Vasculitis: variety of findings in chest.

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Learning objectives

Vasculitis is a group of diseases in which inflammation of the vessel wall occurs causing the destructuring of the blood vessels. Lungs are commonly affects and involvement can be from the main pulmonary arteries to the alveolar capillaries. Pulmonary vasculitis includes various pathologies and there are no specific radiological findings. The combination of symptoms, signs, laboratory data and certain image patterns, can help to guide the diagnosis. In this paper, a review of the different forms of presentation of thoracic involvement in vasculitis is performed.

Background

Classically, vasculitis has been classified according to the histological findings.

At the conference in Chapel Hill, a new classification based on the size of the vessels involved and the presence of ANCA was proposed. Fig. 1 on page 2

Drawing on this classification, the different entities that produce thoracic involvement is discussed.

In primary vasculitis of large vessels such as Takayasu arteritis there is thoracic involvement. In giant cell arteritis may exist chest involvement but in this entity as in polyarteritis nodosa and Kawasaki disease usually have no lung involvement.

Moreover, within the entities included in the classification of Chapel Hill, the lung is involved more often in those idiopathic primary small vessel vasculitis associated with ANCA (Wegener granulomatosis now known as granulomatosis with polyangiitis, microscopic polyangiitis and Churg-Strauss syndrome).

There are other vasculitis (not initially included in this classification) that also produce pulmonary disease, such as systemic autoimmune diseases, for example, Behçet's disease and systemic lupus erythematosus.

Images for this section:
Fig. 1: * Some vasculitis of small and large vessels can compromise the vessels of medium caliber, but vasculitis of large and medium vessels do not affect smaller vessels than the arteries.
Findings and procedure details

LARGE VESSEL VASCULITIS # THORACIC INVOLVEMENT:

Takayasu arteritis:

This entity causes chronic idiopathic stenosis of large vessels with a predilection for the aorta, its branches and it may also affect the pulmonary arteries. This disease is more common in Asia and usually affects young women.

It is characterized by granulomatous inflammation of the arterial wall with intimal proliferation, fibrosis of the media and adventitia, which may develop stenosis, occlusion, dilatation and poststenotic aneurysm formation.

# Radiological findings:

- Irregular contour of the descending aorta, linear calcification of the aortic wall, aortic arch ectatic, cardiomegaly and decreased of the pulmonary vasculature.

- On unenhanced CT # vascular wall (aorta or pulmonary) hyperdense, with or without calcifications. The mediastinal fat adjacent of the vessel may be involved as a sign of active inflammatory process.

- Contrast CT:

  # In inflammatory phase, there are wall thickening of pathological vessel (1-4 mm) and contrast enhancement. Fig. 2 on page 11

  # In fibrotic phase, the wall does not enhance after contrast administration. Stenosis, aneurysmal dilation or occlusion of segmental or subsegmental pulmonary arteries can be seen which may be accompanied by vascular mosaic pattern in lung parenchyma.

- The RM also provides information on the activity of Takayasu's disease, showing contrast enhancement in the inflammatory phase.

- Positron emission tomography (PET) shows greater fluorodeoxyglucose uptake by the diseased arteries.
# Differential Diagnosis:

Atherosclerosis, Buerger's disease, giant cell arteritis and syphilitic aortitis

**Giant cell arteritis (GCA):**

Giant cell arteritis is a granulomatous vasculitis that affects large and medium vessels as the aorta, its branches and branches of extracranial carotid mainly the temporal artery Fig. 3 on page 12. The inflammation occurs in the middle layers and adventitia with presence of giant cells, causing destruction by fibrinoid necrosis of the vessel wall.

This entity occurs in elderly patients (over 50 years) and is often associated with polymyalgia rheumatica.

Symptoms that may occur in addition to the classic clinical picture (thickening of temporal artery, temporal headache, jaw claudication and visual loss) are: fever, fatigue, dry cough, pleuritic chest pain and dyspnea.

**Radiological findings:**

- When there is involvement of the aorta, the findings are similar to those described in Takayasu's disease: presence of wall thickening Fig. 4 on page 13 and Fig. 5 on page 14, stenosis and possibility of developing a thoracic aortic aneurysm. However, interpretation of results is more difficult than in Takayasu arteritis because GCA affects elderly patients with concomitant arteriosclerosis.

- Pulmonary involvement is a rare manifestation of GCA. Presences of pulmonary nodules or pulmonary infiltrates due to alveolar hemorrhage have been described.

**Behçet disease:**

Behçet's disease has been included in this section because it affects large vessels, although this is considered variable vessel vasculitis in the International Chapel Hill Consensus.

It mainly affects men from Middle East and over 20-30 years. Clinical findings are: recurrent aphthous ulcers, skin lesions, genital ulcers and uveitis. The transmural
vascular inflammation with fibrosis or thinning of muscular and elastic arteries may form aneurysms. Venous or arterial thrombi can be seen.

**# Radiological findings:**

a) Aneurysms of the pulmonary artery:

- Single or multiple, unilateral or bilateral, which may be thrombosed.

- The presence of airspace consolidation in perianeurysmatic location is indicative of impending rupture.

b) Arterial thrombotic occlusion of the pulmonary vasculature Fig. 6 on page 15:

- Secondary parenchymal abnormalities: infarcts, pulmonary oligohemia, atelectasis and pleural effusions associated with heart attacks.

c) Thrombotic occlusion of the superior vena cava.

d) Pulmonary hemorrhage secondary to vasculitis.

**MEDIUM VESSEL VASCULITIS:**

This group includes **polyarteritis nodosa** and **Kawasaki disease** in which there is involvement of medium caliber vessels, mainly visceral arteries (mesenteric arteries, liver, kidney ...) and coronary arteries producing aneurysmal dilatation of them Fig. 7 on page 16. **Lung affectation is very rare.**

**SMALL VESSEL VASCULITIS # PULMONARY INVOLVEMENT:**

**ANCA-associated granulomatous vasculitis:**

As discussed previously, the lung is involved more often in those idiopathic primary small vessel vasculitis associated with ANCA (Wegener granulomatosis now known as granulomatosis with polyangiitis, Churg-Strauss and microscopic polyangiitis).
This group includes those vasculitis involving small vessels such as arterioles, capillaries and venules. In these cases find cutaneous manifestations such as purple, glomerulonephritis and pulmonary hemorrhage by capillary involvement.

**Granulomatosis with polyangiitis - Wegener's granulomatosis:**

It is characterized by *necrotizing granulomatous inflammation involving the upper respiratory tract* (sinusitis, otitis, ulcers ...), *lower respiratory tract* (cough, chest pain, dyspnea and hemoptysis) and *renal glomerulonephritis* (hematuria, proteinuria and azotemia).

**# Radiological findings:**

a) Pulmonary nodules and masses are common findings in patients with Wegener's granulomatosis (up to 70% of patients at the time of diagnosis or during the course of the disease).

- They correspond to areas of parenchymal necrosis, granulomatous inflammation and vasculitis.

- The lesions may be single or multiple (generally less than 10), with variable size and randomly distributed Fig. 8 on page 17 Fig. 9 on page 18 Fig. 10 on page 19. Up to 50% may cavitate. Fig. 11 on page 20

- Nodules may show a ground glass surrounding area (halo sign) Fig. 11 on page 20, which represents adjacent parenchymal hemorrhage or an inverted halo (sign atoll) due to the presence of an area of organizing pneumonia in the periphery of the focal hemorrhage.

- With treatment, the nodules usually resolve. Complete resolution may not occur and residual areas of scarring can be seen.

b) Airspace consolidation, patchy infiltrates or, less frequently, diffuse ground-glass opacities are the second most common radiographic finding.

- Consolidations and ground-glass opacities reflect pneumonitis or alveolar hemorrhage. They may have a peripheral or peribronchial distribution.

- The presence of bilateral and diffuse ground-glass opacities represent a diffuse alveolar hemorrhage with capillaritis and they appear in the clinical context of dyspnea, hemoptysis and anemia Fig. 12 on page 21
c) Lower airway involvement:

- It usually manifests as involvement of a short segment of the trachea (more frequently in the subglottic portion).
- Circumferential wall thickening, smooth or nodular, including membranous pars is observed.
- Bronchial wall thickening and bronchiectasis can be seen.

# Differential Diagnosis Table 1 on page 22

**Churg-Strauss syndrome**

The Churg-Strauss disease or allergic granulomatosis with angiitis, is clinically characterized by asthma, fever and eosinophilia. In the histological study, there are granulomatous inflammation with abundant eosinophils and necrotizing vasculitis affecting vessels of small and medium caliber, mainly in the lung.

Histopathologic lung presentation can combine extravascular presence of granulomas, vasculitis and eosinophilic pneumonia.

**# Radiological findings:**

a) Airspace consolidation:

- In most cases, the alterations include patchy, no segmental, transient and predominantly peripheral airspace consolidation or ground glass areas Fig. 13 on page 23.

b) Pulmonary nodules:

- They are sometimes bilateral, small or large, which can be coalescent. Cavitation is rare.

c) Pleural effusion:

- Approximately 30% of patients presented unilateral or bilateral pleural effusion.
**# Differential Diagnosis:**

Chronic eosinophilic pneumonia, simple eosinophilic pneumonia (Löffler syndrome).

**Microscopic polyangiitis (MPA)**

The MPA is a granulomatous vasculitis of small vessel not associated with the presence of ANCA.

MPA is the most common pulmonary-renal syndrome and it can simultaneously affect other organs such as the nervous or musculoskeletal system, gastrointestinal tract or the heart.

**# Radiological findings:**

a) Consolidation of airspace:

- Bilateral patchy ground-glass opacities representing alveolar hemorrhage with capillaritis Fig. 14 on page 24.
- Predominate in the perihilar regions, middle and lower lung fields, respecting the apex and costrophrenic angles.
- There may be thickening of interlobular septa acquiring cobblestone pattern Fig. 15 on page 25.

b) Pulmonary nodules:

- In some cases with halo sign representing hemorrhagic nature of these lesions.

c) Vascular tree in bud, with the presence of centrilobular nodules Fig. 16 on page 26.

d) It may be changes of pulmonary fibrosis in relation to repeat bleeding.

e) Pleural effusion is a rare finding.

**# Differential Diagnosis:**
Goodpasture’s syndrome, other ANCA associated vasculitis (In MPA, the involvement of sinus and upper respiratory tract is rare) and systemic lupus erythematosus.

**Cryoglobulinemic vasculitis:**

Approximately 2% of patients with cryoglobulinemic vasculitis have pulmonary vasculitis, which usually manifests as a diffuse alveolar hemorrhage.

**Henoch-Schönlein purpura:**

Pulmonary involvement is rare and is caused by necrotizing capillaritis. Ig A deposit in the alveolar basement membrane causes an immune mediated pneumonitis and leukocytoclastic capillaritis. It is commonly manifested as diffuse alveolar hemorrhage.

**Cutaneous leukocytoclastic angiitis:**

Pulmonary involvement is rare.

Summary table of the thoracic involvement in vasculitis Fig. 17

Images for this section:
**Fig. 1:** * Some vasculitis of small and large vessels can compromise the vessels of medium caliber, but vasculitis of large and medium vessels do not affect smaller vessels than the arteries.
Fig. 2: Contrast-enhanced axial chest CT using the mediastinal window settings shows mural thickening and contrast enhancement of the thoracic aorta.
Fig. 3: Doppler ultrasound study shows hypoechoic thickening of the wall of the temporal artery in a patient with temporal arteritis.
Fig. 4: Axial section chest CT, mediastinal window. 77 year old male patient diagnosed with GCA. See the wall thickening of the ascending thoracic aorta.
**Fig. 5:** Axial section chest CT, mediastinal window. Sequence without contrast, early and late arterial phase respectively. See the progressive contrast enhancement of the ascending thoracic aorta (being more evident in late phase) correlates with inflammatory aortitis.
Fig. 6: Forms of large vessel vasculitis. The third image shows thrombotic occlusion of the main pulmonary arteries in Behçet disease.
Fig. 7: Contrast-enhanced axial chest CT, long axis-2 cameras and 3D Volume Rendering reconstructions. 43 year old woman with a history of Kawasaki disease in childhood. See the presence of aneurysms in the right coronary artery.
Fig. 8: 77 year old female with Wegener’s granulomatosis. CT thorax, lung window (axial view) shows pulmonary nodules and subpleural airspace consolidation in right lower lobe.
**Fig. 9:** CT thorax, lung window (axial view). Lung nodule in a patient with Wegener disease.
Fig. 10: PET-CT study. The pulmonary nodule has high SUV but a repeated image after 30 minutes shows a drop of activity of 30% which is quite unusual for cancer.
Fig. 11: CT thorax, lung window (axial view). During evolution, the nodule cavitates showing a thick and irregular wall. Also there is a ground glass halo around it.
Fig. 12: Fig 3 and Fig. 4: Chest radiograph and CT thorax, lung window (axial view). Pulmonary hemorrhage and cavitated nodules (red arrow) in a patient with Wegener's granulomatosis.
<table>
<thead>
<tr>
<th>Pathology</th>
<th>Nodules</th>
<th>Size</th>
<th>Distribution</th>
<th>Cavitation</th>
<th>Other findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>Wegener's granulomatosis</td>
<td>Multiple</td>
<td>A few millimeters to 10 cm</td>
<td>Bilateral and aleatory, can be peribronchovascular</td>
<td>In 50% of lesions &gt; 2 cm</td>
<td>Halo or atoll signs</td>
</tr>
<tr>
<td>Metastasis</td>
<td>Multiple</td>
<td>Variable</td>
<td>Bilateral and aleatory</td>
<td>Unusual, except squamous primary tumor, sarcoma and transitional cell</td>
<td>Lymphadenopathy</td>
</tr>
<tr>
<td>Infection</td>
<td>Variable</td>
<td>Less than 10 mm</td>
<td>Peripheric or miliary</td>
<td>Rare</td>
<td>Consolidation, tree in bud</td>
</tr>
<tr>
<td>Sarcoidosis</td>
<td>Multiple</td>
<td>2–10 mm</td>
<td>Perilymphatics</td>
<td>Rare</td>
<td>Distortion of lung architecture, symmetrical lymphadenopathy</td>
</tr>
<tr>
<td>Rheumatoid arthritis</td>
<td>Solitary or Multiple</td>
<td>5–7 mm</td>
<td>Peripherics</td>
<td>Less common</td>
<td>Subcutaneous nodules</td>
</tr>
</tbody>
</table>

**Table 1:** Differential Diagnosis of multiple pulmonary nodules.
Fig. 13: CT paranasal sinuses without contrast and CT thorax, lung window (axial view). Woman of 54 years, history of asthma and eosinophilia. Images show sinus abnormality and patchy pulmonary infiltrates in patient with p-ANCA antibodies positive, the diagnosis was Churg-Strauss disease.
Fig. 14: Chest radiograph and CT thorax, lung window (axial view). 77 year old woman with patchy bilateral pulmonary infiltrates, renal failure (biopsy: necrotizing glomerulonephritis), gastrointestinal symptoms (diarrhea) and pANCA +, diagnosis of MPA.
Fig. 15: Chest radiograph and CT thorax, lung window (axial view). Airspace consolidation and ground glass pulmonary opacities with interlobular septa thickening ("crazy paving" area) mainly in the right upper lobe findings consistent with alveolar hemorrhage in a patient with pulmonary-renal syndrome and anti-MPO-ANCA IgG, the diagnosis was MPA.
**Fig. 16:** CT thorax, lung window (axial view). 70 year old woman diagnosed with MPA. The scan shows centrilobular nodules and presence of vascular tree in bud pattern (see magnified image at right).
## LARGE VESSEL VASCULITIS

<table>
<thead>
<tr>
<th>Condition</th>
<th>Characteristics</th>
</tr>
</thead>
</table>
| Takayasu arteritis | - Women under 50 years.  
- Granulomatous inflammation of the aorta and its branches.  
- It can affect the pulmonary arteries.  
- **Image:**  
  - Thickening and enhancement of the affected vessel wall. |
| Temporal arteritis  | - Patients over 50 years.  
- Associated with rheumatic polymyalgia  
- Granulomatous arteritis of the aorta and its branches, mainly the temporal artery.  
- Do not usually affect the pulmonary arteries.  
- **Image:**  
  - Thickening and enhancement of the affected vessel wall. |

## MEDIUM VESSEL VASCULITIS

<table>
<thead>
<tr>
<th>Condition</th>
<th>Characteristics</th>
</tr>
</thead>
</table>
| Polyarteritis nodosa | - Necrotizing inflammation of arteries of medium and small size.  
- There is usually no pulmonary involvement. |
| Kawasaki disease   | - It often affects children.  
- Arteritis with involvement of small, medium and large arteries.  
- The coronary arteries are often affected, with aneurysmal dilatation. There may be involvement of the aorta and its branches. There are usually no pulmonary involvement. |

## SMALL VESSEL VASCULITIS

<table>
<thead>
<tr>
<th>Condition</th>
<th>Characteristics</th>
</tr>
</thead>
</table>
| Wegener's granulomatosis   | - Granulomatous inflammation and necrotizing vasculitis of small and medium vessels.  
- Often necrotizing glomerulonephritis.  
- c-ANCA +  
- **Image:**  
  - Single or multiple pulmonary nodules that may be cavitate.  
  - Patchy or diffuse ground-glass pulmonary opacities.  
  - Lower airway involvement |
| Churg-Strauss syndrome     | - Granulomatous inflammation rich in eosinophils and necrotizing vasculitis affecting vessels of small and medium-sized  
- Associates asthma, eosinophilia and involvement of the upper respiratory tract.  
- p-ANCA +  
- **Image:**  
  - Consolidation or patchy ground-glass pulmonary opacities (non-segmental, transient and peripheral distribution).  
  - Non-cavitated pulmonary nodules. |
| Microscopic polyangiitis   | - Necrotizing vasculitis with few or no immune deposits that affects small vessels (capillaries, venules or arterioles).  
- Necrotizing glomerulonephritis  
- Most common pulmonary-renal syndrome  
- p-ANCA +  
- **Image:**  
  - Bilateral patchy alveolar consolidations or ground-glass opacities → alveolar hemorrhage.  
  - Pulmonary nodules that may have the halo sign. |

**Fig. 17:** Summary table of the thoracic involvement in vasculitis
Conclusion

Thoracic involvement is a common manifestation in this group of systemic diseases. Although there are no specific radiological findings, it is important to know the different forms of presentation and, with the clinical guidance, suspect the diagnosis.

Personal information

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References


