MD seminar

Diffuse alveolar hemorrhage

Definition

- Medical emergency
- Pulmonary hemorrhage originating from pulmonary microcirculation (arterioles, capillaries, venules)
- @ intrapulmonary hemorrhage/pulamonary alveolar hemorrhage/ pulmonary capillary hemorrhage/ microvascular lung hemorrhage.

Etiology

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

Cardinal symptoms

1. Hemoptysis
2. Shortness of breath
3. Cough

recurrent episodes

Physical examination

Respiratory system: nonspecific

Examination of ocular, nasopharynx, cutaneous evidence of vasculitis

+/- fever

investigation

CXR: 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	+	-	-	-
IPC	+	-	-	-
HSP	+	+	+	+

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

- Drugs: penicillamine: DAH+ immune complex mediated GN
- Drugs: with capillaritis- prophythiouracil, phenytoin, mitomycin.
- Trimellite anhydride: DAH
- Mitral stenosis: all DAH without renal/ systemic manifestations needs ECHO to r/o MS.
- Coagulation disorder: DIC, ITP, TTP,acquired vit K defn, APLA syndrome

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.