1. DACH ANCA VASKULITIS FORUM 2023
12. & 13. MAI 2023

CSL Vifor

Einführung
ANCA-Vasculitis - Update zur ANCA-assoziierten Vaskulitis (AAV)

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Bern

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DE-AVA-2300045

VASAS
VASCULITIS
Association
Switzerland

Medizinisches Zentrum Monbijou

Einführung ANCA-Vasculitis

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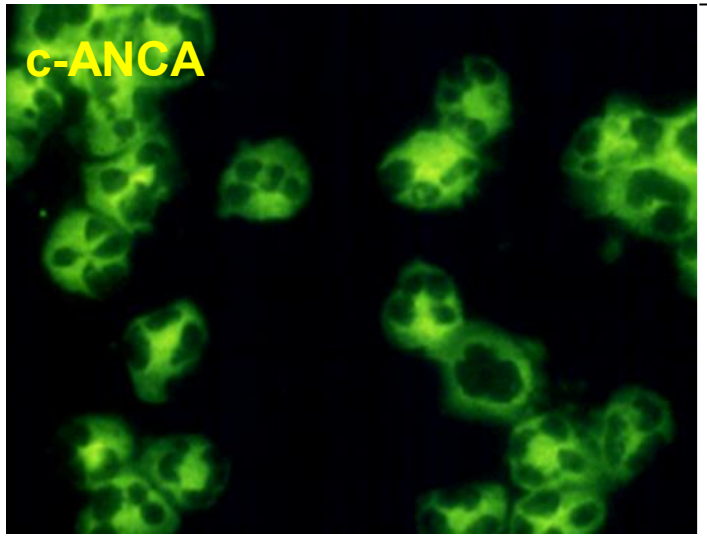
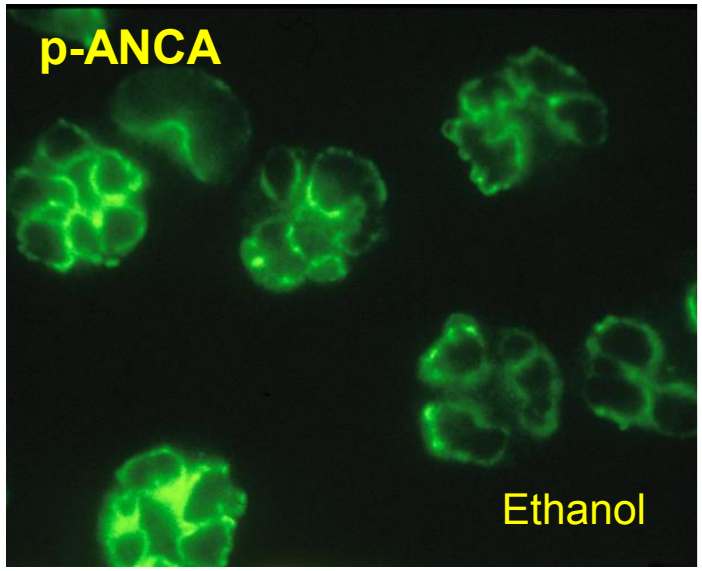
- Speaker, advisory fees and research support
  - Roche, MSD, Abbvie, Pfizer, Amgen, Chugai, BMS, GSK, Vifor, Astra-Zeneca

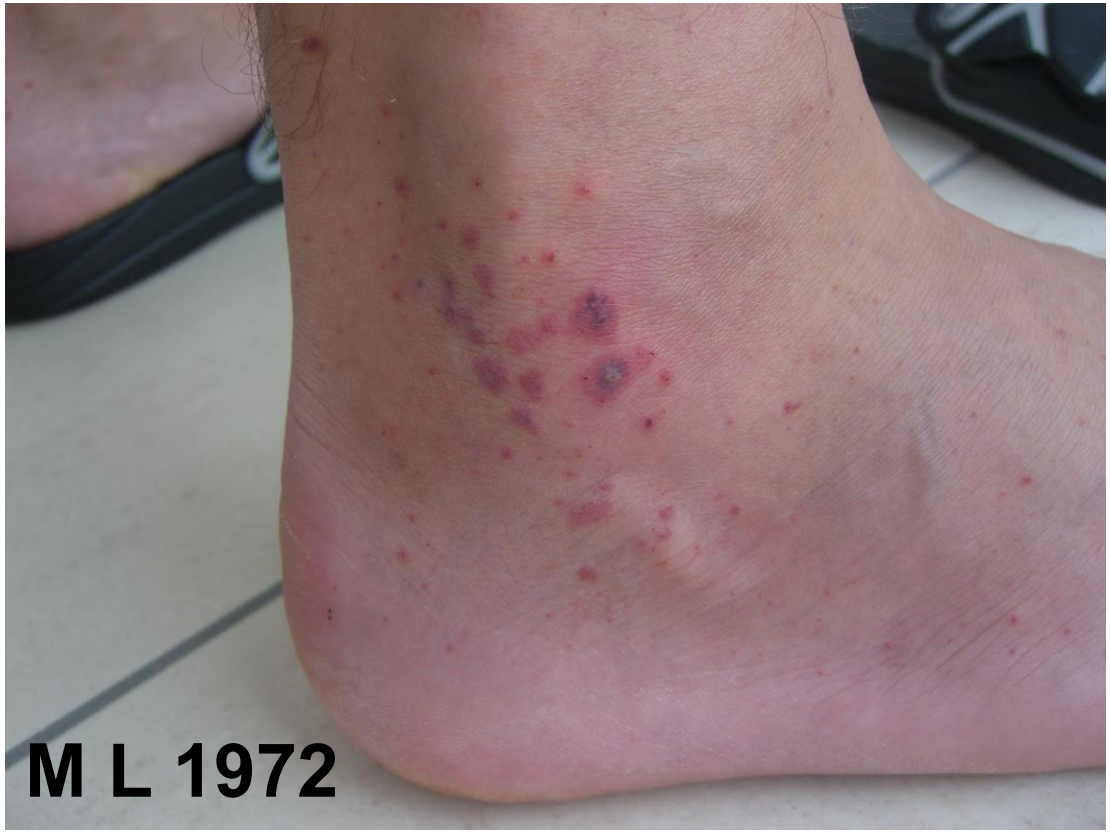
ANCA-assoziiert

Nekrotisierend

Pauci-immun

Fluoreszenz	Antigen	Assoziierte Krankheit
	Proteinase 3 (PR3)	GPA Cave: Infekt Medikamente
	Myeloperoxidase (MPO) Andere Antigene: Elastase, Lactoferrin	MPA eGPA RA SLE IBD

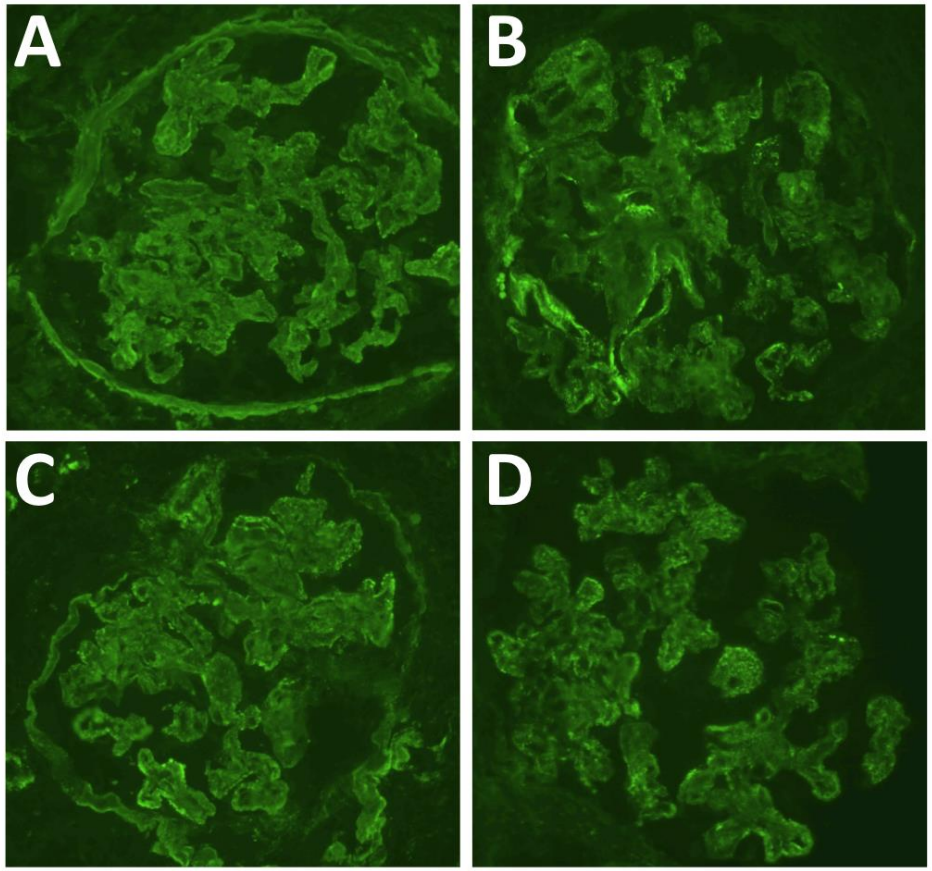
nekrotisierend



Pauci-immun

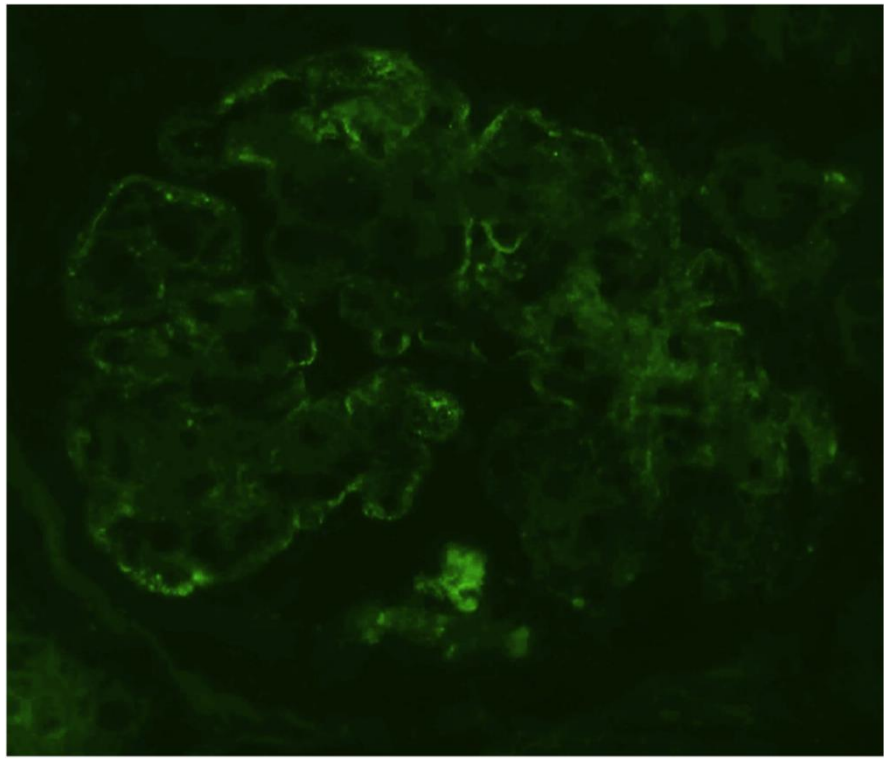
Lupus-Nephritis-Full-House

Immunfluoreszenz eines Glomerulums, “full house” Bild mit Deposition von IgG (A), IgA (B) Komplement C5b-9 (C) und C3 (D) (als hellgrüne Ablagerungen erkennbar)

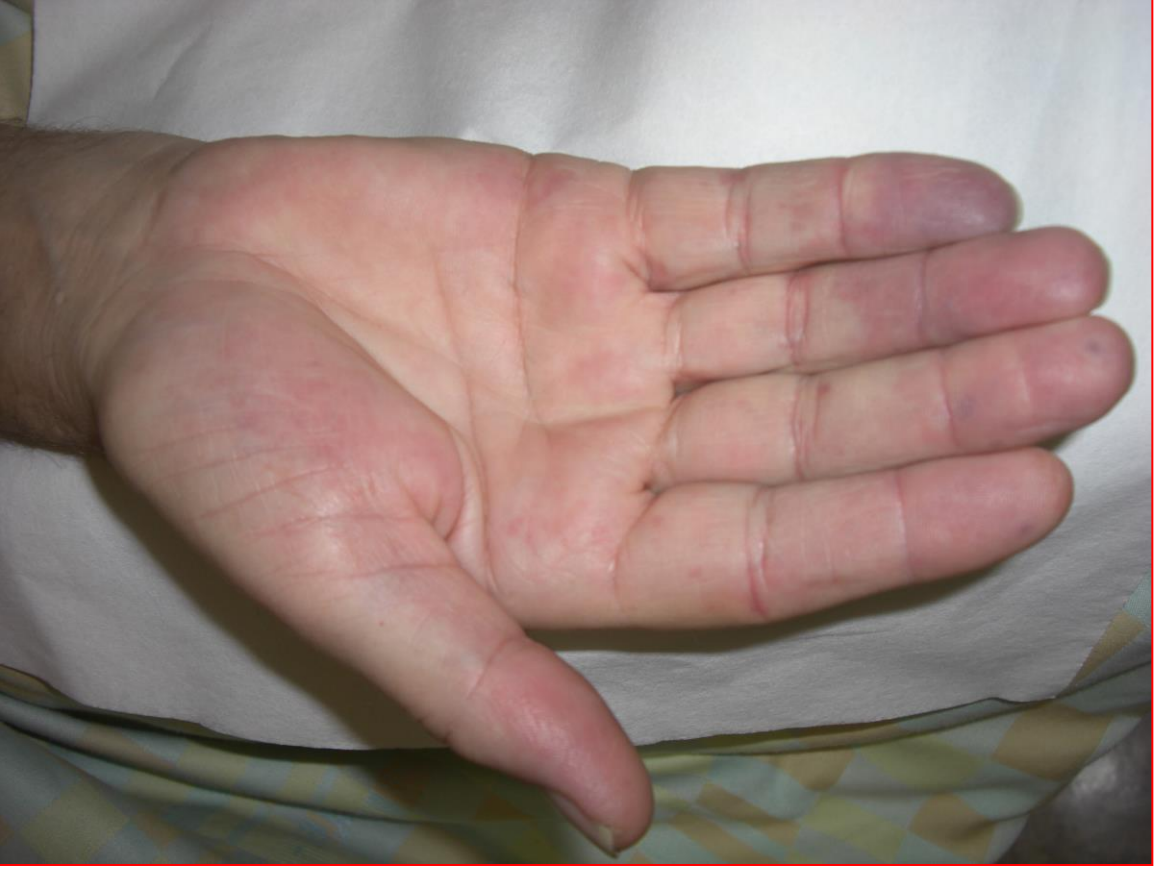


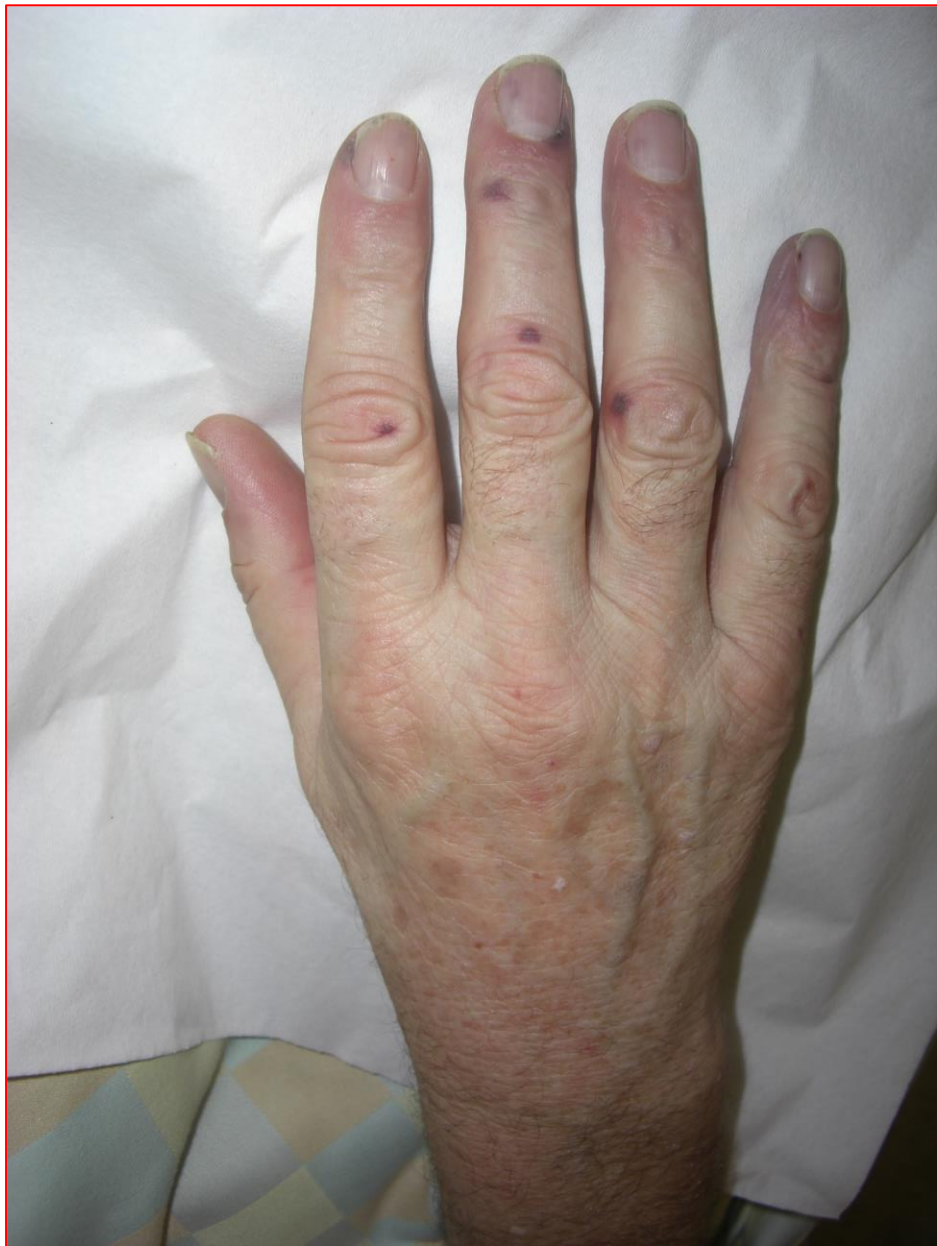
ANCA-Pauciimmun-GN

Immunfluoreszenz eines Glomerulums, kaum Depots (hellgrün) erkennbar



Engramme aus dem Alltag

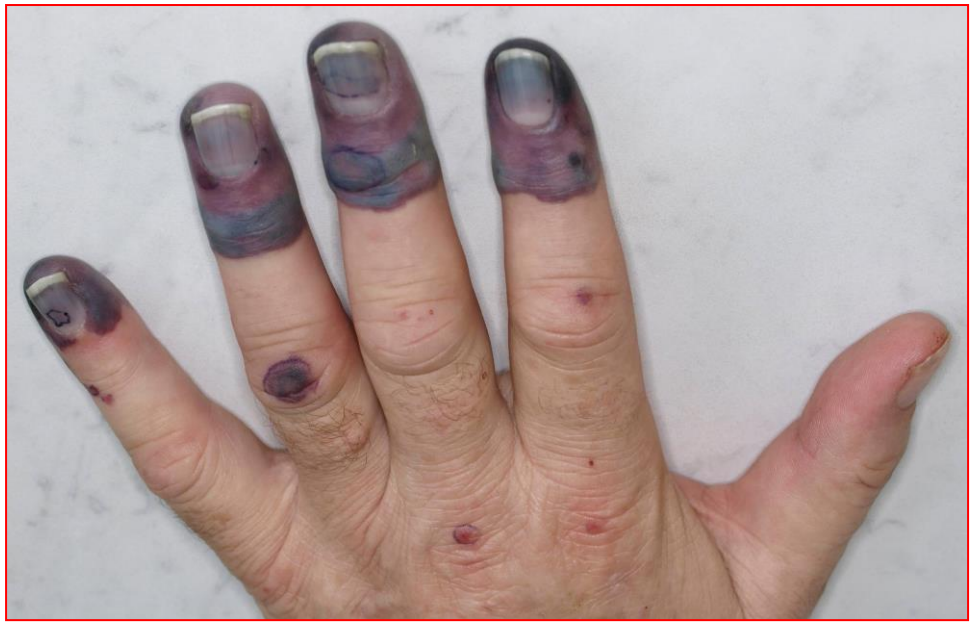


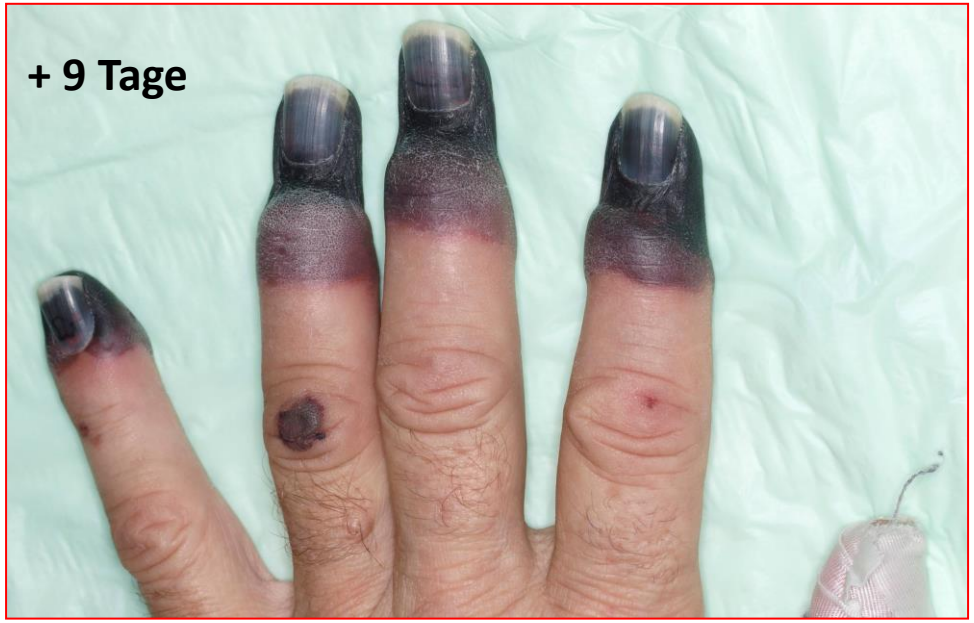


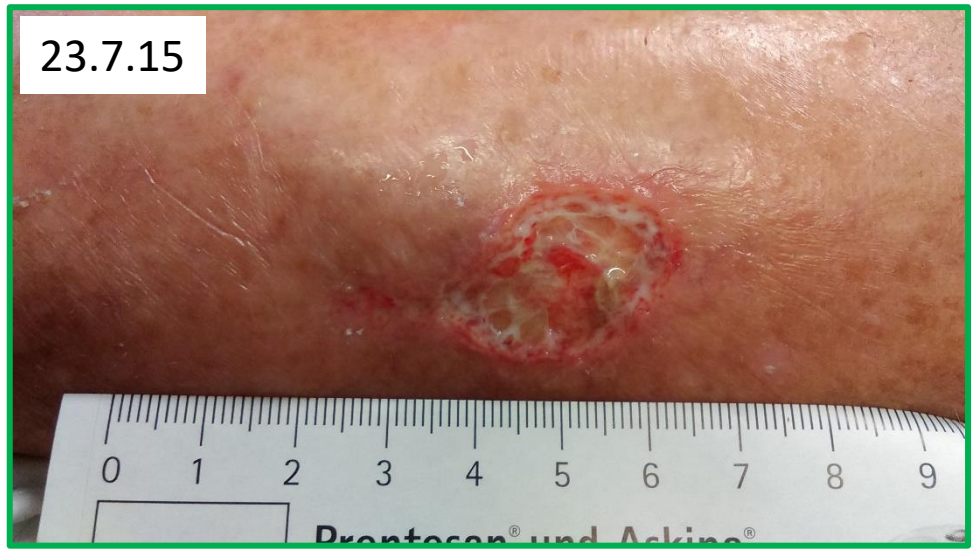
- 64 jähriger Mann
- Seit 10 Tagen kalte Akren
- Schmerzen

Anamnese: bland?
Strukturell – funktionell,
Welcher Gefässtyp?

Gefässe



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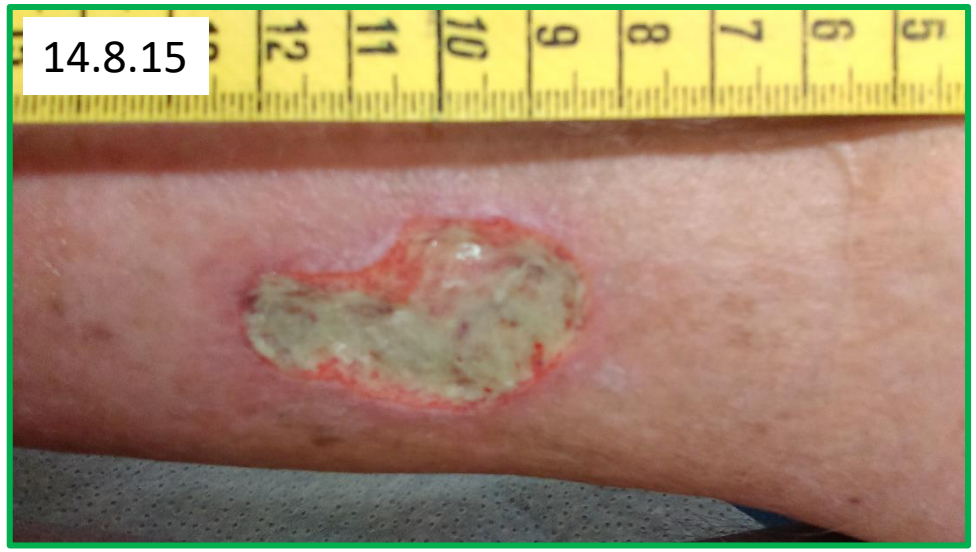
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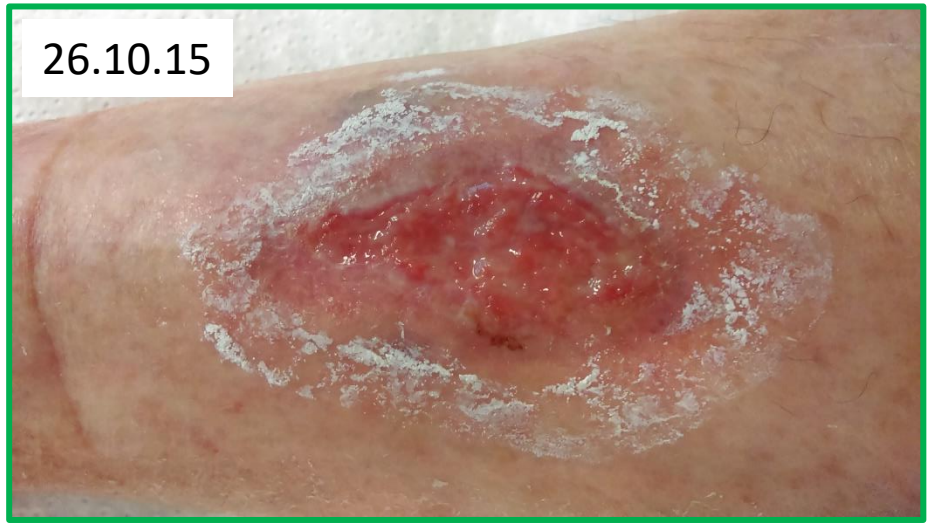
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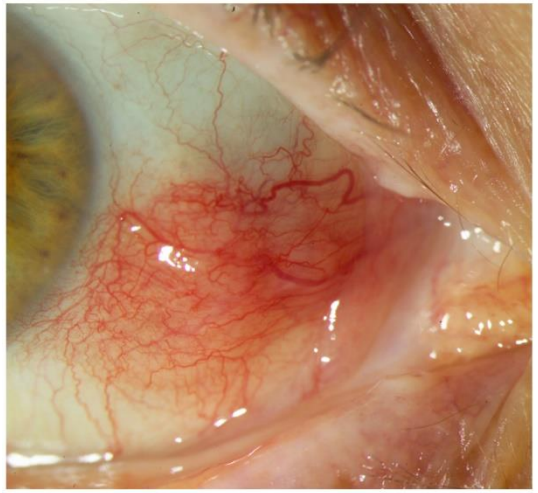
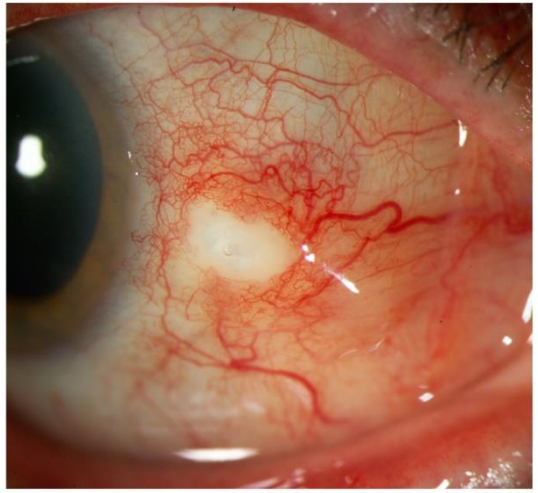
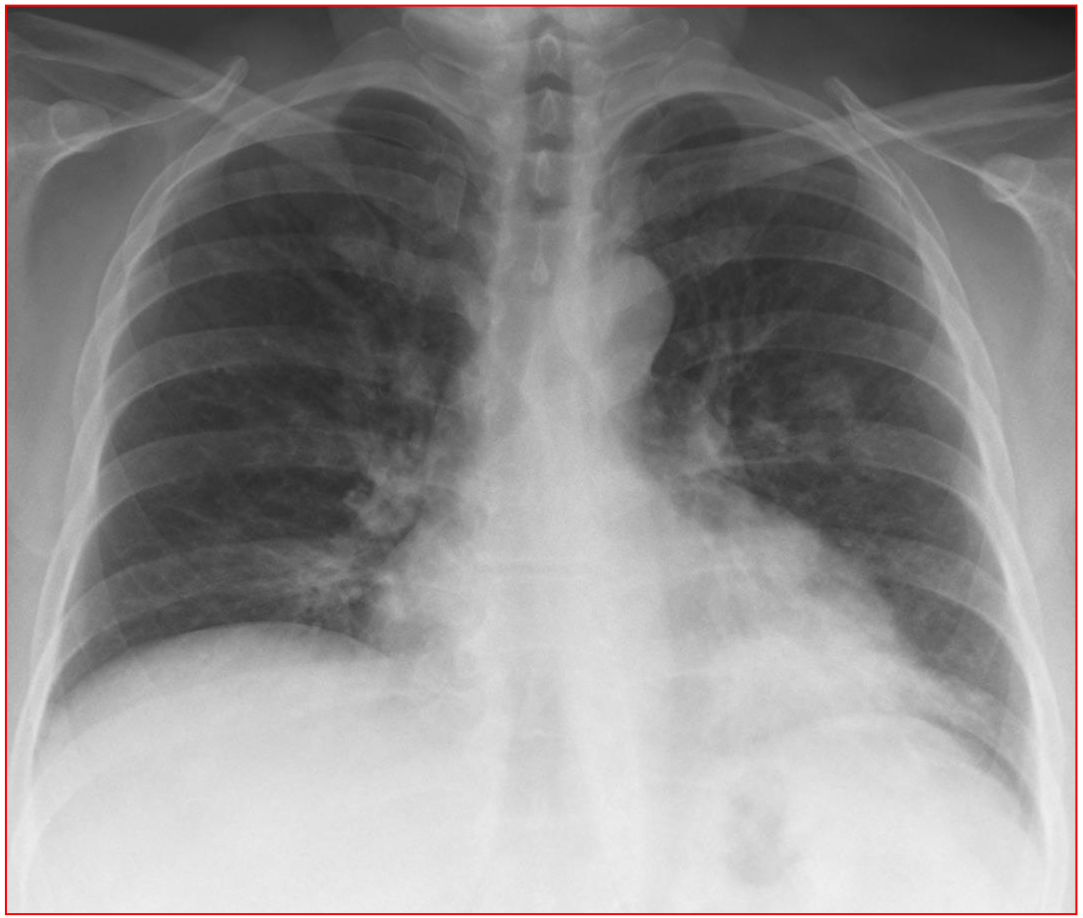
Auge: noduläre Skleritis





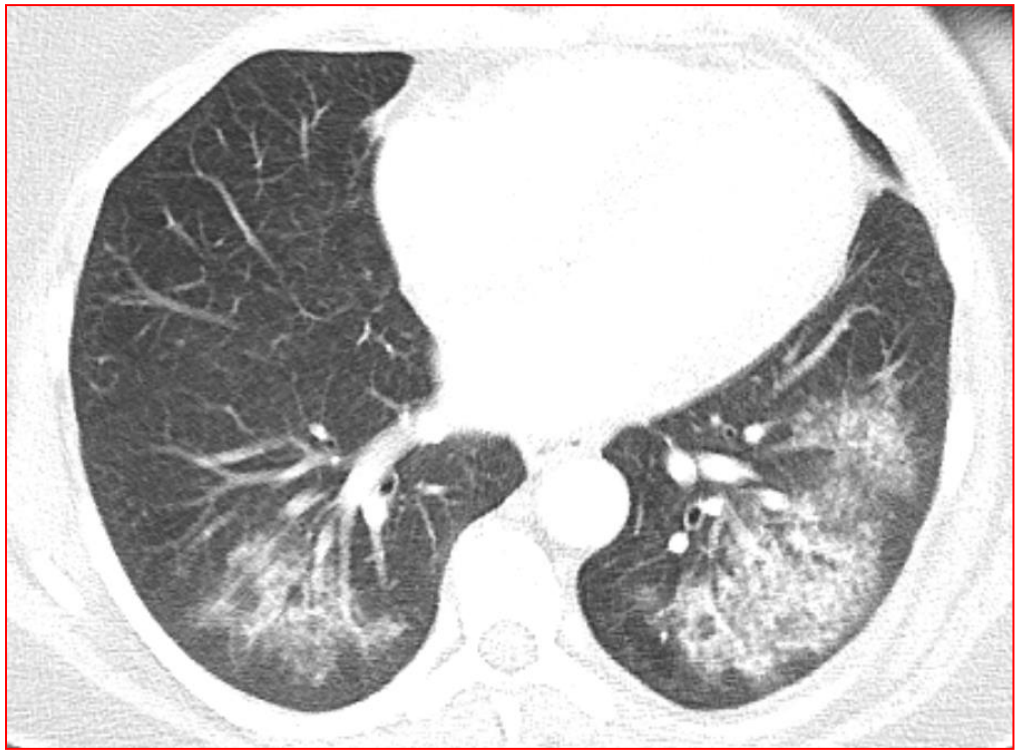
Lunge

Klinische Angaben: Husten mit leichtem Auswurf, Rasselgeräusche basal beidseits. Infiltrate?

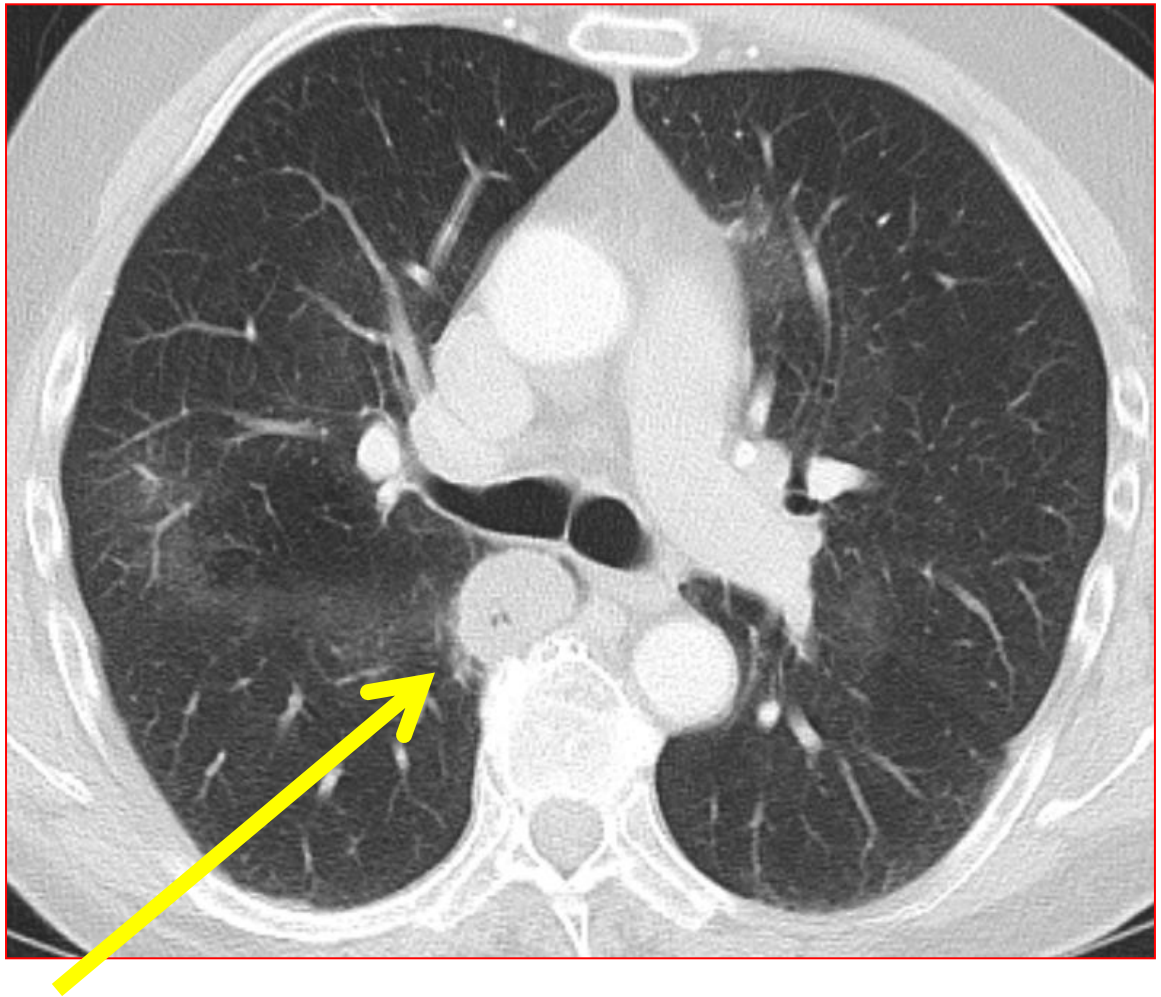


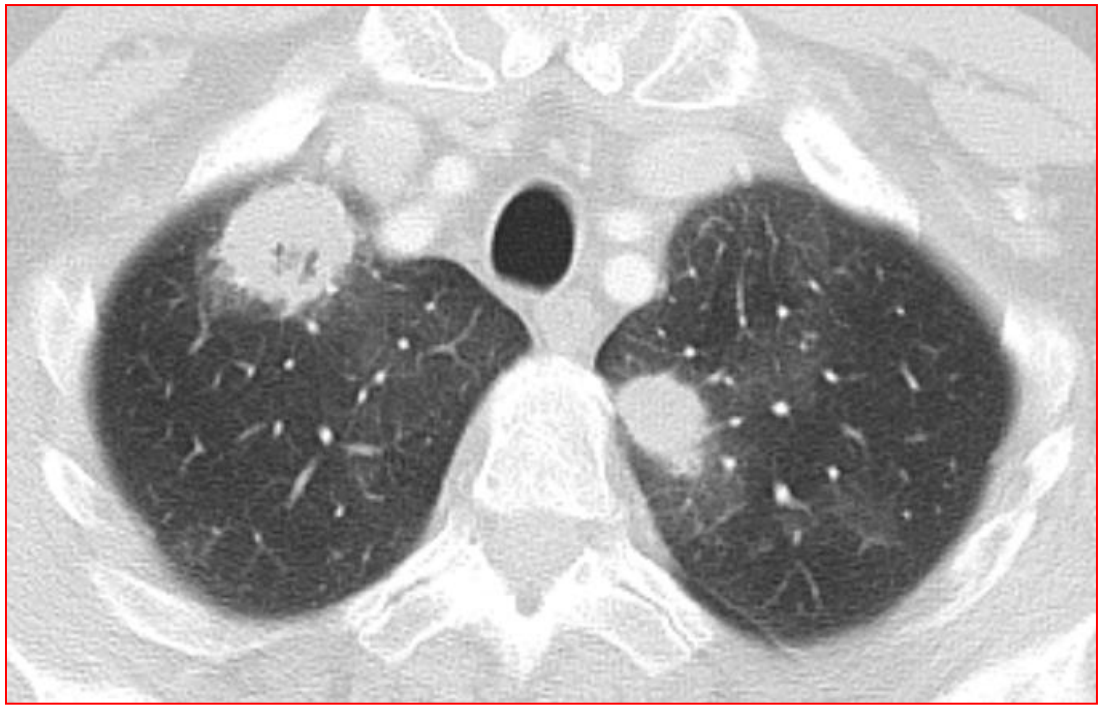
- Normozytäre normochrome Anämie
- ANCA
- CRP
- Urinstatus

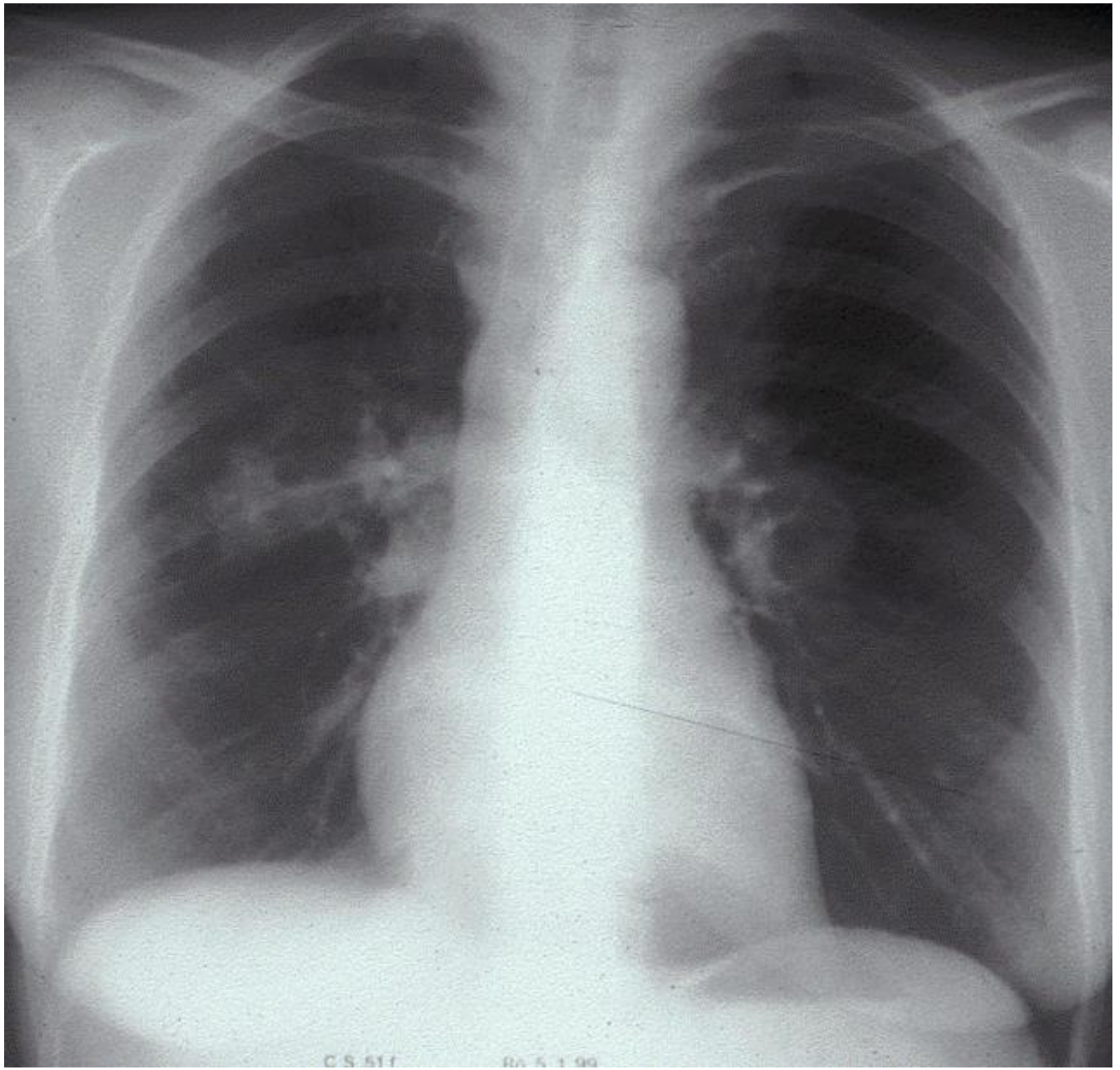
Azinäre und ground glass Infiltrate im dorsobasalen Unterlappen beidseits bei Lungenblutung im Rahmen einer ANCA-assoziierten Kleingefässvaskuliitis



Granulomatosis

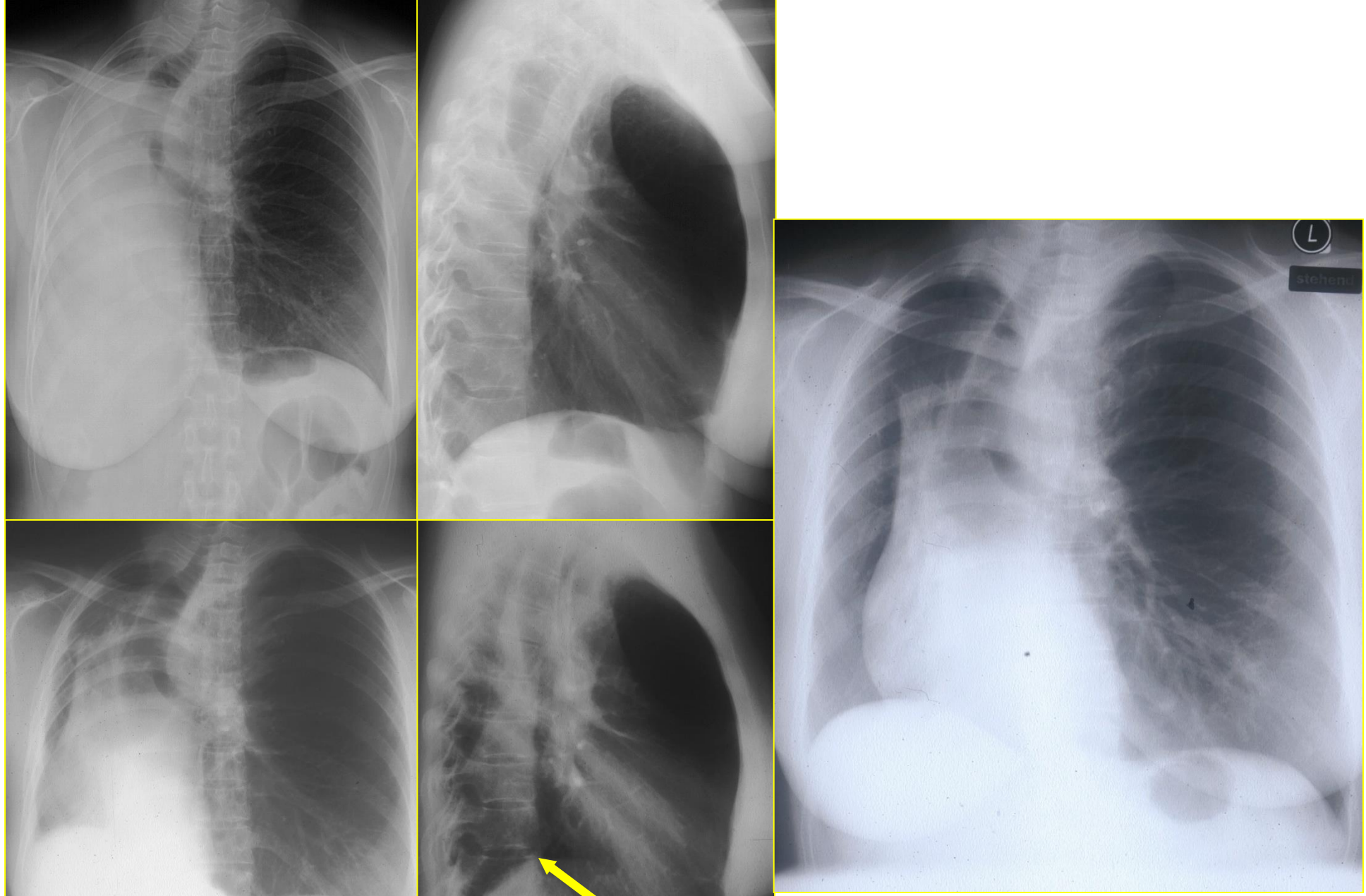






ANCA negativ
Granulomatose

Januar 1999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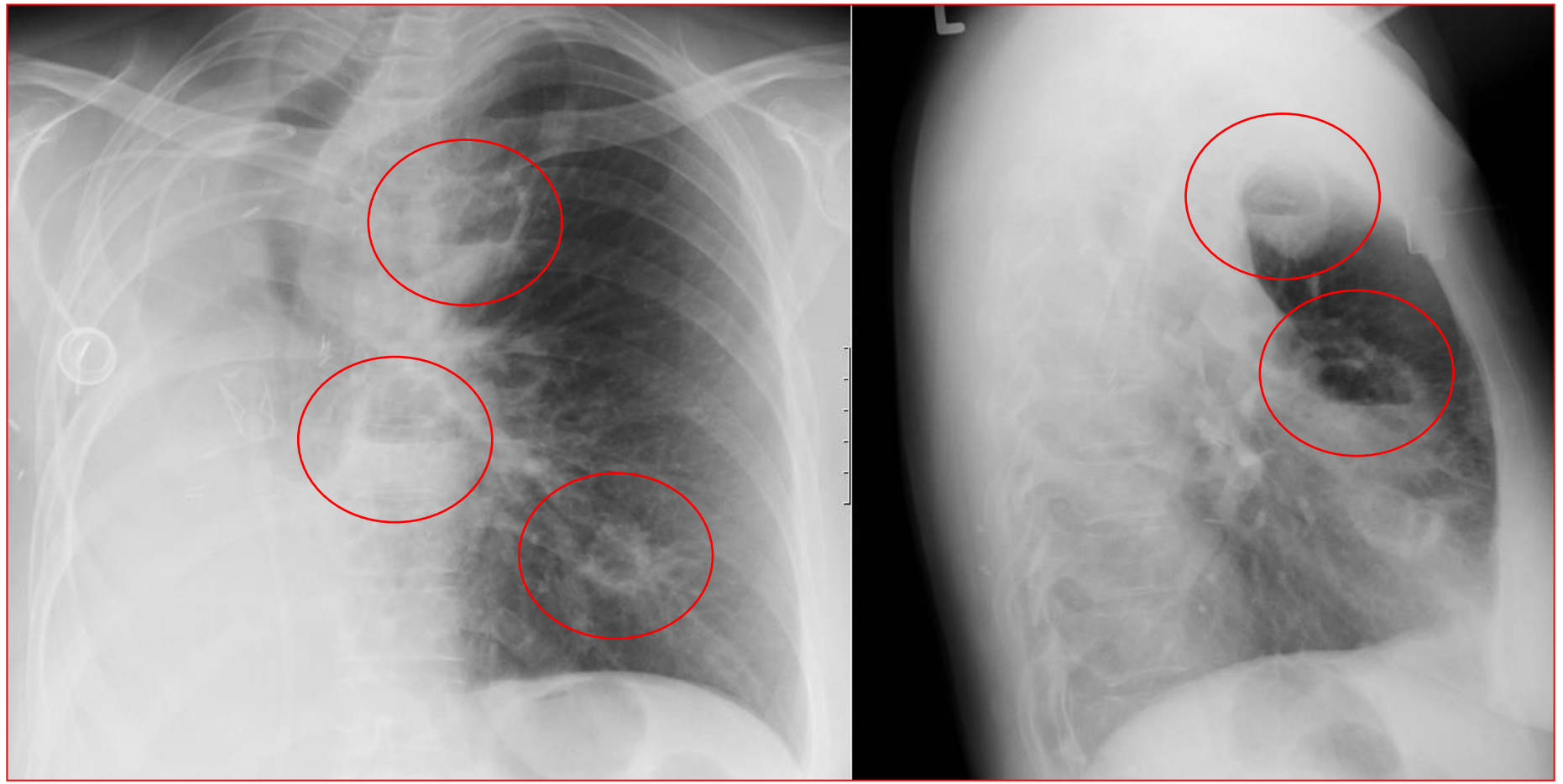

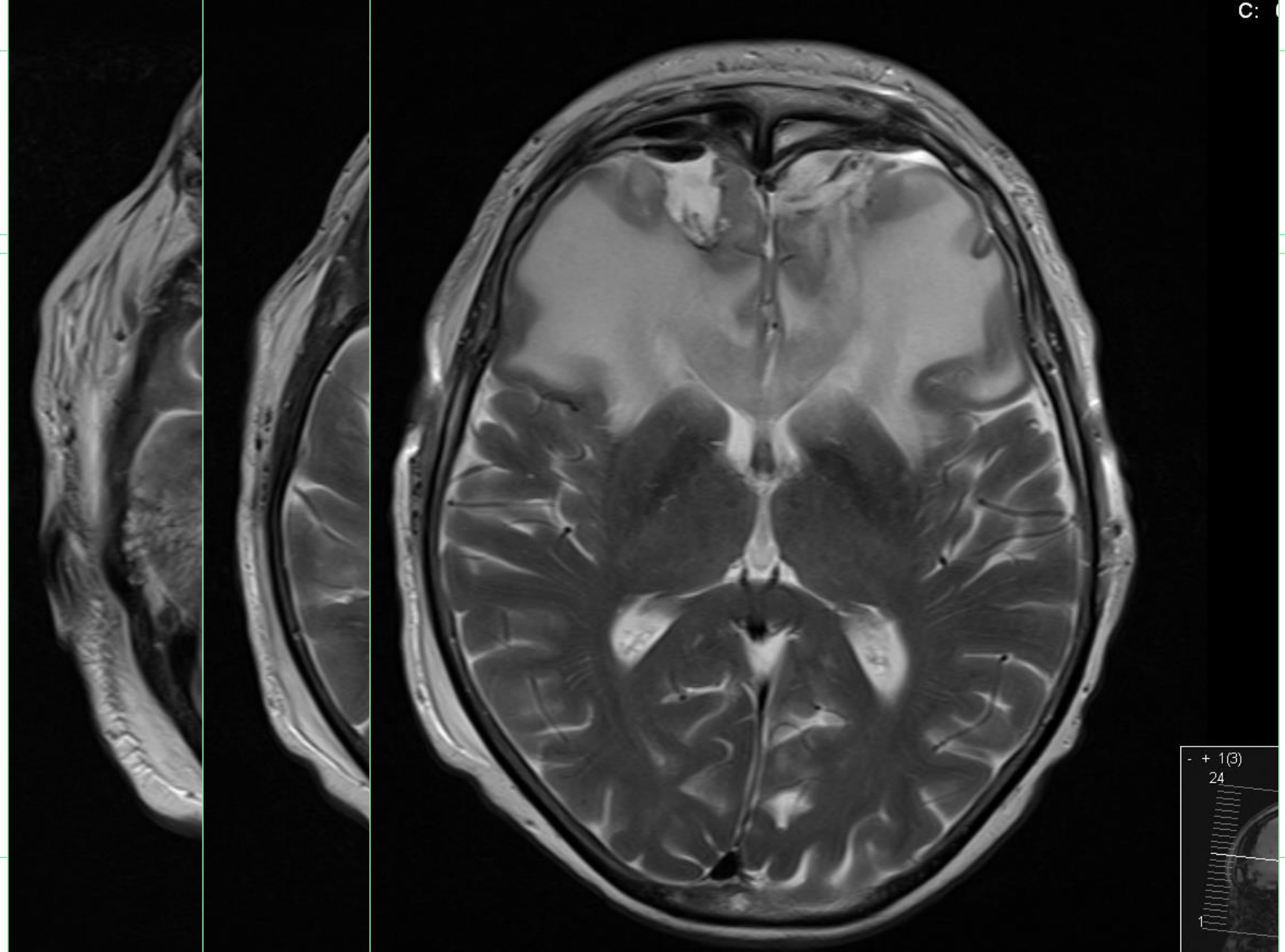
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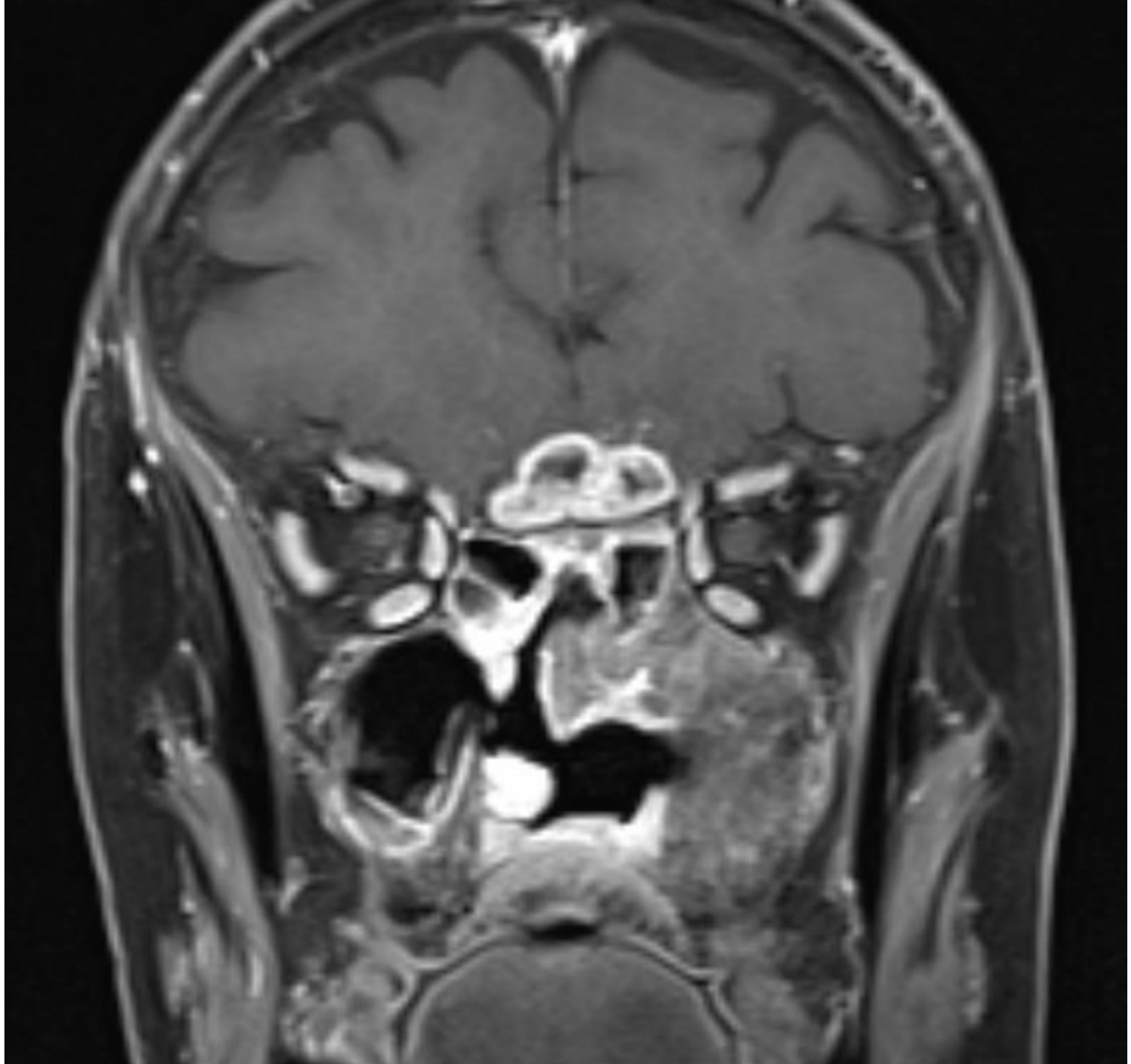

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Histology of eGPA

- 3 types of lesions: ❖ **Diagnosis**
  - Necrotizing vasculitis
  - Tissue infiltration by eosinophils
  - Extravascular granulomas
- 4 categories of lung histology:
  - Eos, necrotizing vasculitis, granuloma ❖ **Yes**
  - Eos, necrotizing vasculitis ❖ **Likely**
  - Eos + necrosis of other organ tissues ❖ **Possible**
  - Pulmonary eos only ❖ **No value**

Nervensystem

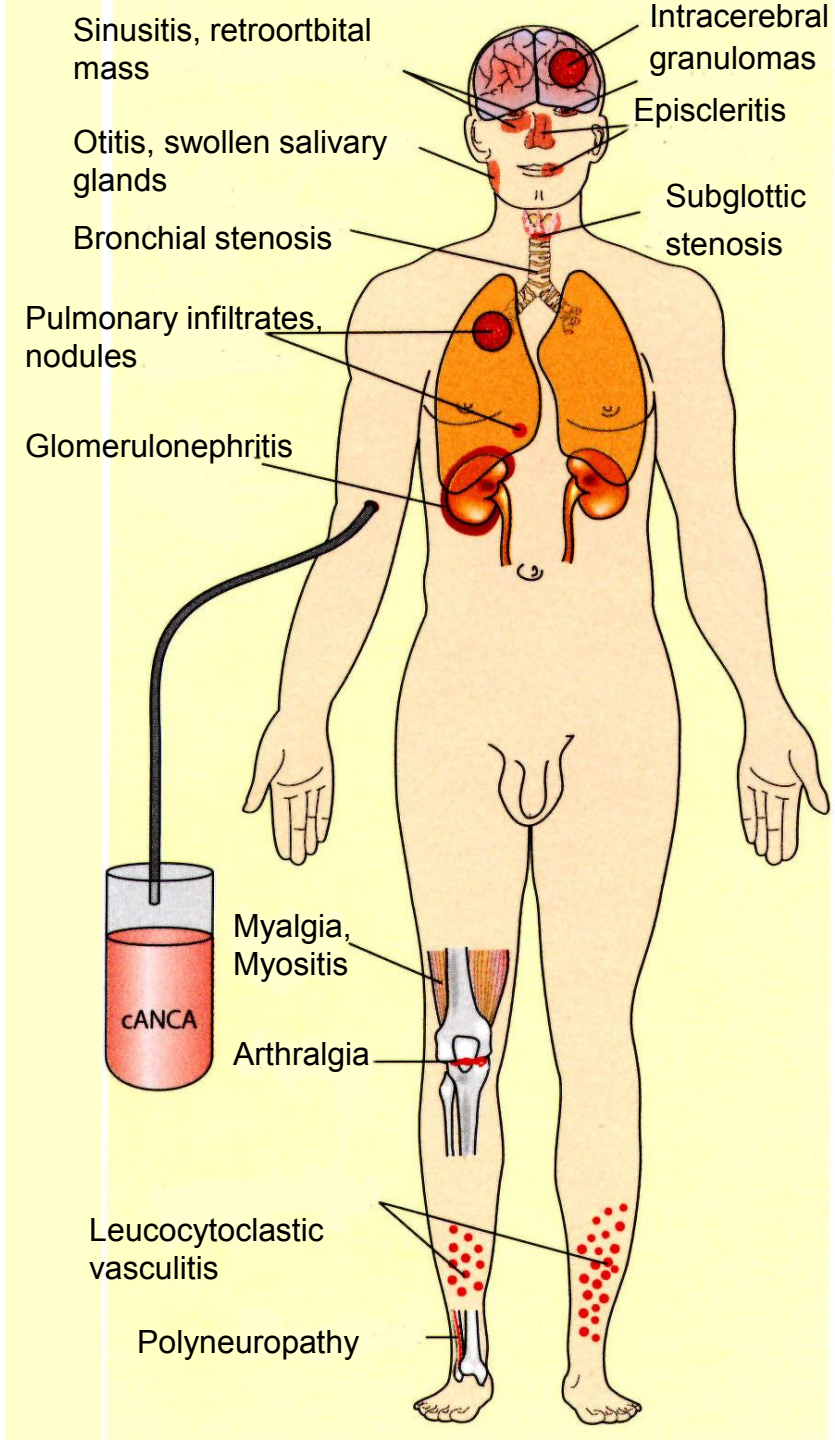
Neurapraxie – Axonotmesis - Neurotomie

Fragen

a. Axonal
b. Demyelinisierend

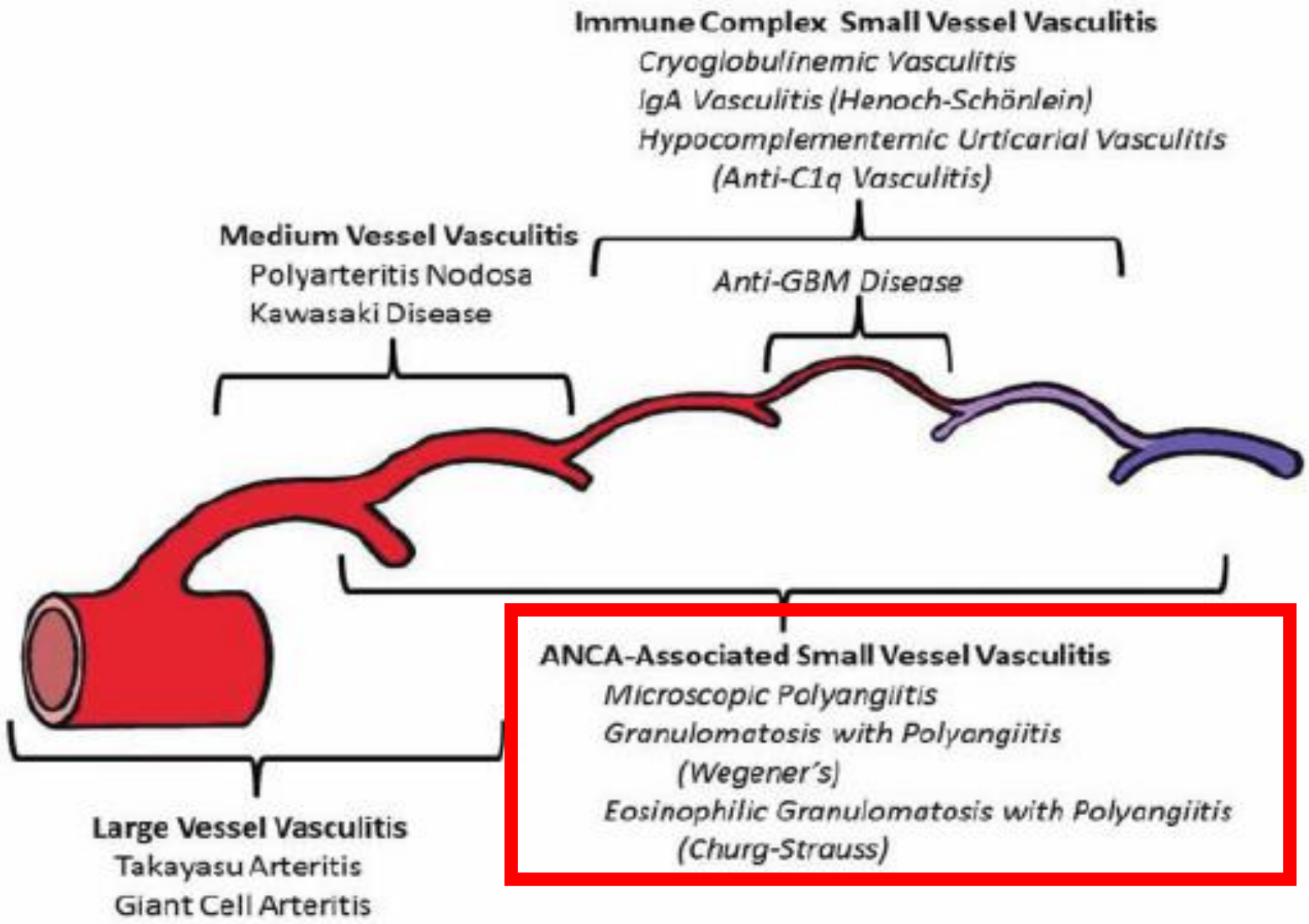
a. Chronisch progredient
b. Plötzlich

a. Definitiver Schaden
b. Funktionelle Erholung



- Lungen
- Nieren
- Nervensystem
- Muskeln, Gelenke
- Haut

The big 5: GPA, MPA, eGPA, RA, Infekt



+ RA-Vaskulitis

+ Infektvaskulitis

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Fazit

- Multiorganbefall
  - Mehrere Spezialitäten / Netzwerk
- Dynamik
  - Fast-track-Clinics / Netzwerk
- Schleichende Verschlechterung
  - Granulomatose: langsames invasiv-destruktives Wachstum
- Klinische Untersuchung
- «patient eduction»
  - Urinkontrollen / Blutdruck / Kenntnis über Notfallsituationen

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Die Fachpersonen können bei Vifor Pharma Switzerland AG eine vollständige Kopie des zitierten Prüfungsberichts anfordern.

Gekürzte Verschreibungsinformationen

Deutschland

▼ Dieses Arzneimittel unterliegt einer zusätzlichen Überwachung. Dies ermöglicht eine schnelle Identifizierung neuer Sicherheitsdaten. Angehörige der Gesundheitsberufe werden gebeten, alle Verdachtsfälle von unerwünschten Wirkungen zu melden.

TAVNEOS®▼ 10 mg Hartkapseln: Wirkstoff: Avacopan. **Zusammensetzung:** Jede Hartkapsel enthält 10 mg Avacopan. Sonstige Bestandteile mit bekannter Wirkung: 245 mg Macrogolglycerolhydroxystearat (Ph.Eur.). **Anwendungsgebiete:** Tavneos ist in Kombination mit einem Rituximab- oder Cyclophosphamid-Dosierungsschema indiziert zur Behandlung erwachsener Patienten mit schwerer aktiver Granulomatose mit Polyangiitis (GPA) oder mikroskopischer Polyangiitis (MPA). **Gegenanzeigen:** Überempfindlichkeit gegen den Wirkstoff oder einen der sonstigen Bestandteile. **Nebenwirkungen:** Sehr häufig: Übelkeit, Kopfschmerzen, erniedrigte Leukozytenzahl, Infektion der oberen Atemwege, Diarrhö, Erbrechen, Nasopharyngitis, erhöhte Werte in Leberfunktionstests. Häufig: Pneumonie, Rhinitis, Harnwegsinfektion, Sinusitis, Bronchitis, Gastroenteritis, Infektion der unteren Atemwege, Zellulitis, Herpes zoster, Influenza, Orale Candidose, Oraler Herpes, Otitis media, Neutropenie, Schmerzen im Oberbauch, erhöhte Kreatinphosphokinase im Blut. Gelegentlich: Angioödem. **VERSCHREIBUNGSPFLICHTIG. Fachinformation beachten. Pharmazeutischer Unternehmer:** Vifor Fresenius Medical Care Renal Pharma France, 100-101 Terrasse Boieldieu, Tour Franklin La Défense 8, 92042 Paris La Défense Cedex, Frankreich. **Stand der Information:** Februar 2022

Österreich

▼ Dieses Arzneimittel unterliegt einer zusätzlichen Überwachung. Dies ermöglicht eine schnelle Identifizierung neuer Sicherheitsdaten. Angehörige der Gesundheitsberufe werden gebeten, alle Verdachtsfälle von unerwünschten Wirkungen zu melden.

Tavneos® Fachkurzinformation: Tavneos®10mg Hartkapsel. **Zusammensetzung:** Jede Hartkapsel enthält 10 mg Avacopan. Sonstige Bestandteile mit bekannter Wirkung:245 mg Macrogolglycerolhydroxystearat(Ph.Eur). **Anwendungsgebiete**: Tavneos® ist in Kombination mit einem Rituximab- oder Cyclophopamid-Dosierungsschema indi- ziert zur Behandlung erwachsener Patienten mit schwerer aktiver Granulomatose mit Polyangiitis (GPA) oder mikroskopischer Polyangiitis (MPA). **Gegenanzeigen**: Überempfindlichkeit gegen den Wirkstoff oder einen der sonstigen Bestandteile. Pharmakotherapeutische Gruppe: L04AJ05 Complement Inhibitors **ATC-Code:** L04AJ05. **Inhaber der Zulassung:** Vifor France, 100-101 Terrasse Boieldieu Tour Franklin La Defense 8 92042 Paris La Defense Cedex, Frankreich. Rezept- und apothekenpflichtig. Weitere Angaben zu Warnhinweisen und Vorsichtsmaßnahmen für die Anwendung, Wechselwirkungen mit anderen Arzneimitteln oder sonstigen Wechselwirkungen, Schwangerschaft und Stillzeit und Nebenwirkungen sowie Gewöhnungseffekten sind der veröffentlichten Fachinformation zu entnehmen. Stand der Information: Letzter Stand Fachinformation

Schweiz

▼Dieses Arzneimittel unterliegt einer zusätzlichen Überwachung. Für weitere Informationen, siehe Fachinformation TAVNEOS® auf www.swissmedicinfo.ch.

Tavneos®: Z: Avacopan. **I:** Tavneos, als ergänzende Therapie zu einer immunsuppressiven Standardbehandlung auf Basis von Rituximab oder Cyclophosphamid mit Glukokortikoiden, ist für die Behandlung erwachsener Patienten mit schwerer aktiver ANCA Vaskulitis (GPA/MPA) indiziert. **D:** Orale Einnahme morgens und abends 2x täglich 30 mg (3 Kapseln zu je 10 mg) mit Nahrung. **KI:** Überempfindlichkeit gegen den Wirkstoff oder einen der Hilfsstoffe. **VM:** Hepatotoxizität; Angioödem; Überwachung des Blutbildes (weisse Blutkörperchen); Schwere Infektionen; Reaktivierung des Hepatitis-B-Virus; Herzbeschwerden; Bösartige Tumore; Macroglycerinhydroxystearat. **S/S:** Eine Anwendung während der Schwangerschaft und bei Frauen im gebärfähigen Alter, die keine Verhütungsmethode anwenden, ist nicht empfohlen. Es ist nicht bekannt, ob Avacopan in die Muttermilch ausgeschieden wird. Der Nutzen des Stillens für das Kind sollte gegen den Nutzen der Behandlung für die Patientin abgewogen werden. **UW:** Sehr häufig: Infektion der oberen Atemwege, Nasopharyngitis; Kopfschmerzen; Erbrechen, Durchfall, Übelkeit; erhöhter Lebertest; verminderte Anzahl weisser Blutkörperchen. Häufig: Lungenentzündung, Infektion der unteren Atemwege, Influenza, Bronchitis, Zellulitis, Infektion der Harnwege, Herpes zoster, Sinusitis, orale Candidose, Herpes im Mundbereich, Otitis media, Rhinitis, Gastroenteritis; Neutropenie; Oberbauchschmerzen; Anstieg der Kreatinphosphokinase im Blut. Gelegentlich: Angioödeme. **IA:** Avacopan ist ein Substrat von CYP3A4. Die gleichzeitige Verabreichung von Induktoren oder Inhibitoren dieses Enzyms kann die Pharmakokinetik von Avacopan beeinflussen. Siehe Fachinformation. **P:** Tavneos 10 mg: 30 und 180 Hartkapseln. **Liste B**. Detaillierte Informationen**:** www.swissmedicinfo.ch. Stand der Information**:** September 2022. **Zulassungsinhaberin:** Vifor Fresenius Medical Care Renal Pharma Ltd., St. Gallen. **Vertrieb:** Vifor Pharma Switzerland AG, CH-1752 Villars-sur-Glâne | CH-AVA-2300011