Eosinophilic Granulomatosis with Polyangiitis (Churg-Strauss Syndrome)

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First Description

- In 1951, **Churg and Strauss** first described the syndrome in 13 patients who had:

1. asthma,
2. eosinophilia,
3. granulomatous inflammation,
4. necrotizing systemic vasculitis, and
5. necrotizing glomerulonephritis

Introduction

- In 2012 : the name of ***(Churg-Strauss Syndrome)*** has been replaced with **eosinophilic granulomatosis with polyangiitis (EGPA)** with the aim of focusing on the histopathology of the disease.
- EGPA is a rare systemic necrotizing vasculitis that affects **small-to-medium-sized vessels** and is ass. with severe **asthma** & peripheral blood and tissue **eosinophilia**.

[Guideline] Groh M, Pagnoux C, Baldini C, Bel E, Bottero P, Cottin V, et al. Eosinophilic granulomatosis with polyangiitis (Churg-Strauss) (EGPA) Consensus Task Force recommendations for evaluation and management. Eur J Intern Med. 2015 Sep. 26 (7):545-53

Introduction (cont.)

- Like granulomatosis with polyangiitis (Wegener granulomatosis), and the microscopic polyangiitis , EGPA is an **antineutrophil cytoplasmic antibody (ANCA)–associated vasculitis**
- It is **characterized** by extravascular necrotizing granulomas (usually rich in eosinophils), eosinophilia, and tissue infiltration by eosinophils.
- **However**, these abnormalities do not always coexist.

Keogh KA, Specks U. Churg-Strauss syndrome: clinical presentation, antineutrophil cytoplasmic antibodies, and leukotriene receptor antagonists. Am J Med. 2003 Sep. 115(4):284-90

Introduction (cont.)

- EGPA is considered a disease that situated between **primary systemic vasculitis** (small-vessel vasculitis associated with (ANCAs) & **hypereosinophilic disorders** .
- **Any organ** can be affected, but the lungs, skin, sinuses, cardiovascular system, kidneys, peripheral nervous system, CNS, joints, and GI tract are most commonly affected.

Simon H.U., Rothenberg M.E., Bochner B.S., Weller P.F., Wardlaw A.J., Wechsler M.E. Refining the definition of hypereosinophilic syndrome. J. Allergy Clin. Immunol. 2010 Jul;126(1):45–49

Pathophysiology

- EGPA is a granulomatous small-vessel vasculitis.
- The cause of this allergic angiitis and granulomatosis is **unknown**.
- No data have been reported regarding the role of immune complexes or cell-mediated mechanisms in this disease, although autoimmunity is evident with the presence of **hypergammaglobulinemia, increased levels of immunoglobulin E (IgE), rheumatoid factor, and ANCA.**

Hellmich B, Ehlers S, Csernok E, Gross WL. Update on the pathogenesis of Churg-Strauss syndrome. Clin Exp Rheumatol. 2003 Nov-Dec. 21(6 Suppl 32):S69-77

Genetics

- EGPA is **not** inherited or genetic disease.
- Familial form is extremely **rare**
- **HLA-DRB4** positivity may be a genetic risk factor for the development of EGPA , and may increase the likelihood of vasculitic manifestations of the disease.

Vaglio A, Martorana D, Maggiore U, Grasselli C, Zanetti A, Pesci A, et al. HLA-DRB4 as a genetic risk factor for Churg-Strauss syndrome. Arthritis Rheum. 2007 Sep. 56(9):3159-66

Epidemiology

- **Frequency** : incidence in United States is 1-3 cases per 100,000 adults per year. The international incidence is approximately 2.5 cases per 100,000 adults per year
- **Mortality/Morbidity** :The principal causes are myocarditis and myocardial infarction secondary to coronary arteritis. With treatment, the 1-year survival rate is 90% and the 5-year survival rate is 62%

Keogh KA, Specks U. Churg-Strauss syndrome: update on clinical, laboratory and therapeutic aspects. Sarcoidosis Vasc Diffuse Lung Dis. 2006 Mar. 23(1):3-12.

Sex :more common in males than in females

Age :from 15-70 years , The mean age at diagnosis is around 50 years.

- But the disease can occur at any age
- EGPA in pediatric patients is well described, but mostly as case reports

Boyer D, Vargas SO, Slattery D, Rivera-Sanchez YM, Colin AA. Churg-Strauss syndrome in children: a clinical and pathologic review. Pediatrics. 2006 Sep. 118(3):e914-20

causes

- Causes of EGPA are **unknown**.
- EGPA is possibly an **allergic** or **autoimmune** reaction to an environmental agent or drug
- Several case reports have described drug-induced forms of EGPA. **Mesalazine-induced** EGPA has been reported in a patient with Crohn disease

Sharma A., Dogra S., Sharma K. Granulomatous vasculitis. Dermatol. Clin. 2015 Jul;33(3):475–487.

Possible risk factors

- History of asthma or nasal problems.
- Most patients diagnosed with EGPA have a history of nasal allergies, chronic sinusitis or asthma, which is often severe or hard to control

Moosig F. Eosinophilic granulomatosis with polyangiitis: future therapies. Presse Med. 2013 Apr;42(4 Pt 2):510–512.

Clinical picture

- The syndrome has **3 phases or stages** :

1. **Prodromal or allergic stage**
2. **Eosinophilic infiltrative disease** (eg, eosinophilic pneumonia or eosinophilic gastroenteritis)
3. **Systemic medium- and small-vessel vasculitis** with granulomatous inflammation : Systemic symptoms & life-threatening vasculitis occur in this phase.

- **However**, the phases do not necessarily follow one another consecutively, and may overlap

Pagnoux C, Guillevin L. Churg-Strauss syndrome: evidence for disease subtypes? Curr Opin Rheumatol 2010; 22:21

Prodromal or Allergic stage

- The prodromal stage is characterized by allergy.
- Almost **all patients** experience **asthma** and/or **allergic rhinitis** which may require systemic corticosteroid treatment
- The allergic rhinitis may produce symptoms such as **rhinorrhea** and **nasal obstruction**, and the formation of nasal **polyps** that require surgical removal, often more than once.
- **Sinusitis** may also be present.
- This stage may persist for **years**

"*Churg-Strauss syndrome: clinical and serological features of 19 patients from a single Italian centre". Rheumatology. Oxford. 41 (11): 1286–94. doi:10.1093/rheumatology/41.11.1286. PMID 12422002 (2002)*

Eosinophilic stage

- Characterized by an abnormally **high level** of **eosinophils** in the blood and tissues.
- The symptoms of hypereosinophilia depend on which part of the body is affected, but mostly cause **eosinophilic pneumonia or gastroenteritis**
- **Symptoms** may include weight loss, Fever, asthma, cough, abdominal pain, and gastrointestinal bleeding.
- This stage can last **months or years.**

Jennette JC, Falk RJ, Andrassy K, et al. Nomenclature of systemic vasculitides. Proposal of an international consensus conference. Arthritis Rheum. 1994 Feb. 37(2):187-92

Vasculitic stage

- The third & final stage, is **vasculitis**, lead to reduction of blood flow to various organs.
- **Blood clots** may develop within the damaged arteries in severe cases, particularly in arteries of the abdominal region
- Many patients experience severe abdominal pain ; due to **peritonitis** and/or **ulcerations** and **perforations** of the gastrointestinal tract, but occasionally due to **acalculous cholecystitis** or granulomatous **appendicitis** or **pancreatitis**

Churg A. Pulmonary angiitis and granulomatosis revisited. Hum Pathol. 2002 Oct. 14(10):868-83

Vasculitic stage (cont.)

- The most serious complication of this stage is **heart disease**, which is the main cause of death
- **Kidney** complications have been reported as being less common than other types of vasculitis
- The vasculitic phase usually develops **within 3 years** after the onset of asthma, although it may be delayed for several decades
- ttt of asthma with corticosteroid **mask** other features of the syndrome & delay the diagnosis

"Churg-Strauss syndrome: clinical and serological features of 19 patients from a single Italian centre". Rheumatology. Oxford. 41 (11): 1286 doi:10.1093/rheumatology/41.11.1286. PMID 12422002 (2002)

symptoms

- **Constitutional symptoms** : Malaise, fatigue, wt loss (70%), fever (57%), myalgias (52%)
- **Asthma** in 97% of pts , may precede vasculitis by up to 10 years or, less frequently, may coincide with the appearance of vasculitis.
- Asthma, often with onset during adulthood,& tends to be severe &corticosteroid-dependent

Sinico R.A., Bottero P. Churg-Strauss angiitis. Best. Pract. Res. Clin. Rheumatol. 2009 Jun;23(3):355–366.

Symptoms (cont.)

- Peripheral neuropathy - **Mononeuritis multiplex** (most frequent form, in about 77% of pts), it is the second most common manifestation, it cause pain & numbness in extremities
- **Upper respiratory manifestation** :Allergic rhinitis, Paranasal sinusitis (60%) & recurrent nasal polyposis (50 %).
- **Lower respiratory system manifestation** or Pulmonary symptoms (37%), including cough and hemoptysis due to alveolar hemorrhage

Moosig F. Eosinophilic granulomatosis with polyangiitis: future therapies. Presse Med. 2013 Apr;42(4 Pt 2):510–512

Symptoms (cont.)

- **Arthralgias or arthritis (40%)**
- **Skin manifestations** (49%) Purpura - Skin nodules, urticarial rash, necrotic bulla & digital ischemia
- **Cardiac manifestations** (14%)- Symptoms related to heart failure, myocarditis, pericarditis, constrictive pericarditis, and myocardial infarction
- **Gastrointestinal (GI) manifestations** (31%) - Symptoms related to GI vasculitis, eosinophilic gastritis, or colitis; these include abdominal pain , diarrhea & GI bleeding

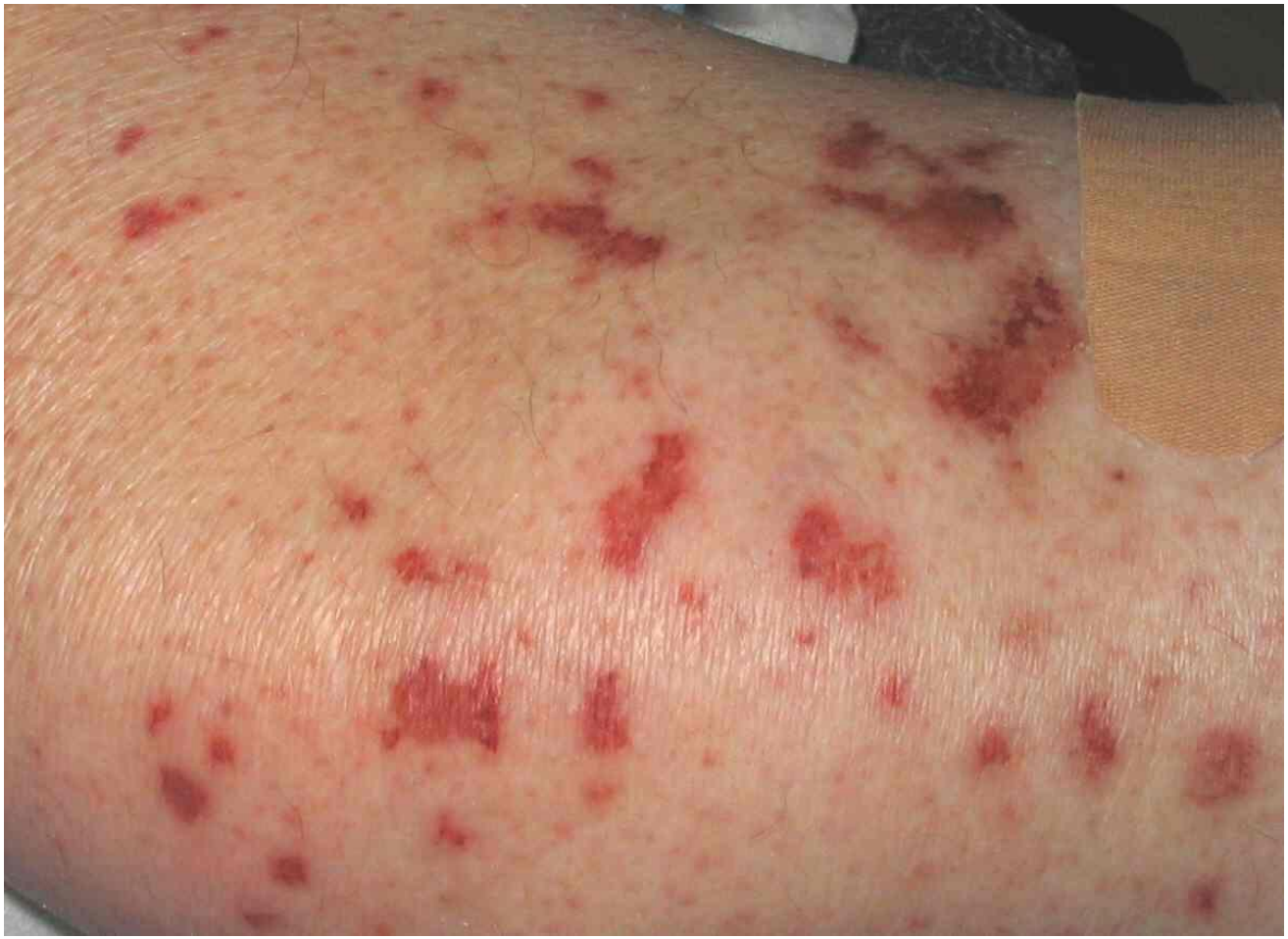
Baldini C., Talarico R., Della Rossa A., Bombardieri R. Clinical manifestations and treatment of Churg–Strauss syndrome. Rheum. Dis. Clin. North Am. 2010 Aug;36(3):527–543

Symptoms (cont.)

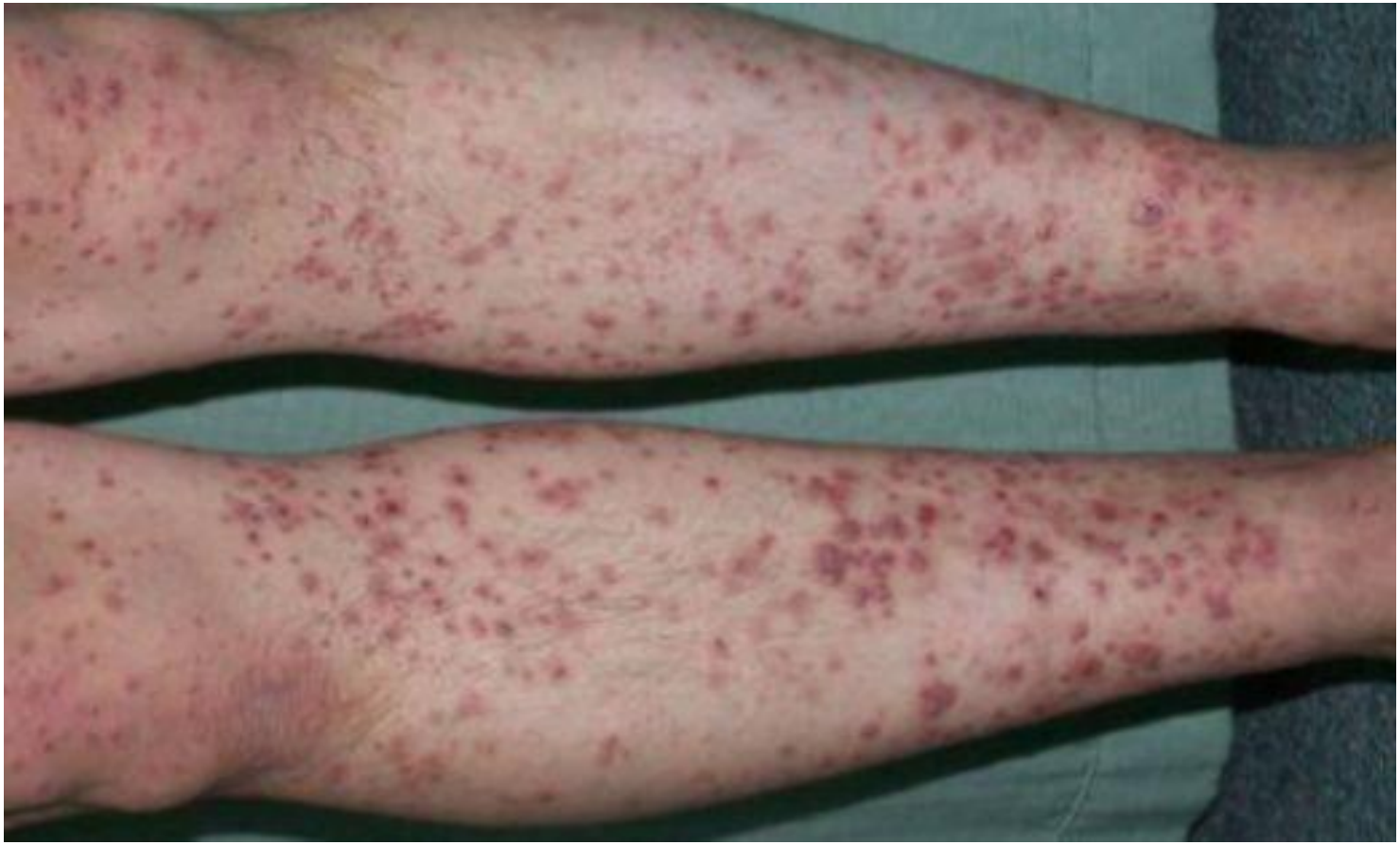
- **Renal symptoms**: as hypertension and uraemia is less common than in other vasculitic disorders associated with ANCA.
- **Less frequent symptoms** - Symptoms related to stroke (5 %) , fits, and coma , ophthalmologic involvement, and others
- Presence of renal, cardiac, or neurologic involvement indicates a **worse prognosis**

Woolnough K., Wardlaw A. Eosinophilia in pulmonary disorders. Immunol. Allergy Clin. N. Am. 2015 Aug;35(3):477492

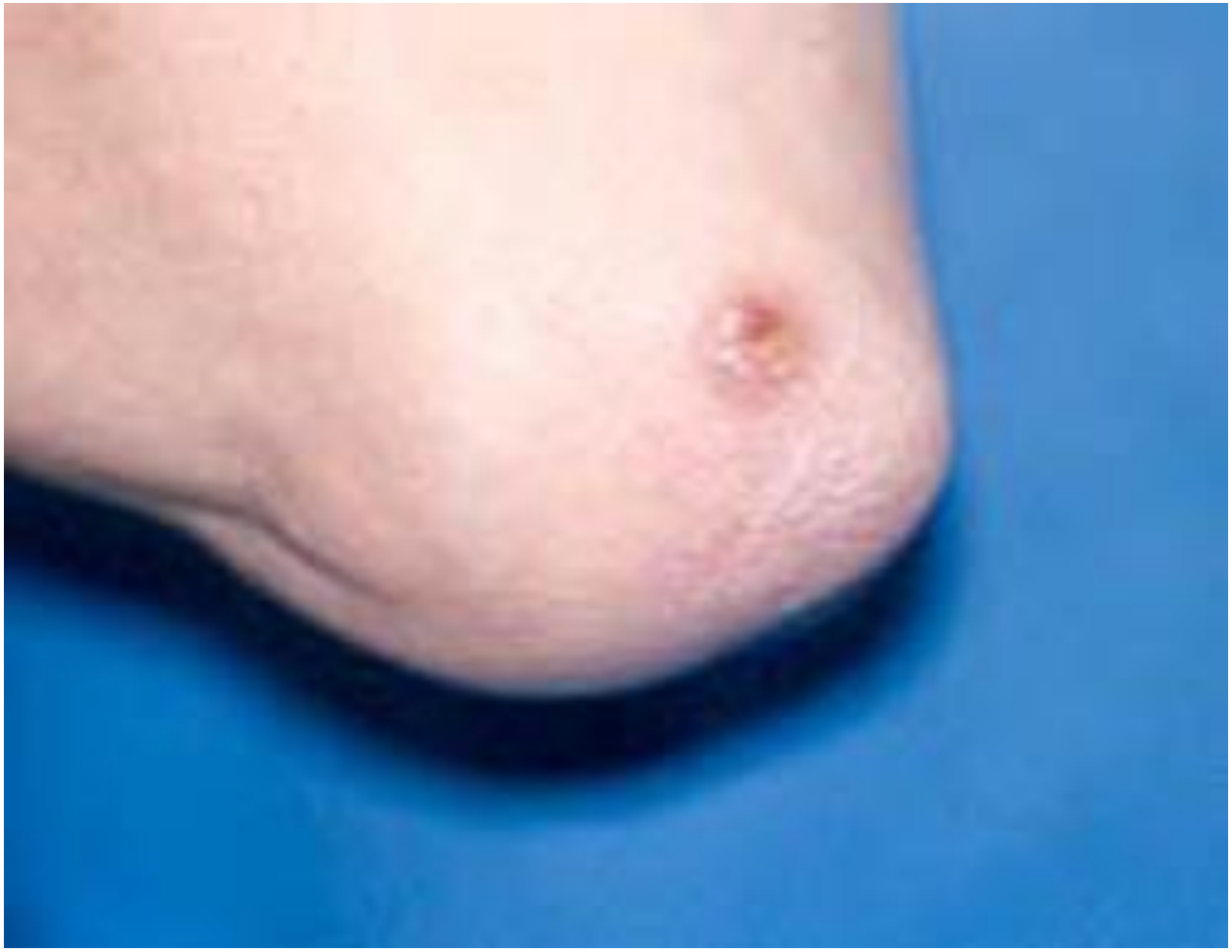
Skin rashes of EGPA



Skin rashes of EGPA



Sucutanous nodule in pt with EGPA



Differential Diagnoses

- **Other eosinophilic disorders** :

1. Parasitic infections,
2. Drug reactions .
3. Hypereosinophilic syndrome
4. Allergic bronchopulmonary aspergillosis
5. Acute eosinophilic pneumonia and chronic eosinophilic pneumonia

- **Other ANCA associated vasculitis**

Hypereosinophilic Syndrome(HES)

- **Characterized** by:

1- Marked and persistent (more than 6 month) eosinophilia (>1500 cells/µL),

2- Signs and symptoms of organ involvement

3- No identifiable cause of eosinophilia (as parasitic & viral infections, drugs, allergies, tumors, autoimmune or immunological dis)

Gotlib, J. World Health Organization-defined eosinophilic disorders: 2012 update on diagnosis, risk stratification, and management. Am. J. Hematol. 2012, 87, 903–914.

Hypereosinophilic Syndrome (cont.)

- **Four Groups of HES:**

(a) myeloid and lymphoid neoplasms with eosinophilia

(b) chronic eosinophilic leukemia

(c) lymphocytic HES

(d) idiopathic HES

Hypereosinophilic Syndrome (cont.)

- **Idiopathic HES** may substantially overlap with EGPA since cardiac & lung manifestations may be similar, but pts with idiopathic HES **rarely** present either asthma or nasal polyps , or vasculitic complications (e.g., purpura, glomerulonephritis, neuropathy)
- In Idiopathic HES tissue biopsies **do not** show vasculitis, and **ANCA** are typically **negative**

Khoury, P.; Zagallo, P.;Klion, A.D. Serum biomarkers are similar in Churg-Strauss syndrome and hypereosinophilic syndrome. Allergy 2012, 67, 1149–1156.

Allergic bronchopulmonary aspergillosis (ABPA)

- ABPA is a **fungal** infection simulates an EGPA but **limited** to the respiratory tract (asthma , CXR infiltrate & peripheral blood eosinophilia)
- **Diagnosis** :

1. Isolation of Aspergillus in bronchoalveolar lavage (BAL) or sputum, and
2. Elevated serum levels of specific IgE to Aspergillus .

Sinico, R.A.; P. Churg-Strauss angiitis. Best Pract. Res. Clin. Rheumatol. 2009, 23, 355–366.

Acute Eosinophilic Pneumonia

- **Causes** :

1. Idiopathic
2. Toxic inhalation & recent smoking
3. Drugs as amiodarone
4. Infection as HIV & candida

- **Clinically**: fever , cough and respiratory failure, **without** asthma or other systemic symptoms
- **Investigation** :pulmonary infiltrates and a BAL fluid rich in eosinophils. **NO** peripheral eosinophilia **NO** ANCA
- **TTT :** high dose of steroid & some times ventilation

Wechsler, M.E. Pulmonary eosinophilic syndromes. Immunol. Allergy Clin. N. Am 2007, 27,477–492.

Chronic Eosinophilic Pneumonia

- **Causes**

1. Idiopathic (most common)
2. Toxic inhalation
3. Drugs as antibiotic and amiodarone
4. Infection : parasite & fungal as aspergillus

- **Clinically** : fever, cough, asthma 50%, allergic rhinitis 50% and systemic symptoms such as weight loss & night sweats
- **Investigation** : peripheral eosinophilia but **absence** of ANCA & other organ manifestations can help differentiate this from EGPA.
- **TTT** corticosteroid 30-40 mg and tappering gradually

Wechsler, M.E. Pulmonary eosinophilic syndromes. Immunol. Allergy Clin. N. Am 2007, 27,477–492.

Other ANCA associated Vasculitis

- **Granulomatosis with Polyangiitis** : cavitated pulmonary nodules, nasal crusts and sinus bone erosions, more renal affection & c-ANCA positive (90%) NO eosinophilia or asthma
- **The Microscopic polyangiitis**, rarely shows high eosinophilia and rare involvement of the upper airway while its renal complications are often more severe than in EGPA , associated with p-ANCA (80%)

Workup of EGPA

- Testing **aims** to **establish** the diagnosis and the **extent** of organ involvement & to **distinguish** EGPA from other eosinophilic disorders.
- Diagnosis of EGPA is suggested by **clinical** findings and results of routine **laboratory** tests but should usually be confirmed by **biopsy** of lung or other affected tissue

Gotlib J. World Health Organization-defined eosinophilic disorders: 2014 update on diagnosis, risk stratification, and management. Am. J. Hematol. 2014 Mar;89(3):325–337

Guidelines of Workup of EGPA

- **European guidelines from the EGPA Consensus Task Force (2015) recommend the following**:

1. Serologic testing for toxocariasis and HIV
2. Specific IgE and IgG for Aspergillus species
3. Testing for Aspergillus spp. in sputum, bronchoalveolar lavage fluid, or both
4. Peripheral blood smear (for dysplastic eosinophils or blasts)
5. ANCA with (ELISA)
6. Chest computed tomography (CT) scan

Groh M,Cottin V, et al. Eosinophilic granulomatosis with polyangiitis (Churg-Strauss) Consensus Task Force recommendations for evaluation and management. Eur J Intern Med. 2015 Sep. 26 (7):545-53

Guidelines of Workup of EGPA

- **Additional investigations** should be guided by patient-specific clinical findings.
- **The EGPA Consensus Task Force encourages obtaining biopsies** from pts with suspected EGPA.
- ***In the correct clinical context*** (asthma with eosinophilia or systemic manifestations, or eosinophilia with extrapulmonary disease), a biopsy showing small- or medium-vessel vasculitis, **strongly** supports a diagnosis of EGPA.

Groh M,Cottin V, et al. Eosinophilic granulomatosis with polyangiitis (Churg-Strauss) Consensus Task Force recommendations for evaluation and management. Eur J Intern Med. 2015 Sep. 26 (7):545-53

Laboratory Studies

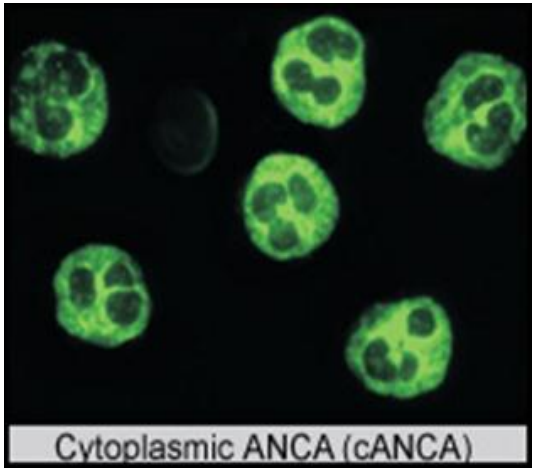
- **CBC** with differential typically demonstrates eosinophilia, usually with at least 10% eosinophils (or 5000-9000 eosinophils/µL), and anemia.
- **ESR** and **CRP** level are usually elevated.
- In patients with **renal involvement** serum creatinine levels are elevated. Urine analysis demonstrates proteinuria, microscopic hematuria, and red blood cell casts.
- **perinuclear-ANCA (p-ANCA):**positive (40%)

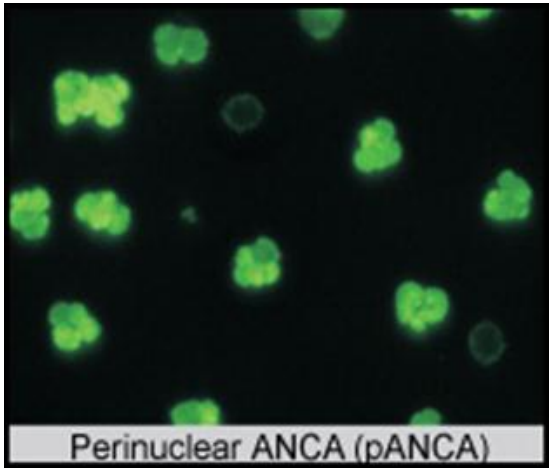
Sablé-Fourtassou R, Cohen P, Mahr A, Pagnoux C, et al. Antineutrophil cytoplasmic antibodies and the Churg-Strauss syndrome. Ann Intern Med. 2005 Nov 1. 143(9):632-8

Laboratory Studies (cont.)

antineutrophil cytoplasmic antibody (ANCA)

- ANCAs were originally described based on their **immunofluorescence patterns**
- **Types : cytoplasmic (c-ANCA)** and **perinuclear (p-ANCA)**





ANCA (cont.)

- The antigens responsible for these patterns have also been identified

1. **proteinase 3** (PR3) for c-ANCA
2. **myeloperoxidase** (MPO) for p-ANCA

- **c-ANCA** positive in Granulomatosis with polyangiitis (Wegener's granulomatosis) (90%)
- **p-ANCA** positive in EGPA (40%) & Microscopic polyangiitis (80%)

Specks U, Merkel PA, Seo P, Spiera R, Langford CA, Hoffman GS, et al. Efficacy of remission-induction regimens for ANCA-associated vasculitis. N Engl J Med 2013; 369: 417-27

ANCA (cont.)

- EGPA **Subsets** of Clinical Manifestations according to presence of ANCA:

1. **ANCAs−ve** : more eosinophilia, pulmonary infiltration and cardiac manifestation
2. **ANCAs+ve**: more vasculitis as purpra , Glomerulonephritis and Peripheral neuropathy

Gendelman, S.; Zeft, A. Childhood-onset eosinophilic granulomatosis with polyangiitis (formerly Churg-Strauss syndrome): A contemporary single-center cohort. J. Rheumatol. 2013,40, 929–935.

ANCA in Other Diseases

- **Connective tissue diseases**: SLE & rheumatoid arthritis,
- **Infections**: as HIV
- **Inflammatory bowel disease**: Ulcerative colitis & Crohn's disease
- **Other autoimmune GI diseases** :Sclerosing cholangitis, autoimmune hepatitis
- **Drug-induced ANCA**: Hydralazine & propylthiouracil

Laboratory Studies (cont.)

- Elevated **serum IgE levels**
- **Hypergammaglobulinemia**
- +ve results for **rheumatoid factor** at low titer
- Elevated levels of **eosinophil cationic protein** (ECP), **soluble interleukin-2 receptor** (sIL-2R), and **soluble thrombomodulin** (sTM), which is a marker of endothelial cell damage

Della Rossa A, Baldini C, Tavoni A, et al. Churg-Strauss syndrome: clinical and serological features of 19 patients from a single Italian centre. Rheumatology (Oxford) 2002; 41:1286.

Laboratory Studies (cont.)

- **Bronchoalveolar lavage** (BAL), eosinophilia is evident in 33% of cases.
- The laboratory tests that might correlate with **disease activity** include ESR and peripheral blood eosinophilia and ANCA in some pts

Sablé-Fourtassou R, Cohen P, Mahr A, Pagnoux C, Mouthon L, Jayne D, et al. Antineutrophil cytoplasmic antibodies and the Churg-Strauss syndrome. Ann Intern Med. 2005 Nov 1. 143(9):632-8

Imaging Studies

- As **chest radiography** and **chest computed tomography (CT)**.
- Other imaging studies are indicated for the complications of the disease and specific organ-system involvement, including **abdominal CT scanning** for pancreatitis, **coronary angiography** for myocardial ischemia and infarction, and **echocardiography** for congestive heart failure

Choi YH, Im JG, Han BK, Kim JH, Lee KY, Myoung NH. Thoracic manifestation of Churg-Strauss syndrome: radiologic and clinical findings. Chest. 2000 Jan. 117(1):117-24.

Chest x-ray

- Chest radiography findings are as follows:

1. **Pulmonary opacities** can be found in 75% of cases of EGPA;
2. Opacities usually are **bilateral**, **peripheral**, and **patchy**.
3. **Cavitation** is rare.
4. **Pulmonary infiltrates** may be transient.

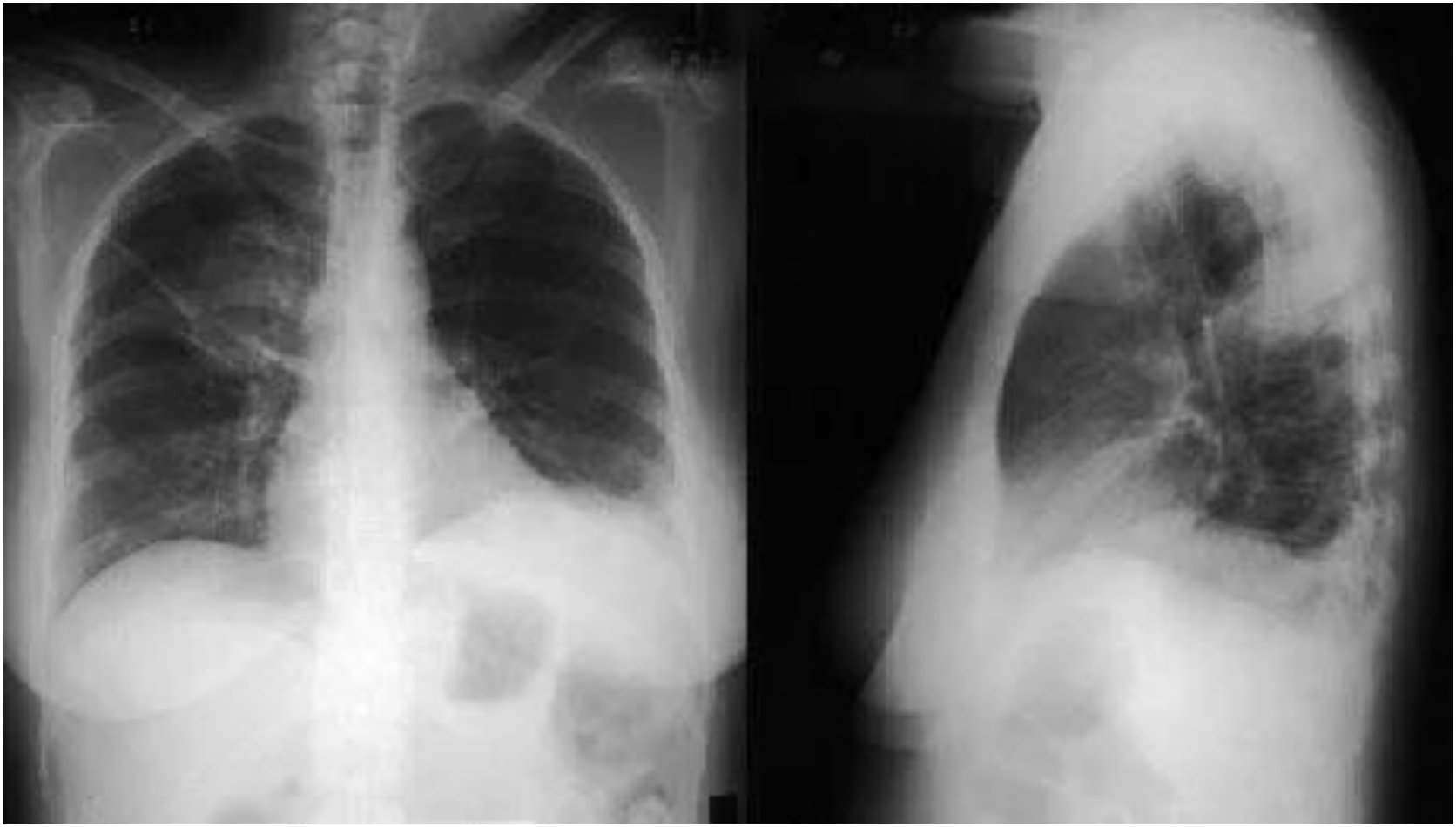
Chest x-ray (cont.)

5. **Extensive opacities** in the setting of a drop in hemoglobin levels suggest massive intra-alveolar hemorrhage as a result of pulmonary alveolar capillaritis

6. **Pleural effusions** are observed in 5%-30% of cases and can be eosinophilic.

7. **Hilar nodal enlargement** has occasionally been reported.

Transient pulmonary infiltrates in a patient with EGPA



CT chest

- Findings include peripheral areas of consolidation with ground-glass attenuation .
- Much less commonly, parenchymal nodules (from 5 mm to 3.5 cm), with cavitation or air bronchograms, can be observed.
- Bronchial dilatation and bronchial wall thickening may also be visible.

Choi YH, Im JG, Han BK, Kim JH, Lee KY, Myoung NH. Thoracic manifestation of Churg-Strauss syndrome: radiologic and clinical findings. Chest. 2000 Jan. 117(1):117-24

Other Tests

- **ECG** for cardiac manifestations
- **Echocardiography** should be done to all pts at baseline and repeated over time if symptoms and/or signs of heart failure develop
- **Gastrointestinal endoscopy** for GI bleeding
- **Electromyelography (EMG)** and **nerve conduction** for peripheral neuropathies

Biopsy

- Biopsy from the affected organ (lung, skin, kidney , muscle or nerve) is helpful in confirming the diagnosis.
- If no localizing finding exists, obtaining nerve or muscle biopsy may be considered
- Histologic Findings : necrotizing granulomas, & necrotizing vasculitis involving small arteries and venules associated with eosinophilic infiltration
- kidney biopsy results may show focal or crescentic glomerulonephritis

Classification criteria and definitions

- **The 2012 Chapel Hill Consensus Conference** defined EGPA as an eosinophil-rich and necrotizing granulomatous inflammation involving the respiratory tract with necrotizing vasculitis of small- and medium-sized vessels in association with asthma and eosinophilia.

1990 ACR classification criteria

1. Asthma
2. Eosinophilia of > 10% in peripheral blood
3. Paranasal sinusitis
4. Pulmonary infiltrates, may be transient, on chest imaging
5. Histologic evidence of vasculitis with extravascular eosinophils
6. Mononeuritis multiplex or polyneuropathy

If ≥ 4 criteria are present, sensitivity is 85%, and specificity is 99.7%.

Risk stratification

- **The French Vasculitis Study Group (2011)** has developed a five-point system ("five-factor score") (FFS) that predicts the risk of death in EGPA using clinical presentations. These factors are:

1. Reduced renal function (creatinine >1.58 mg/dl)
2. Proteinuria (>1 g/24h)
3. Gastrointestinal hemorrhage, infarction, or pancreatitis
4. Involvement of the central nervous system
5. Cardiomyopathy

Risk stratification (cont.)

- The **lack of any of these factors** indicates milder case, with a five-year mortality rate of 11.9%.
- **One factor** indicates severe disease, with a five-year mortality rate of 26%,
- **Two or more** indicate very severe disease: five-year mortality rate 46%

Guillevin, L.; Pagnoux, The five-factorscore revisited: Assessment of systemic necrotizing vasculitides based on the French Vasculitis Study Group (FVSG) cohort. Medicine 2011, 90, 19–27.

Treatment

- **Glucocorticoids** alone are usually adequate for the ttt of mild cases (FFS = 0) of EGPA
- Other immunosuppressive drugs as **(cyclophosphamide, methotrexate, azathioprine)** may be added, depending on severity (FFS ≥ 1) & usually are necessary in fewer than 20% of patients.
- **Rituximab**, is useful in treatment of steroid-resistant cases, as well as for prevention and treatment of relapse.

Beasley R, Bibby S, Weatherall M. Leukotriene receptor antagonist therapy and Churg-Strauss syndrome: culprit or innocent bystander?. Thorax. 2008 Oct. 63(10):847-9

Treatment (cont.)

- The anti-IgE monoclonal antibody **omalizumab** used as a corticosteroid-sparing effect in refractory or relapsing EGPA
- Major life-threatening organ involvement require ttt with **pulse** doses of intravenous **corticosteroids** and other cytotoxic agents as Cyclophosphamide.
- **Cyclophosphamide** is typically given in intravenous pulses for 3 months; afterward, pts are converted to oral **mycophenolate**, **azathioprine**, or **methotrexate** for maintenance therapy

Giavina-Bianchi P, Giavina-Bianchi M, Agondi R, Kalil J. Three months' administration of anti-IgE to a patient with Churg-Strauss syndrome. 2007 May. 119(5):1279;

Treatment (cont.)

- **Mepolizumab** is an interleukin-5 antagonist monoclonal antibody & is FDA approved for the treatment of EGPA in 2017
- **Plasma exchange** has been studied in EGPA and other ANCA-positive vasculitides, without a clear benefit.
- **Immune globulin** given as a monthly infusion, to pts who haven't responded to other treatments.

Wechsler ME, Akuthota P, Langford CA, et al. Mepolizumab or Placebo for Eosinophilic Granulomatosis with Polyangiitis. N Engl J Med. 2017 May 18. 376 (20):1921-1932

THANK YOU