

27/2/51



14/3/51

Systemic vasculitis (Pulmonary renal syndrome)



- **Immune complex disease: SLE, Idiopathic crescentic immune complex GN**
- **Goodpasture disease**
- **Microscopic polyangiitis**
- **Wegener granulomatosis**
- **Churg-Strauss syndrome**

Feature	SLE	Goodpasture disease	Microscopic Polyangiitis	Wegener's granulomatosis	Churg-strauss syndrome
Small-vessel vasculitis	+	+	+	+	+
Signs/ symptoms	SLE	PR	PR	Sinusitis/ OM	Allergic phage/Asthma
Necrotizing granulomas	-	-	-	+	+
ANCA	-	-	p-ANCA	c-ANCA	p-ANCA
Anti-PR3	-	-	-	+	-
Anti-MPO	-	-	+	-	+
C3/C4	L	N	N	H	N
Anti-GBM Ab	-	+	-	-	-
ANA/ Immunologic	Anti-Sm, Anti ds-DNA, Anticardiolipin	-	-	-	+
Renal Bx	GN	IgG+C3	Pauciimmune segmental necrotising GN	Pauciimmune segmental necrotising GN	Focal segmental GN

Differential diagnosis: small-vessel vasculitis



Feature	HSP	Cryoglobulinemic vasculitis	Microscopic Polyangiitis	Wegener's granulomatosis	Churg-strauss syndrome
S/S of small-vessel vasculitis	+	+	+	+	+
IgA-dominant immune deposits	+	-	-	-	-
Cryoglobulins in blood vessels	-	+	-	-	-
ANCA in blood	-	-	+	+	+
Necrotizing granulomas	-	-	-	+	+
Asthma and eosinophilia	-	-	-	-	+

Refractory pulmonary hemorrhage



■ Immunosuppressive therapy

Cyclophosphamide, Methotrexate

Azathiopine

Jara LJ. *Curr Rheumatol Rep.* 2003 Apr;5(2):107-15.

Badcha H. *Semin Arthritis Rheum.* 2004 Jun;33(6):414-21.

■ Plasma exchange and plasmapheresis

**Report in : Anti-GBM disease, Lupus,
Wegener's granulomatosis
Microscopic polyangiitis**

Gallagher H. *Am J Kidney Dis.* 2002 Jan;39(1):42-7.

Barile LA. *Lupus.* 1997;6(5):445-8.

McCarthy LJ. *J Clin Apher.* 1994;9(2):116-9.

Refractory pulmonary hemorrhage



■ Intravenous immunoglobulin (IVIg)

O'riodan A. Ren Fail. 2005;27(6):791-4.

Sherer Y. Autoimmun Rev. 2006 Feb;5(2):153-5.

■ Rituximab

Hermle T. Clin Nephrol. 2007 Nov;68(5):322-6.

Walsh M. Kidney Int. 2007 Sep;72(6):676-82.

■ Aminocaproic acid

Wanko SO. Biol Bloo Marrow transplant 2006;12:949-56.

■ Recombinant factor VIIa

Carriollo-Esper R. Gac Med Mex. 2007 Jan-Feb;143(1):83-6.

Betensley AD. Am J Respir Crit Care Med 2002; 166:1291.

Pulmonary hemorrhage: When will CXR be clear?



- **Retrospective review of DAH**
- **Consolidations or ground-glass opacities were identified mainly in the middle fields(73%)**
- **Complete follow-up showed consolidation opacities in 36/60 zones, which persisted in 16/60 after 7 days and in 11/60 after 15 days**
- **After 7 days - partial regression of consolidation opacities**
- **After 15 days - markedly diminished**

Infection in immunocompromised host



- **Cardiopulmonary involvement in pediatric SLE: a twenty-year retrospective analysis**
- National Taiwan University Hospital

J Microbiol Immunol Infect.2007;40:525-31.

Infection in immunocompromised host



- Pneumonia: most common pulmonary manifestation 36.9%
- *Pseudomonas aeruginosa*: most common pathogen 14%
- *Mycoplasma*, *Aspergillus* and tuberculosis
- Others: *Stenotrophomonas maltophilia*
Klebsiella pneumoniae
Staphylococcus aureus
Candida spp.
Pneumocystis carinii
Cytomegalovirus

Infection in immunocompromised host



- **Community-acquired pneumonia in Thai patients with SLE**
- Retrospective chart review
- SLE patients, age >16 years
- Division of Rheumatology, Chiang Mai University

Southeast Asian J Trop Med Public Health. 2007 May;38(3):528-36.

Infection in immunocompromised host



- Mycobacterium tuberculosis 30%
- Nocardia spp. 15%
- Aspergillus spp. 12.5%
- Staphylococcus aureus 7.5%
- Pneumocystis carinii 5%
- Haemophilus influenzae 5%
- Escherichia coli 5%
- Pseudomonas aeruginosa 5%
- Others : Acinetobacter baumannii, Burkholderia pseudomallei, and Strongyloides stercoralis

Diagnosis of CMV pneumonitis



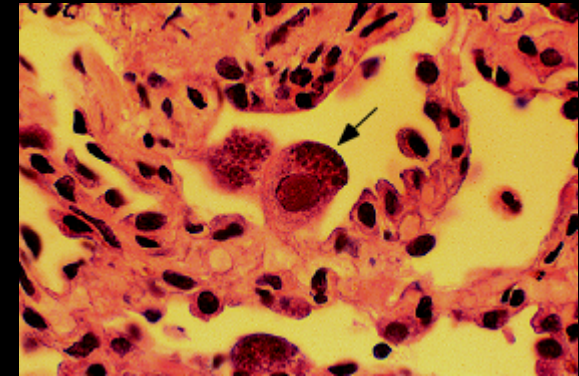
- Positive CMV culture of bronchoalveolar lavage fluid (BAL)
 - Not established as either a sensitive or a specific test for CMV pneumonitis (sensitivity 50% and very low specificity)

Mann M. Am J Respir Crit Care Med. 1997 May;155(5):1723-8.

Diagnosis of CMV pneumonitis



■ **Transbronchial or surgical lung biopsy demonstrating pneumonitis and cytopathic effect consistent with CMV**



■ **Likely to be more specific for CMV pneumonitis, but are almost certainly insensitive due to the patchy nature of the disease**

Rodriguez-Barradas MC. Clin Infect Dis. 1996 Jul;23(1):76-81.

Dammy M. Arch Pediatr. 1994 Feb;1(2):137-42.

Sommer SE. Chest. 1991 Sep;100(3):856-8.

Diagnosis of CMV pneumonitis



- Evidence of CMV antigens or typical cytopathic changes induced by CMV seen on cytologic examination of BAL fluid cells.
 - CMV PCR (Blood/BAL)
 - Quantitative CMV DNA assay/ Real time PCR (sensitivity 90.9%, specificity 20%)
 - BAL viral load > 500,000 copies/ml

Lawrence D. Curr Opin Infect Dis 2007;20:408-411.

Diagnosis of CMV pneumonitis



- Evidence of CMV antigens or typical cytopathic changes induced by CMV seen on cytologic examination of BAL fluid cells.
 - Combining PCR with antibody methods may improve both specificity and positive predictive

Value.Cathomas G. Blood 1993;81:1909-14.

- pp65 Antigen (Blood)

Drew WL. Curr Opin Infect Dis. 2007 ;20(4):408-11.

Honda J. Scand J Infect Dis. 2001;33(9):702-5.

CMV: When to be treated?



- No available guidelines in medical literature on the use of antiviral agents for the management of symptomatic CMV infection
- No data issued from clinical trials
- **Observation reported** indicate that treatment seems worth to be used in chronic inflammatory disease treated by immunosuppressive agents.

CMV: choice of treatment?



- Ganciclovir (5 mg/kg IV every 12 hours)
: considered the drug of choice
- Foscarnet is an alternative therapy
- Intravenous immunoglobulin therapy or
CMV hyperimmune globulin



8/1/51



10/1/51



10/1/51



13/1/51



7/2/51: CT ratio 0.61



9/2/51: CT ratio 0.54

T=0