Pulmonary involvement in systemic vasculitis: imaging of parenchymal and airways alterations and differential diagnosis

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Learning objectives

- To describe the most common systemic vasculitis that cause pulmonary manifestations, illustrating their typical/atypical features as seen on chest-CT and their possible changes over time and therapy.
- To illustrate their peculiar imaging patterns to help in the differential diagnosis among the various vasculitis and other infectious/inflammatory or neoplastic diseases.
- To underline the importance of clinical data and close cooperation with the referring physician to possibly achieve the definitive diagnosis.
Background

Systemic primary vasculitis are a group of idiopathic diseases in which the sole or predominant histological feature is primarily represented by an inflammatory insult to the vessel walls with vessel destruction.

Several classifications of Systemic Vasculitis have been proposed over time, but the Chapel-Hill Consensus Conference classification is still considered as the most useful and practical; this classification is based on the size of vessels principally involved (large, medium and small vessels; see Table 1).

Chapel-Hill Consensus Conference: systemic vasculitis nomenclature

Large vessel Vasculitis

Giant Cell (temporal) arteritis
Granulomatous arteritis of aorta and its major branches (mainly extracranial branches of carotid artery)

Takayasu's arteritis
Granulomatous arteritis of aorta and its major branches (usually young patients)

Medium vessel Vasculitis

Polyarteritis nodosa
Vessel necrotizing inflammation without renal involvement

Kawasaki disease
Arteritis (usually coronary arteries in young patients) associated with mucocutaneous lymph node syndrome

Small vessel Vasculitis

ANCA-associated granulomatous vasculitis (formerly Wegener's granulomatosis)
Granulomatous inflammation. Usual pulmonary and renal involvement

Microscopic polyangiitis
Necrotizing vasculitis with few or no immune deposits; necrotizing glomerulonephritis and pulmonary involvement are common

Henoch-Shonlein purpura
Vasculitis with IgA immuno deposits, typically involving skin, gut and kidneys; arthralgias or arthritis

Essential cryoglobulinemic vasculitis
Vasculitis with cryoglobulin immune deposits associated with serum
Cutaneous leukocytoclastic angiitis
Isolated cutaneous involvement

Churg-Strauss Syndrome
Granulomatous inflammation with eosinophil-rich infiltration; typical respiratory tract involvement associated with asthma and eosinophilia

Table 1. Chapel-Hill Consensus Conference on systemic vasculitis nomenclature; vasculitis classification is based on the size of the vessels mainly affected (large, medium, small); vasculitis most frequently showing pulmonary involvement are underlined.

Although the exact pathogenesis of vasculitis is still matter of discussion and research, an immunological dysfunction is highly suspected, as suggested by many clinical, pathological and serological data derived from affected patients.

Also genetic and environmental factors (drugs or infectious microorganisms) would play a key role in the genesis and development of disease.

Three main immunological mechanisms of vessel damage have been identified and the genesis of each vasculitis would be primarily based on one of them:

- Deposition of immune complexes;
- Antibody-mediated vascular disease;
- Cell-mediated immunity and granulomatous reaction.

In particular, the investigations on the antibodies directed against peri-nuclear and cytoplasmatic neutrophil components (ANCA: antineutrophil cytoplasmatic antibodies) allowed the identification of a sub-group of ANCA-associated vasculitis (see Table 2) characterized by similar clinical features, involvement of small vessels and response to immunosuppressive treatment. Two indirect immunofluorescent staining patterns have been described: cytoplasmic ANCA (c-ANCA) and perinuclear ANCA (p-ANCA).

<table>
<thead>
<tr>
<th>Vasculitis</th>
<th>ANCA association</th>
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<tbody>
<tr>
<td>ANCA-associated Granulomatous Vasculitis</td>
<td>c-ANCA 85%  p-ANCA 10%</td>
</tr>
<tr>
<td>Churg-Strauss Syndrome</td>
<td>c-ANCA 10%  p-ANCA 40%-75%</td>
</tr>
<tr>
<td>Microscopic Polyangiitis</td>
<td>c-ANCA 15%-45% p-ANCA 45%-60%</td>
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Table 2. ANCA-associated vasculitis; respective frequency (%) of association with c-ANCA and p-ANCA is reported.
A pulmonary involvement can be identified in all systemic vasculitis, but the conditions that are most frequently accompanied by lung alterations can be summarized as follows: **ANCA-associated granulomatous Vasculitis** (formerly Wegener’s Granulomatosis), **Churg-Strauss Syndrome**, **Microscopic Polyangiitis**, **Goodpasture Syndrome**, **Behçet’s Disease** and **Takayasu Arteritis**.

Primary vasculitis are rare diseases and their clinical signs and symptoms are often similar to those of other systemic diseases, infections or malignancies.

Small-vessel vasculitis can cause nonspecific symptoms as fever, myalgias, arthralgias, and malaise; a multiple organ systems involvement can be found, with various clinical scenarios such as renal failure, uveitis, sinus disease, shortness of breath and rashes. Such clinical associations should always arise the suspicion of a small-vessel vasculitis when another cause is still unknown.

On the other side, large-vessel vasculitis can cause clinical signs and symptoms related to ischemia and unexplained inflammatory syndromes.

Since overlapping imaging features and similar clinical presentations are very common, the differential diagnosis among the different vasculitis or other inflammatory/infectious and neoplastic diseases can be extremely challenging. For these reasons, a deep knowledge of the peculiar imaging patterns of each disease, integrated with the patient’s clinical background derived from a close cooperation with the referring clinician, is necessary to achieve the correct diagnosis.
When assessing a chest-CT in a case of suspect vasculitis, a radiologist can face an incredibly wide spectrum of imaging presentations, ranging from no abnormalities (no pulmonary involvement) to extensive lung alterations.

When detected, a lung involvement can derive from pathological alterations affecting any anatomical or functional component of the lung: vessels (pulmonary and/or systemic), parenchyma, airways (trachea and bronchi).

The main imaging patterns suggesting a lung involvement are represented by: parenchymal nodules and masses (with or without cavitations), diffuse or focal ground-glass opacities, parenchymal consolidations, tracheal or bronchial abnormalities (wall thickening, stenosis), vascular abnormalities (aneurysms, wall thickening, stenosis and inflammation), pleural disease; recently, a non fortuitous association with pulmonary fibrosis has come to evidence, especially in ANCA-associated vasculitis.

Although overlapping patterns are very common, characteristic imaging features and their associations can help distinguishing a disease from the other. In this process, the knowledge of the clinical background of the patient is crucial.

Moreover, each alteration can show significant changes over time, related to the natural evolution of disease (acute versus chronic alterations) and therapeutic interventions, which can also lead to a complete resolution with no residual alterations.

**ANCA-associated granulomatous vasculitis (formerly Wegener's Granulomatosis):**

ANCA-associated granulomatous vasculitis is a rare multi-organ disease (prevalence of 3/100,000 in the USA), which can affect all ages but is most common in adults 50-69 years of age; men are slightly more frequently affected than women.

All organs can be affected; the most frequently affected sites are: the upper respiratory tract (sinusitis and nasal ulceration), kidneys and lower respiratory tract. The lower respiratory tract (airways and lungs) is affected in the 90% of patients.

**Radiological manifestations of the disease:**

* Nodules and masses:

The most typical radiological manifestation of lung disease at chest-CT is represented by nodules and masses (90% of cases) (Fig. 1 on page 16). In active disease, nodules
are expression of granulomatous inflammation and necrosis, with tendency towards cavitation.

- Nodules dimension ranges from few millimeters to masses of 10 cm;
- Margins are more commonly smooth than irregular;
- Nodules are frequently bilateral (75% of cases), with random distribution and no lobar predilection; occasionally they show a predominant or exclusive peripheral (subpleural) distribution, or, less commonly, have a peribronchovascular localization;
- The halo-sign (a rim of ground-glass opacity surrounding the pulmonary lesion, non specific) can be detected in up to 15% of nodules, and can be expression of perinodular hemorrhage and/or inflammation.
- After contrast media administration, a hypodensity of the core of the nodule can be identified as expression of central necrosis; in a small number of cases a feeding vessel can also be seen. Calcifications are rare;
- With progression of disease nodules tend to increase in number and size, and cavitation is common in nodules greater than 2 cm (50% of cases);
- Cavitations are usually thick-walled and show irregular inner margins; less frequently - and often after therapy - they can show thin and smooth walls; sometimes an air-full level can be detected.

Airspace consolidations and other parenchymal alterations:

The second most common radiological manifestation is represented by airspace consolidations (25%-50% of cases), which may occur with or without nodules and masses (Fig. 2 on page 16).

Patchy or diffuse ground-glass opacities are seen less commonly (20%-30% of cases). **Ground-glass opacity** (GGO) is defined as an hyperdensity which does not obscure underlying bronchial walls or vascular structures. GGO may correspond to partial or total alveolar filling, thickening of the alveolar septa with partial alveolar collapse, pulmonary fibrosis or an increase in size of the capillary vasculature. When occurring in ANCA-associate granulomatous Vasculitis, airspace consolidations or ground-glass opacities may reflect either vasculitic pulmonary disease in the form of pneumonitis or alveolar hemorrhage.

- Areas of consolidations usually show a random distribution; sometimes they appear as subpleural wedge-shaped lesions mimicking pulmonary infarcts, or have a peribronchial distribution;
Airspace consolidations are characterized by quite variable shapes and appearances, ranging from dense and localized alterations to bilateral, patchy or confluent consolidations, sometimes involving a whole lobe in extension (Fig. 3 on page 17);

- Bilateral, diffuse GG opacities can be seen in a small proportion of patients;

- In a small number of cases **centrilobular small nodules** (Fig. 4 on page 18) of variable density and **branching linear opacities** (« tree in bud » pattern), septal and intralobular linear opacities (**reticular pattern**) have been described;

- **Pleural effusions**, unilateral or bilateral and of variable extent are quite uncommon;

- In a small proportion of patients (15%) hilar and/or mediastial lymph node enlargement can be detected.

*Tracheal and bronchial involvement:*

**Tracheal** and **bronchial involvement** is a common manifestation in ANCA-associated granulomatous vasculitis, being reported in up to the 15%-25% and 40-70% of patients respectively.

- Tracheal stenoses are usually **subglottic** (this area should always be included in the study acquisition volume), can be smooth or irregular, most commonly circumferential and about 2-4 cm long (Fig. 5 on page 18 and Fig. 6 on page 19);

- The tracheal rings can show calcifications, thickening and cartilaginous erosions.

- Mucosal thickening maybe symmetrical or asymmetrical, irregular or ulcerated; a possible involvement of the vocal cords must be assessed (Fig. 7 on page 20);

- Segmental and subsegmental bronchial walls can be thickened and stenotic, resulting in possible airway obstruction and atelectasis (Fig. 8 on page 20 and Fig. 9 on page 21);

- Bronchiectasis (Fig. 10 on page 22) and patchy air trapping can be found in the 10%-20% and 30% of cases respectively.

*Imaging Follow-up:*

Follow-up studies after therapy (immunosuppressive treatment) can show, in the majority of cases, a complete resolution of GG opacities, of nodules and a reduction in dimension or complete resolution of the masses (Fig. 11 on page 23 and Fig. 12 on page 23);

- Cavitated lesions may develop in thin-walled cavitations. A reduction of tracheal or bronchial wall thickening can be observed as well (Fig. 13 on page 23).
Fibrotic changes (intralobular linear opacities and septal thickening) and bronchiectasis tend to remain stable.

**Differential Diagnosis:**

The main radiologic differential diagnoses include other diseases that may essentially result in multiple nodules and masses, with or without cavitations. The knowledge of the clinical background of the patient (upper respiratory symptoms, laboratory data suggesting a glomerulonephritis and presence of serum c-ANCA) is essential in the formulation of diagnosis.

- **Infections** (septic embolism, multiple lung abscesses). They are usually associated with bacteremia and tend to involve mainly the lower lobes and are rarely greater than 3 cm;

- **Neoplasms** (hematogenous metastases, lymphoma, Kaposi sarcoma when peribronchovascular lesions predominate). Metastases principally involve the lower lobes; cavitations are uncommon in lymphoma; the fast changes of nodules and masses dimensions and distribution demonstrate argue against malignancy;

- **Organizing pneumonia.**

**Churg-Strauss Syndrome:**

Churg-Strauss Syndrome is a very rare disease (annual incidence: 1-3/million), mainly affecting middle-aged patients (40-50 years old), clinically characterized by asthma and allergic rhinitis, peripheral hypereosinophilia and necrotizing vasculitis, virtually present in all cases.

**Radiological manifestations of the disease:**

The most common imaging feature (present in up to 90% of cases) is represented by bilateral areas of **GGO** or **consolidations**.

- Their distribution is usually symmetric, with a peripheral predominance and no lobar predilection; less commonly, they have a peribronchial or patchy random distribution (Fig. 14 on page 24);

- **Septal lines** can be seen in approximately 50% of cases; the interlobular septal thickening can be explained either by the presence of edema secondary to cardiac involvement or by an eosinophil infiltration of the septa;

- **Nodules** or **masses** with dimensions ranging from few millimeters to 3,5 cm can be uncommonly found, with or without association with a halo-sign or cavitations (very rare);
- Airway involvement is rare, consisting of centrilobular nodules, tree-in-bud pattern, bronchial dilatation, and bronchial and bronchiolar wall thickening; these alterations are related to asthma in most of the cases (which is almost always present), or, less commonly, to an eosinophilic involvement of the bronchial wall.

- Unilateral or bilateral pleural effusions are relatively frequent (10%-50% of cases) and may be related to heart failure resulting from cardiomyopathy or by eosinophilic pleuritis;

- Lymph node enlargement and pericardial effusion are uncommon findings.

**Imaging Follow-up:**

Follow-up studies after therapy show, in the majority of cases, a complete resolution of all parenchymal abnormalities (clinical remission achieved in more than 90% of patients); airways alterations (bronchial wall thickening and dilatations) tend to be less respondent to treatment.

**Differential Diagnosis:**

The main radiologic differential diagnoses include other diseases that may present with transient, patchy GGOs or consolidations without any lobar predilection in patients with a history of asthma. The differential diagnosis of Churg-Strauss Syndrome is essentially based on the knowledge of the systemic manifestations of disease, including rash, peripheral neuropathy and presence of serum p-ANCA.

- **Simple and chronic pulmonary eosinophilic pneumonia:** their radiological manifestations can be very similar to those of Churg-Strauss Syndrome, with patchy, transient and migratory non segmental GGOs or consolidations, associated with eosinophilia. In case of chronic pulmonary eosinophilic pneumonia, the consolidations would involve only or predominantly the outer thirds of the lungs.

- **Allergic bronchopulmonary aspergillosis:**

- **Infectious pneumonia** (bacterial, fungal, viral); mainly represented by bacterial bronchopneumonia and opportunistic agents, such as *Pneumocystis Jiroveci* and *Cytomegalovirus* (especially in asthmatic patients under corticosteroids therapy);

- **Organising Pneumonia.**

**Microscopic Polyangiitis:**

Microscopic Polyangiitis is a necrotizing non granulomatous vasculitis which represents the first cause of pulmonary-renal syndrome. Its incidence is estimated at 1/100.000/
year with a slight male predominance and mean age at onset of about 50 years. The main clinical features are represented by rapidly progressive glomerulonephritis (90% of cases) and **diffuse alveolar hemorrhage** (DAH, 10%-30% of cases). DAH can be defined by the presence of hemoptysis, diffuse alveolar infiltrates, and a drop in hematocrit level. The most common histologic finding in patients with DAH is capillaritis.

**Radiological manifestations of the disease:**

Imaging features of Microscopic Polyangiitis at CT, nonspecific, are those characterizing a DAH, with GG opacities as the most common radiological manifestation.

- Bilateral (less commonly even unilateral), patchy or diffuse **GGOs** (partial alveolar blood filling; 90% of cases), airspace **consolidations** (complete alveolar blood filling; 70% of cases) with no predominant distribution and thickening of the broncho-vascular bundles (51% of cases).

- Poorly defined or GG **centrilobular nodules** may be predominant in some patients, representing localized foci of hemorrhage;

- After an acute episode of hemorrhage, the hemosiderin-laden macrophages accumulate in the interstitium, causing **interlobular septal thickening** in association with ground-glass opacity (**crazy-paving** pattern);

- As for the other ANCA-associated vasculitis, a non fortuitous association with pulmonary fibrosis has been recently demonstrated, characterized by a persistent reticular pattern, areas of peripheral honeycombing and traction bronchiectasis (**Fig. 15** on page 25);

- **Pleural effusions** can be detected in up to the 15% of cases; **pulmonary edema** in the 6% of cases.

**Imaging Follow-up:**

After therapy, the most of the patients show a complete resolution or a reduction of GGOs and parenchymal consolidations. Septal thickening and mild fibrotic changes may persist in severe and reiterated presentations.

**Differential Diagnosis:**

The diagnosis of Microscopic Polyangiitis should be suspected in patients with rapidly progressive glomerulonephritis, serum p-ANCA and radiological manifestations of hemorrhage.

The main radiological differential diagnosis comprehends all conditions possibly causing alveolar hemorrhage.
- Most cases of DAH are generally caused by capillaritis associated with systemic autoimmune diseases such as **ANCA-associated small-vessel vasculitis**, **Goodpasture syndrome**, and **systemic Lupus Erythematosus**;

- **Hypersensitivity pneumonitis** when the predominant pattern is represented by centrolobular poorly defined or GG nodules.

**Goodpasture Syndrome:**

Goodpasture syndrome is extremely rare (incidence 1/1,000,000/year), it's more common in men than in women with an onset between 20-30 years of age. It is characterized by anti-glomerular basement membrane antibodies (more than 90% of cases), directed against basement membrane antigens of the alveolus and glomerulus.

The most common presenting symptom is hemoptysis, sometimes lifethreatening.

**Radiological manifestations of the disease:**

The radiological manifestations correspond to the typical imaging features of a DAH, with **bilateral GGOs** (Fig. 16 on page 25) and less commonly areas of airspace consolidations.

- GGOs can have a diffuse or patchy distribution.

- Less commonly, small, poorly defined or ground-glass **centrilobular nodules** (Fig. 17 on page 26) and **interlobular septal thickening** can be found.

**Imaging Follow-up:**

Treatment options are given by plasmapheresis and immunosuppressive drugs (corticosteroids and cyclophosphamide). After successful treatment, imaging follow-up can show a complete resolution or reduction of GGOs and parenchymal consolidations. Septal thickening and mild fibrotic changes may persist in severe and reiterated presentations. When a CT scan is performed after 2-3 days from the acute episode, a prevalence of the centrilobular nodules and interlobular septal thickening pattern can be found.

**Differential Diagnosis:**

Goodpasture syndrome should always be suspected in a patient with hemoptysis, bilateral airspace consolidations or GGO and renal disease. The presence of anti-glomerular basement membrane antibodies would confirm the diagnosis.
The radiological differential diagnosis includes all conditions which can cause pulmonary hemorrhage associated with a renal involvement.

- **Connective tissue diseases** (especially Systemic Lupus Erythematosus);

- Other **systemic vasculitis** (especially ANCA-associated granulomatous vasculitis and Microscopic Polyangiitis);

- **Blood aspiration**;

- **Metastasis** from highly vascularized tumors, such as choriocarcinoma.

**Behçet's Disease:**

Behçet's Disease is an uncommon systemic disorder (prevalence of about 1/100.000) characterized by vasculitis and a clinical triad of recurrent oral and genital ulcers and relapsing uveitis. Mean age of onset is about 30 years; men are much more commonly affected than women. Pleuropulmonary involvement is quite infrequent (1%-18% of cases) and mainly characterized by pulmonary arteries aneurysms, hemorrhage and parenchymal infarcts. The principal clinical manifestation is given by hemoptyis, sometimes massive and life threatening. The **Hughes-Stovin syndrome** is considered by many authors as a rare variant of Behçet's Disease, characterized by systemic venous thrombi and pulmonary aneurysms and thrombosis without oral and genital ulcerations and eye involvement.

**Radiological manifestations of the disease:**

Chest-CT with contrast media administration allows the evaluation of presence, location and size of pulmonary aneurysms, their possible complications (thrombosis, pulmonary infarcts) and other parenchymal and vascular associated findings.

- **Pulmonary aneurysms** are, in the most of cases, multiple, bilateral, fusiform or saccular and their dimensions range from 1 to 7 cm in diameter (Fig. 18 on page 26);

- The most common localization of pulmonary aneurysms is the right interlobar artery, followed by lobar and segmental arteries;

- The affected vessel can show signs of inflammation, as **wall-thickening** and **enhancement** after contrast media administration;

- Partial or complete **thrombosis** of the aneurysms is a frequent finding; such occlusions can lead to localized areas of consolidations (wedge-shaped peripheral consolidations, rarely with cavitations) related to **parenchymal infarctions**, focal **atelectasis** and areas of **oligemia**;
- Vasculitis and rupture of pulmonary arteries can lead to alveolar hemorrhage, with focal, patchy or diffused areas of **consolidations** or **GGOs** (Fig. 19 on page 27);

- Associated findings are given by **recurrent pneumonia** and **organizing pneumonia**;

- Unilateral or bilateral **pleural effusions**, as a result of pleural vasculitis, pulmonary infarcts or thrombosis occurring in the superior vena cava.

**Imaging Follow-up:**

Repeated scans over time allow the monitoring of aneurysms formation and the follow-up of their parenchymal complications. Patients receiving immunosuppressive treatment can show reduction or complete resolution (up to the 75% of cases) of pulmonary aneurysms and associated thrombosis (Fig. 20 on page 28).

**Differential Diagnosis:**

Pulmonary artery aneurysms are an uncommon finding, which can be given by a small number of conditions.

- **Infections** (especially mycotic pseudoaneurysms);

- Previous **trauma** (often iatrogenic);

- **Takayasu Arteritis**.

**Takayasu Arteritis:**

Takayasu Arteritis is characterized by a chronic, granulomatous vasculitis mainly affecting aorta and its main branches; the main vascular alterations are given by stenosis or occlusion with possible post-stenotic dilatations and aneurysm formation. Distribution of lesions is usually segmental and patchy. Involvement of the pulmonary arteries can be found in about the 50%-80% of cases, as a late manifestation of disease. Although more common in Japan and Southeast Asia, the disease has been found worldwide, usually affecting young women (10-40 years old). To limit radiation exposure, MRI should be the modality of choice in suspected or proved disease.

**Radiological manifestations of the disease:**

The most common pulmonary manifestation of disease is given by **stenosis or occlusion** and **aneurysm formation** of segmental or subsegmental pulmonary artery branches (less commonly of the lobar or main pulmonary arteries);
- The most frequent localization is the right upper lobe, followed by the middle lobe, the lingula, and the lower lobes (Fig. 21 on page 29 and Fig. 22 on page 30);

- Affected vessels can show wall-thickening and contrast-enhancement in early phases, calcium deposition in chronic phases;

- Vascular alterations can be associated to parenchymal areas of oligemia or pulmonary infarcts.

**Imaging Follow-up:**

Vascular lesions tend to progress over time. A control of disease can be achieved with high-doses of corticosteroids or with cytotoxic drugs in patients non responding to corticosteroids and with frequent relapses.

**Differential Diagnosis:**

The main differential diagnosis for the pulmonary manifestations of disease is given by Behçet's Disease. Integration with the clinical background of the patient and histologic findings are necessary for diagnosis.
Fig. 1: ANCA-associated granulomatous vasculitis in a patient with pulmonary emphysema: axial (a) and coronal (b) CT reformations show two masses with spiculated contours localized in the upper lobes and surrounded by paraseptal and centrilobular emphysematous lesions.

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Fig. 2: ANCA-associated granulomatous vasculitis in a 23 years old patient with multiple nodules, masses (some of which cavitated) and parenchymal consolidations with peripheral and peribronchovascular distribution; most of the lesions are surrounded by a rim of ground-glass opacity ("halo-sign")

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**Fig. 3:** Coronal (a) and axial (b) CT reformations in a patient with ANCA-associated granulomatous vasculitis and pulmonary hemorrhage: bilateral, confluent parenchymal consolidations and ground-glass opacities predominantly distributed in the upper lobes, associated with nodules

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**Fig. 4:** CT study in a patient with ANCA-associated granulomatous vasculitis and recurrent episodes of hemoptysis showing diffuse centrolobular ground-glass opacities

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Fig. 5: Tracheal involvement in a patient with ANCA-associated granulomatous vasculitis: CT shows an irregular mucosal thickening with a multinodular appearance of the inner surface of the trachea as seen on parenchymal (a) and mediastinal (b) window; virtual bronchoscopy confirmed the micronodular pattern (c)

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Fig. 6: Patient affected by ANCA-associated granulomatous vasculitis with airways involvement: CT study demonstrated a subglottic concentric stenosis of the tracheal
lumen evaluated by virtual bronchoscopy (a, retroscopic view from the carena) and Volume Rendering reformations (b)

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Fig. 7: Airways involvement in a patient with ANCA-associated granulomatous vasculitis reporting an asymmetrical tracheal wall thickening (a) and a subglottic stenosis (b)

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**Fig. 8:** Patient with ANCA-associated granulomatous vasculitis: stenosis and irregularity of the inner surface of the right lobar bronchi on a coronal reformation (a) with a diffuse contrast-enhancement of the bronchial walls after intravenous administration (b), reflecting active inflammation.

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Fig. 9: Bronchial airways involvement in a patient with ANCA-associated granulomatous vasculitis: focal concentric stenosis of the origin of the left upper lobar bronchus (a, arrow) and occlusion of the segmental bronchus for the superior segment of the left lower lobe (arrow) with peripheral peribronchovascular consolidations (b)

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Fig. 10: Segmental and subsegmental bronchiectasis and distal airspace consolidations (same patient of fig. 8) of the right lower lobe (a) and middle lobe (b) as a consequence of chronic stenosis/occlusion of the proximal bronchi; a stenosis of the lobar middle bronchus is also demonstrated (b, arrow)
**Fig. 11:** Follow-up evaluation in a patient with ANCA-associated granulomatous vasculitis after immunosuppressive treatment (same patient of fig. 1): follow-up evaluation shows an almost complete resolution with residual cicatrical/fibrotic changes (b) of the bilateral masses localized in the upper lobes (a).

**Fig. 12:** Follow-up evaluation in a patient with ANCA-associated granulomatous vasculitis (same patient of fig. 2) after immunosuppressive treatment: follow-up evaluation (b) shows a resolution of the peribronchovascular consolidation of the right upper lobe (big arrow) and the cavitated mass of the left upper lobe (small arrow) with residual cicatrical/fibrotic changes (b).
**Fig. 13:** Follow-up evaluation in a patient affected by ANCA-associated granulomatous vasculitis with symmetrical, circumferential tracheal involvement at baseline (a): follow-up study demonstrated an almost complete resolution after immunosuppressive therapy (b)

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**Fig. 14:** CT study in a patient with Churg-Strauss syndrome showing areas of randomly distributed, patchy airspace consolidations and ground glass opacities, mainly localized in the right upper lobe
Fig. 15: Patient affected by Microscopic Polyangiitis: bilateral, peripheral and subpleural honey-combing predominantly localized in the lower lobes suggestive of UIP (Usual Interstitial Pneumonia)
**Fig. 16:** Patient with Goodpasture syndrome and pulmonary hemorrhage (recurrent hemoptysis): CT study demonstrates bilateral areas of ground-opacities having medullary distribution within the upper lobes

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![CT images showing ground-opacities in upper lobes](image)

**Fig. 17:** Patient affected by Goodpasture syndrome with hemoptysis and pulmonary hemorrhage: small, ill-defined centrilobular nodules diffusely and homogeneously distributed throughout both lungs

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![Small ill-defined nodules in both lungs](image)
Fig. 18: CT study in a patient affected by Behçet’s disease: axial reformations show a thrombosed pulmonary aneurysm of the left lower branch and an aneurysm of the right lower branch partially thrombosed (a, b); MIP reformations (c) allow an overview of the vascular abnormalities demonstrating a distal occlusion of the right aneurysm with a peripheral wedge-shaped consolidation reflecting pulmonary infarction (small arrow)

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Fig. 19: Parenchymal abnormalities in Behçet’s disease (same patient of figure 18): CT study on parenchymal window shows, at the level of the carena (a), a focal perihilar consolidation surrounded by a ground-glass opacity (big arrow) next to a small aneurysm of a proximal branch of the right pulmonary artery (small arrow); an other focal consolidation surrounded by a ground-glass opacity (big arrow) and a wedge-shaped subpleural consolidation are demonstrated in the lower lobes (b), reflecting respectively pulmonary hemorrhage and infarction

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Fig. 20: Follow-up evaluation in a patient with Behçet's disease: a focal pulmonary aneurysm of a right segmental branch (a) showed a complete resolution after immunosuppressive treatment (b)

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Fig. 21: CT study in a patient affected by Takayasu arteritis: aneurysm of the left pulmonary artery (big arrow) and a mild stenosis of the right pulmonary artery with complete occlusion of the right superior and middle branches (small arrow)

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**Fig. 22:** CT evaluation in a patient affected by Takayasu arteritis reported a focal aneurysm of the pulmonary trunk (arrow)

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Conclusion

Imaging features of pulmonary involvement in systemic vasculitis can be extremely heterogeneous and the differential diagnosis very challenging. Nevertheless, every vasculitis can be characterized by peculiar combinations of clinical and radiological signs; only the integration between the clinical, serological and pathological data derived from a close cooperation with the referring physicians and an appropriate knowledge of the specific radiological patterns of each vasculitis allows the formulation of the possible diagnosis.
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