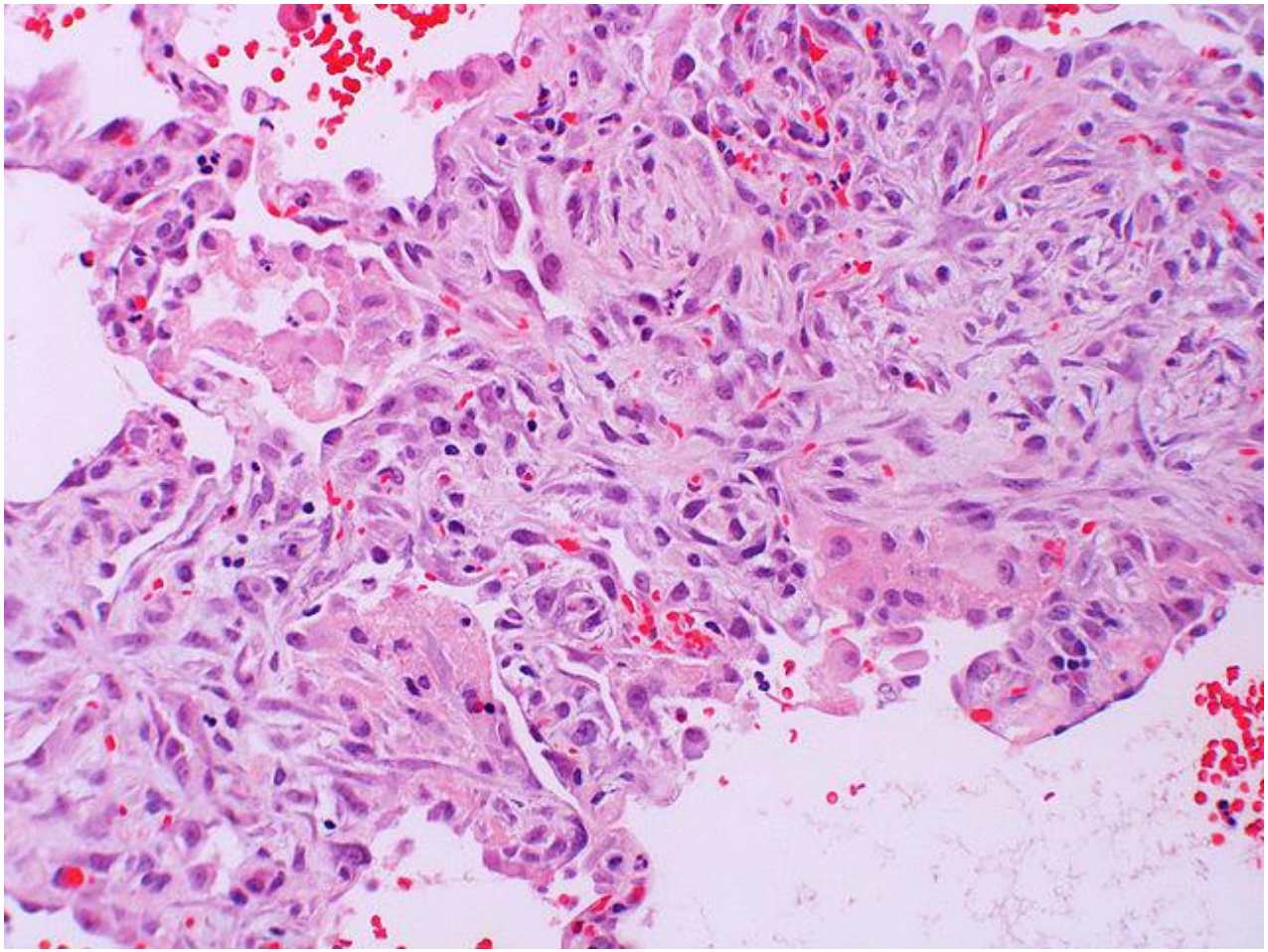
- MALES AFFECTED MORE THAN FEMALES.
- RADIOLOGY AND HISTOLOGY ....CHANGES KNOWN AS UIP = USUAL INTERSTITIAL PNEUMONIA.

UIP

- PATCHY INTERSTITIAL FIBROSIS.
- EARLY IN THE DISEASE: FIBROBLASTIC PROLIFERATION.
- THESES BECOME MORE COLLAGENOUS AND LESS CELLULAR.
- USUALLY EARLY AND LATE LESIONS COEXIST.

- THIS FIBROSIS CAUSES COLLAPSE OF ALVEOLAR WALLS AND FORMATION OF CYSTIC SPACES LINED BY TYPE 2 PNEUMOCYTES = HONYCOMB FIBROSIS.

UIP



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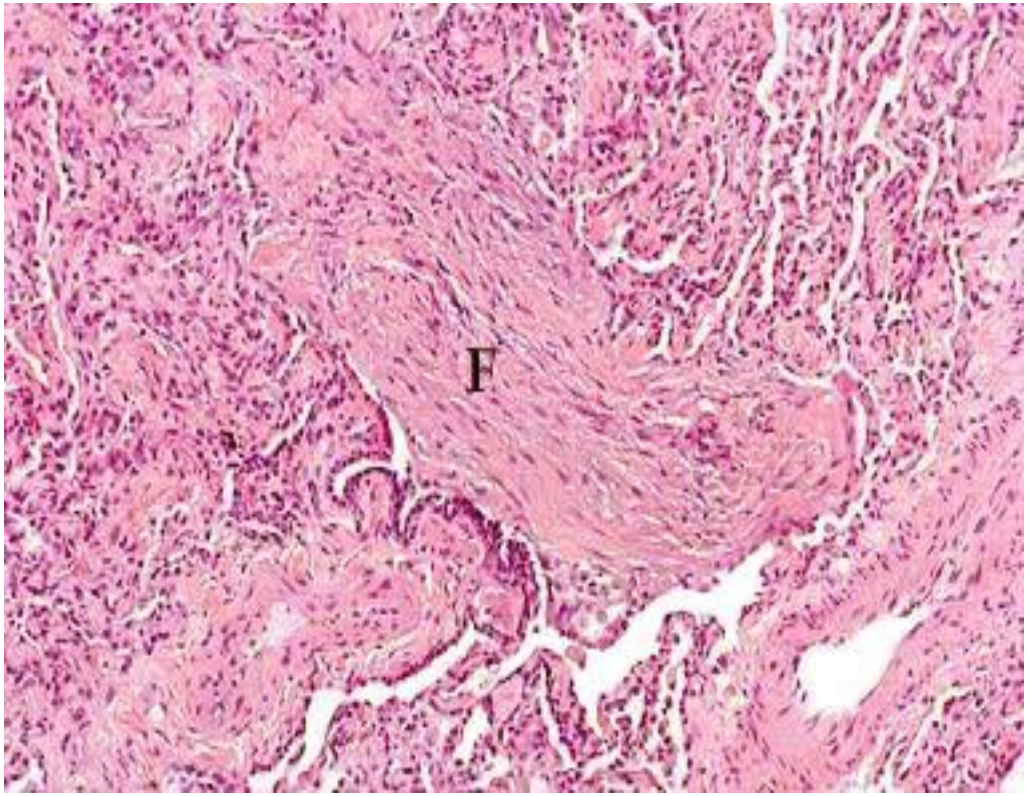
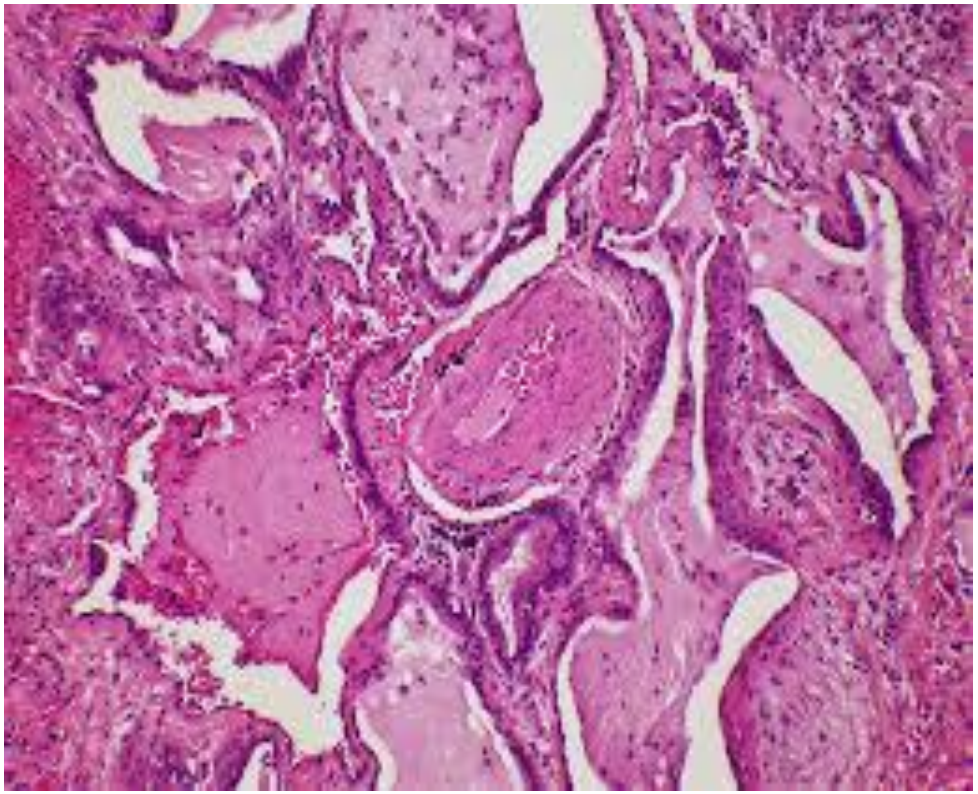


HONEYCOMB LUNG

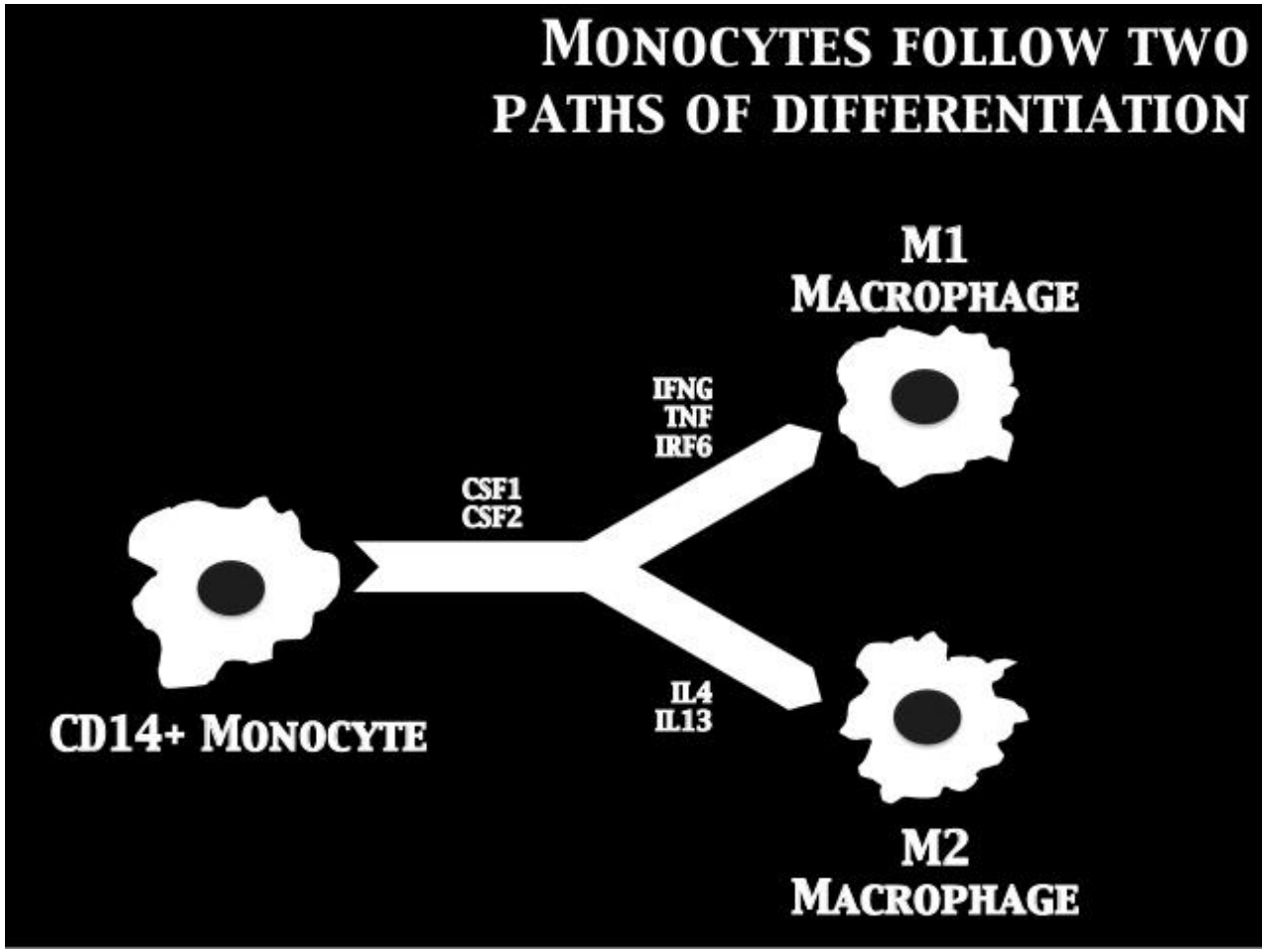
HONEYCOMB LUNG



PATHOGENESIS

- REPEATED CYCLES OF EPITHELIAL INJURY BY UNIDENTIFIED AGENT.
- INFLAMMATORY CELLS AND MEDIATORS PLAY A ROLE.
- M2 MACROPHAGES PROBABLY PLAY AN IMPORTANT ROLE.

M2


MONOCYTES FOLLOW TWO PATHS OF DIFFERENTIATION
M1 MACROPHAGE
IFNG
TNF
IRF6
CSF1
CSF2
CD14+ MONOCYTE
IL4
IL13
M2 MACROPHAGE

CLINICAL FEATURES

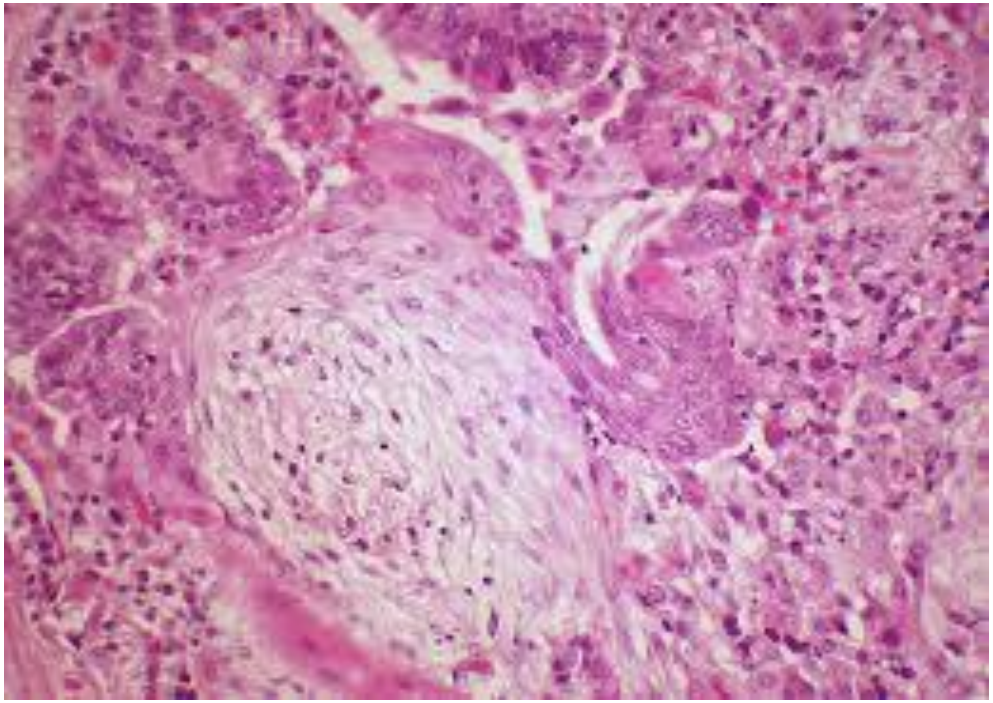
- GRADUAL ONSET OF NONPRODUCTIVE COUGH.
- PROGRESSIVE DYSPNEA.

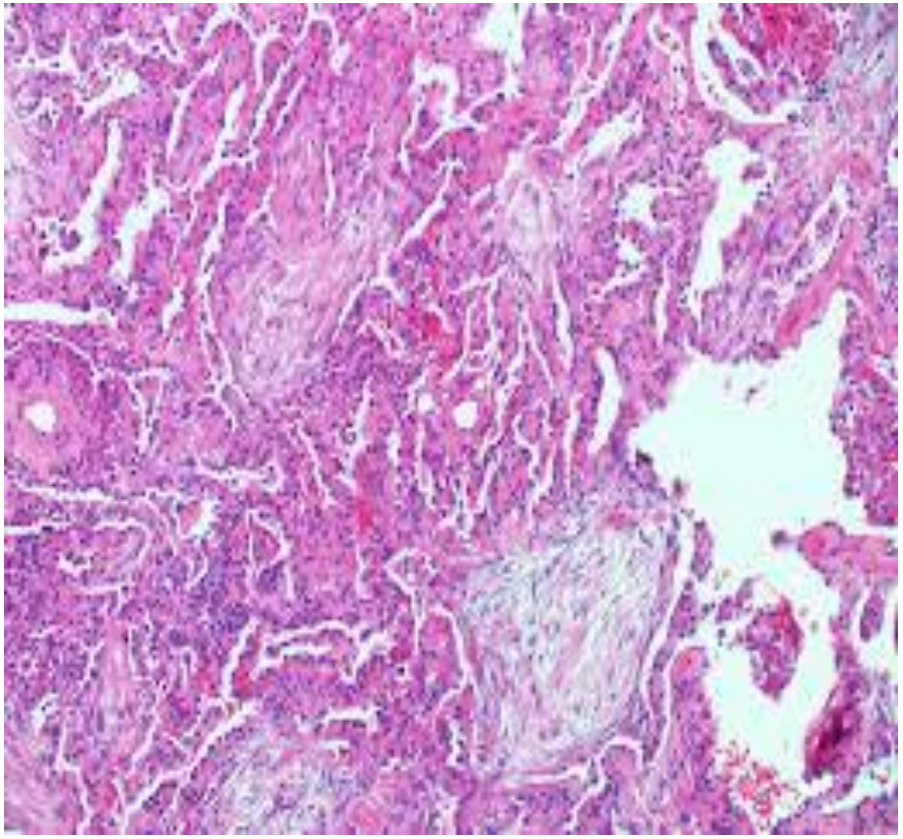
- MEAN SURVIVAL = 3 YEARS.
- LUNG TRANSPLANT IS THE ONLY DEFINITIVE THERAPY.

CRYPTOGENIC ORGANISING PNEUMONIA

- UNKNOWN ETIOLOGY.
- COUGH AND DYSPNEA.
- HISTOLOGICALLY: POLYPOID PLUGS OF LOOSE ORGANISING CONNECTIVE TISSUE -= MASSON BODIES
- UNDERLYING LUNG ARCHITECTURE NORMAL.
- CAN RECOVER SPONTANEOSLY OR NEED STEROIDS FOR 6 MONTHS OR LONGER.

MASSON BODIES





PNEUMOCONIOSES

- REACTION TO INHALATION OF MINERAL DUST.
- MOST COMMON: COAL, SILICA, ASBESTOS.

PATHOGENESIS

- REACTION OF LUNG TO MINERAL DUST DEPENDS ON:

  SIZE

  SHAPE

  SOLUBILITY

  REACTIVITY

SIZE

- PARTICLES > 5 MICROMETER ARE UNLIKELY TO REACH DISTAL AIRWAYS.
- < 0.5 MICROMETER MOVE IN AND OUT OF ALVEOLI WITHOUT BEING LODGED.

- 1- 5 MICRON...MOST DANGEROUS . THEY GET LODGED AT THE BIFURCATION OF DISTAL AIRWAYS.

REACTIVITY

- COAL IS INERT.. LARGE AMOUNT NEEDS TO BE DEPOSITED BEFORE BECOMING CLINICALLY SIGNIFICANT.
- SILICA AND ASBESTOS ARE MORE REACTIVE.

PATHOGENESIS

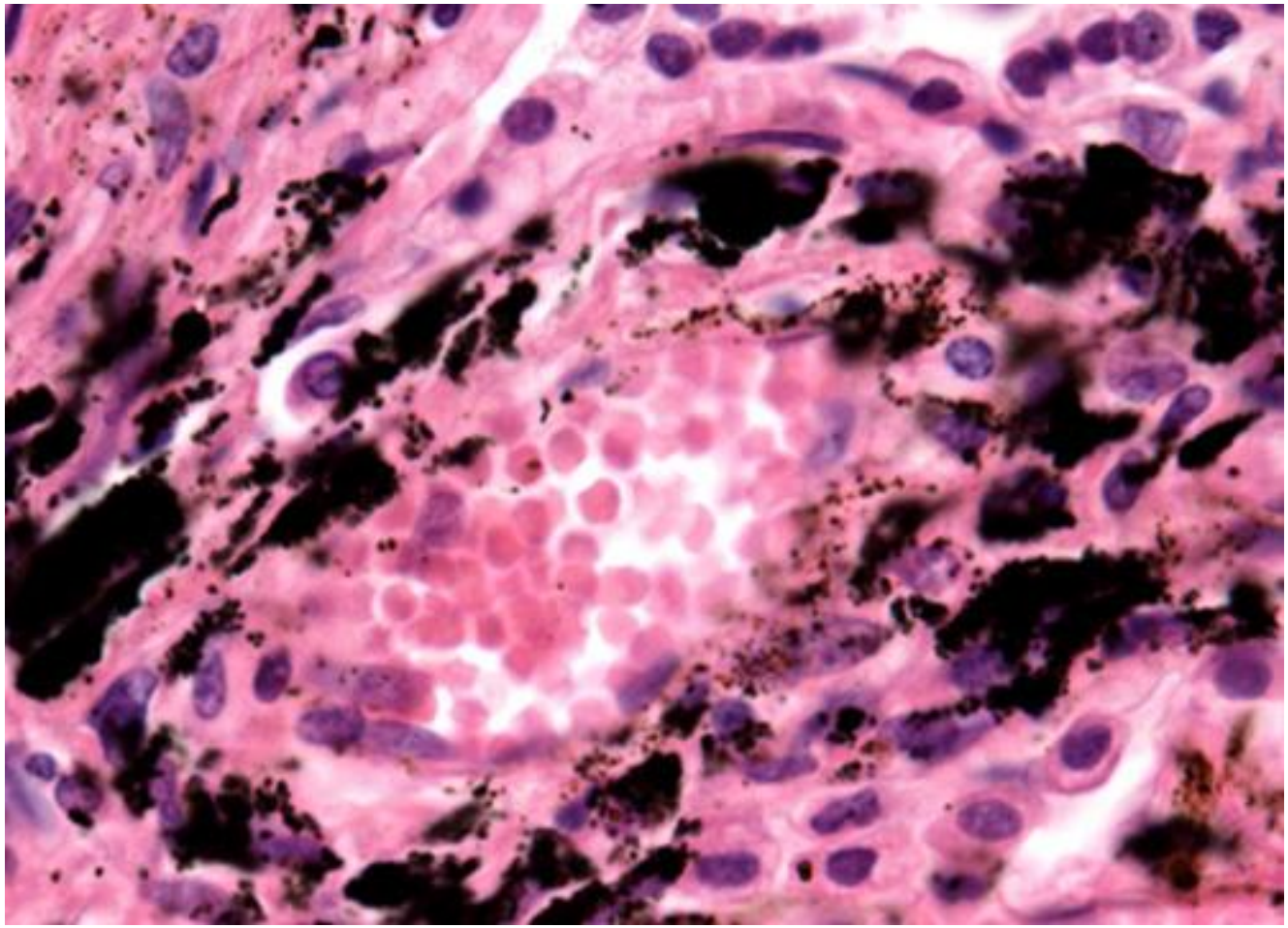
- WHEN PARTICLES ACCUMOLATE , ALVEOLAR MACROPHAGES ENGULF THEM AND CAUSE AN INFLAMMATORY RESPONSE RESULTING IN FIBROSIS.

COAL WORKER'S PNEUMOCONIOSIS

- MAINLY CARBON BUT ADMIXED WITH OTHER CHEMICALS
- CAN CAUSE: ASYMPTOMATIC ANTHRACOSIS.
- : SIMPLE CWP.
- : COMPLICATED CWP

ANTHRACOSIS

- CARBON ENGULFED BY MACROPHAGES.
- ASYMPTOMATIC.
- ALSO SEEN IN SMOKERS AND ALL URBAN DWELLERS.



SIMPLE PNEUMOCONIOSIS

- DUST LADEN MACROPHAGES AND DELICATE NETWORK OF COLLAGEN FIBERS.
- FORM COAL MACULES AND COAL NODULES.
- WITH TIME...EMPHYSEMA.

COMPLICATED CWP

- PROGRESSIVE MASSIVE FIBROSIS

- MULTIPLE SCARS.
- DENSE COLLAGEN AND PIGMENT.

CLINICAL FEATURES

- USUALLY BENIGN DISEASE WITH LITTLE EFFECT ON LUNG FUNCTION.
- PROGRESSIVE MASSIVE FIBROSIS... AFFECTS LUNG FUNCTION.
- NO INCRESED RISK OF CANCER.

SILICOSIS

- THE MOST COMMON CHRONIC OCCUPATIONAL DISEAE .
- INHALATION OF CRYSTALLINE SILICA.

SILICA

- CRYSTALLINE AND AMORPHOUS SILICA.
- CRYSTALLINE IS MORE TOXIC AND FIBRINOGENIC.
- QUARTZ IS MOSTLY IMPLICATED IN SILICOSIS.

- PURE QUARTZ IS MUCH MORE FIBRINOGENIC THAN IF IT IS MIXED WITH OTHER MINERALS.

- INGESTED SILICA CAUSES ACTIVATION OF MACROPHAGES AND RELEASE OF MEDIATORS.
- TNF IS IMPORTANT IN THE PATHOGENESIS, AS ANTI-TNF GIVEN TO MICE EXPOSED TO SILICA CAN BLOCK FIBROSIS.

MORPHOLOGY

- SILICOTIC NODULES: TINY DISCRETE PALE TO BLACKENED NODULES IN THE UPPER ZONES OF LUNGS.
- HISTOLOGICALLY: CONSINTRICALLY ARRANGED HYALINISED COLLAGEN FIBERS SURROUNDING AN AMORPHOUS CENTER.
- CAN PROGRESS TO PMF.

CLINICAL FEATURES

- RESPIRATORY SYMPTOMS USUALLY OCCUR WITH PMF.
- INCREASED SUSCEPTABILITY TO TB. SILICA DEPRESSES IMMUNITY AND IMPAIRS ABILITY OF MACROPHAGES TO PHAGOCYTOSE BACTERIA.
- RELATION TO LUNG CARCINOMA IS CONTROVERSIAL BUT SILICA IS THOGHT TO BE CARCINOGENIC IN HUMANS.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