

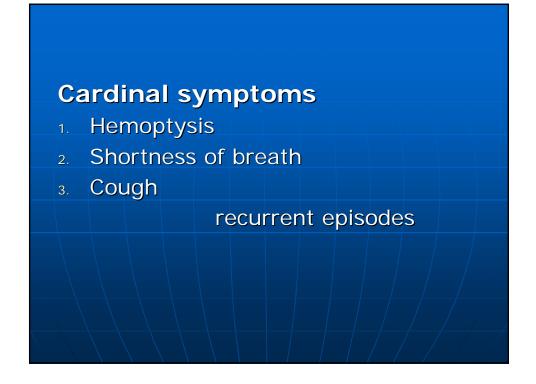
<u>Etiology</u>

With capillaritis

Wegener's granulomatosis Microscopic polyangitis Isolated pulmonary capillaritis Connective tissue disorder Primary antiphospholipid syndrome Mixed cryoglobulinemia Behcets syndrome Henoch scholein purpura Goodpasture's syndrome Pauci-immune glomerulonephritis Immune complex associated glomerulonephritis Drug induced Acute lung allograft rejection

Without capillaritis

Idiopathic pulmonary hemosiderosis Systemic lupus nephritis Good pasture's syndrome Diffuse alveolar damage Penicillamine Timellitic anhydride Mitral stenosis Coagulation disorder Pulmonary veno-occlusive disease Pulmonary capillary hemangiomatosis Lymphangioleiomyomatosis Tuberous sclerosis



Physical examination

Respiratory system: nonspecific Examination of ocular, nasopharynx, cutaneous evidence of vasculitis

+/- fever

investigation

<u>CXR</u>: alveolar infiltrate mostly diffuse + bilateral Pleural effusion rare

Blood investigation:

Low hemoglobin

Raised TLC

ANA/ANCA/Antiglomerular basement membrane antibodies

ESR

Histopathology

Alveolar hemorrhage

May be due to surgical trauma (could be differentiated by finding intra alveolar fibrin, hemosiderin in the alveolar walls and hemosiderin-laden macrophages

Capillaritis:

Fibrin thrombi in capillaries Fibrinoid necrosis of capillary walls Interstitial accumulation of fragmented neutrophils

Nuclear dust adjacent to capillaries

	anemia	Renal disease	cutaneous	arthritis
wegener	+	+	+	+
Micro PAN	+	+	+	+
SLE	+	+	+/-	+
Good pasture's	+	-	-	-
IPH	+	-	-	7
IPC	+		-	
HSP	+		+	+

	ANA	dsDNA	CL	RF	ANCA	ABMA	Tissue stain
WG	+/-	_	Ν	+-	+	-	granular
Mic PAN	+-	-	N	+-	+	-	-
SLE	+	+	low	+	_		granular
GP	-	-	Ν	-	_	÷	Linear
IPH	-	_	Ν	_	_	-	-
IPC		-	N	_	+-	-	-
HSP	-	-	Ν	-	_ /	- / /	granular

Wegener's granulomatosis

Sytemic vasculitis in middle aged adults with necrotizing granulomas in upper and lower respiratory tract cANCA + Focal segmental necrotizing GN DAH with capillaritis(subacute and recurrent) Treated with Corticosteroids and cyclophosphamide Newer agent are IVIG, Cotrimox, Antilymphocyte monoclonal antibodies, tumor necrosis factor inhibitor

Microscopic polyangitis

Small vessel variant of PAN p ANCA + Focal segmental necrotizing GN DAH with capillaritis is common Treated with corticosteroid + cyclophosphamide /azathioprine Short term mortality is 25% 5 yr survival rate is >60%

Good pasture's syndrome

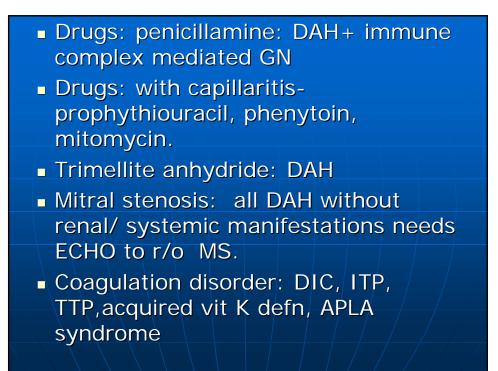
- DAH + GN + ABMA in serum / tissue
- Young smoker
- HLA B7 and HLA DR w2 severe renal disease and poor prognosis.
- Treated with corticosteroids/cyclophosphamide/az athioprine/plasmapheresis/ MMF/ anti CD20
- Predictors of response %of glomerular involvement + renal insufficiency

Collagen vascular disease

- SLE
- <2% of SLE patients have DAH Has to diff. from lupus pneumonitis Mortality of SLE with DAH is 50%
- Rheumatoid arthritis
- Scleroderma
- MCTD
- polymyositis

Idiopathic pulmonary hemosiderosis

- Young children and adults
- Caused: ?linked to Stachybotrys atra ?immune mediated
 20% pediatric patients have LNE and hepatosplenomegaly.
 Diagnosis of exclusion (needs lung biopsy to prove as bland DAH)
 Corticosteroid / azathioprine
 Lung transplant



Mixed cryoglobulinemia:

Purpura+arthritis+hepatitis+GN A/w Hepatitis B / C infections

Leucocyclastic vasculitis.

Behcets syndrome:

5-10% have lung manifestations small vessel vasculitis

Lymphangioleomyomatosis: premenopausal women proliferation of smooth muscle walls of the pulmonary lymphatics. chylothorax. pneumothorax- 40% hemoptysis: 40% focal (DAH is rare) Tuberous sclerosis:

1.mutations in TSC1 and TSC 2 gene

- 2.triad of mental retardation,
- epilepsy, derma angiofibroma.
- 3.lung involvement -1%

4.death due to neurological complication.