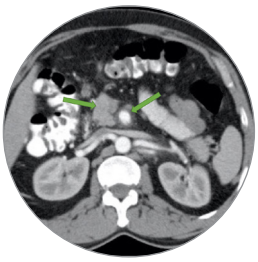

Imaging of Pulmonary and Abdominal Manifestations of Primary Vasculitis

Manifestaciones imagenológicas pulmonares y abdominales de las vasculitis primarias

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Key words (MeSH)
Vasculitis
Takayasu Arteritis
Granulomatosis with polyangiitis
Polyarteritis nodosa
Poliangeítis microscópica
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Churg-Strauss syndrome

Palabras clave (DeCS)
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Síndrome de Behçet
Síndrome de Churg-Strauss.

Summary

Vasculitis are a group of diseases that are characterized by the presence of inflammation in and around the wall of the blood vessels. This produces necrosis and inflammation where the extension of the injury depends on the size and location of the affected vessel. They can have manifestations in any organ, so they are considered systemic diseases. Primary vasculitis are uncommon pathologies and difficult to diagnose because its signs and symptoms are nonspecific and are easily confused with many other disease processes such as infectious, neoplastic or connective tissue. Because of this, it is important to combine clinical, radiological, histopathological and laboratory findings to achieve a correct diagnosis. In this review, we will focus on the role of imaging findings on the diagnosis and monitoring for pulmonary and abdominal manifestations of the most frequent vasculitis.

Resumen

Las vasculitis son un grupo de enfermedades que consisten en la inflamación dentro y alrededor de la pared de los vasos sanguíneos, lo que produce necrosis, y su extensión depende del tamaño y ubicación del vaso afectado. Se pueden manifestar en cualquier órgano, por lo que se consideran enfermedades sistémicas. Las vasculitis primarias son patologías poco comunes y difíciles de diagnosticar, porque sus signos y síntomas son inespecíficos y pueden confundirse con muchas otras patologías —como enfermedades infecciosas, neoplásicas o del tejido conjuntivo—. Por esta razón, para su diagnóstico es importante combinar los hallazgos clínicos, radiológicos, de estudios histopatológicos y de laboratorio. Esta revisión se enfocará en el papel de los hallazgos imagenológicos en el diagnóstico y monitoreo de las manifestaciones pulmonares y abdominales de las vasculitis más frecuentes.

1. Introduction

Vasculitides are a group of diseases characterized by the destruction of blood vessels due to inflammation and necrosis in and around their walls (1); they are considered systemic diseases because they can produce diffuse or local changes in any organ.

There are many types of vasculitis and their classification is a challenge (2). A first classification organizes them into two broad categories according to the underlying pathophysiological process: primary and secondary vasculitis. Primary vasculitides are those in which inflammation occurs mainly within blood vessels, while secondary vasculitides are caused by pathologies such as connective tissue diseases, tumors and infections (3). Primary vasculitides are uncommon (4, 5) and the correct diagnosis of these pathologies involves overcoming the difficulties derived from the great variability of non-specific signs and symptoms and depend on the size and location of the affected vessel.

The most appropriate way to study primary vasculitis is to classify them according to the type of vessel involved: small, medium or large caliber vessels (2). Large vessel vasculitides include those that affect the aorta and its main branches, medium vessel vasculitides involve the arteries of each organ and small vessel vasculitides affect arterioles, venules and capillaries (6). Currently, the most commonly used classification is the 2012 Chapel Hill International Consensus Conference vasculitis nomenclature update, which changed the names and definitions according to scientific advances and added important categories of vasculitis not included in the first version of 1994 (Figure 1) (7, 8).

Knowing the systemic manifestations of patients with vasculitis can help to establish a diagnosis (9). To reach an accurate diagnosis it is important to combine clinical, radiological, histopathological and laboratory findings. In the following, the radiological findings of vasculitis that occur when there is pulmonary and abdominal involvement will be discussed.

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2. Vasculitis of large vessels

2.1 Takayasu's arteritis

Takayasu's arteritis is a granulomatous vasculitis of large vessels, prevalent in young women and Asian populations (10). The estimated annual incidence is 0.12 to 0.26 cases/100,000 (11).

This pathology usually begins with general malaise, fever, night sweats, weight loss, arthralgia and reduced or absent peripheral pulses (10, 12). It is characterized by granulomatous inflammation of the arterial wall, with proliferation of the intima and fibrosis of the media and adventitia, which generates an irregularly distributed thickening of the vessel walls, eventually leading to stenosis, occlusion and, occasionally, post-stenotic dilatation and aneurysm formation (13-15).

It mainly affects the aortic arch, the thoracoabdominal aorta, its branches and the pulmonary arteries (16), which is why Takayasu's arteritis can be classified into 5 types according to the arteries involved (17).

- Type I: Classic type affecting only the branches of the arch.
- Type II:
  - Involvement of the aorta only in its ascending portion and the aortic arch.
  - Affectation of the descending thoracic aorta.
- Type III: Involvement of the thoracic and distal abdominal aorta.
- Type IV: Single involvement of the abdominal aorta and renal arteries.
- Type V: Generalized involvement of all aortic segments.
- Radiological findings in the early stages are thickening of the arterial wall with enhancement and in the late stages, calcium deposit in the walls, stenosis, post-stenotic dilatation, aneurysm formation, occlusion and evidence of increased collateral circulation (9, 18) (Figure 2).

2.2 Giant cell arteritis

It is the most frequent vasculitis of large and medium caliber vessels and affects people over 50 years of age (19). It occurs most commonly in northern European populations and a prevalence of 278 per 100,000 persons has been reported in the United States (13, 20).

Giant cell arteritis mainly affects the carotid arteries and the temporal artery, but can also occur in the aorta, its branches and central pulmonary arteries (16, 21). The most frequent signs and symptoms are temporary headache, jaw claudication and loss of vision. When it occurs in the aorta and its branches, it may present as general malaise, fever, night sweats, weight loss, arthralgias and decreased or absent peripheral pulses (3, 13).

The scenographic findings are wall thickening to the point of stenosis, post-stenotic dilatation, aneurysm formation, occlusion, calcification and mural thrombi (13, 22) (Figure 3).

2.3 Behçet's disease

It is a chronic multisystem vasculitis that can occur in vessels of any size, of the arterial and venous circulation, and occurs most frequently in people between 20 and 30 years of age (23, 24). This disease is found worldwide, but is most common in Eastern Mediterranean and East Asian countries, where prevalences of up to 30 cases per 100,000 people have been reported (25, 26).

It is characterized by ocular abnormalities (uveitis) and ulcers affecting the mouth and genitalia. Pulmonary involvement may occur in 10% of cases and involves the gastrointestinal tract in 10% to 50% of patients (23, 27).

The systemic manifestations of this vasculitis affect the arteries more than the veins. In arteries, it generates inflammation of the vasa vasorum of the tunica media, with destruction of elastic fibers and forms aneurysms (23, 25). The aorta is the most frequently affected artery and the pulmonary arteries occupy the second place; Behçet's disease is the most common cause of pulmonary artery aneurysm (28, 29).

In gastrointestinal manifestations, this vasculitis affects the small vessels of the intestinal wall, most frequently the venules (30). The main sites of involvement are the terminal ileum and cecum; the upper gastrointestinal tract, esophagus and stomach may also be affected, although this is rare (27, 31). Some complications may include perforation, hemorrhage and fistulas (9, 32).

The abdominal findings in scanography are concentric thickening of the intestinal wall with a marked enhancement with contrast medium. In the digestive tract, a large ovoid or irregular ulcer will be visualized with a marked thickening of the intestinal wall (9).

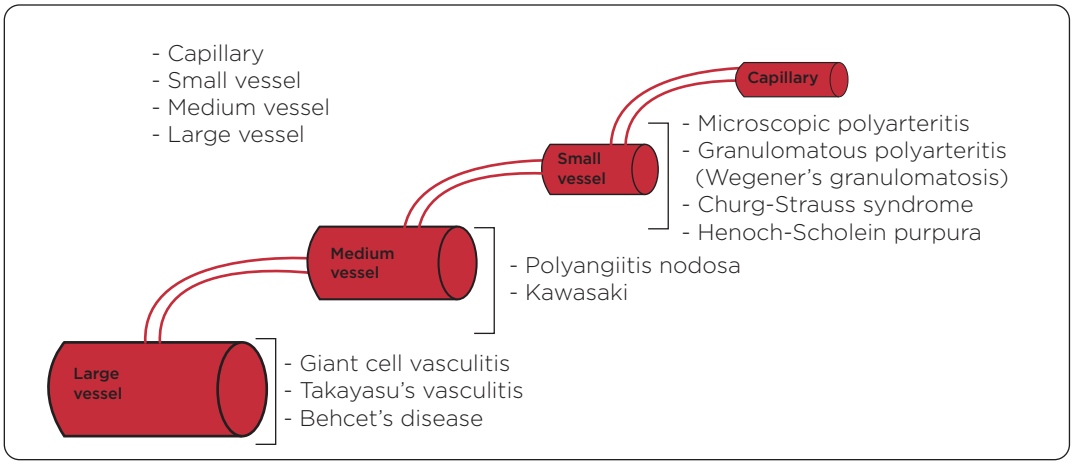

Figure 1. Classification of vasculitis according to Chapel Hill consensus, 2012.
Source: Own elaboration.

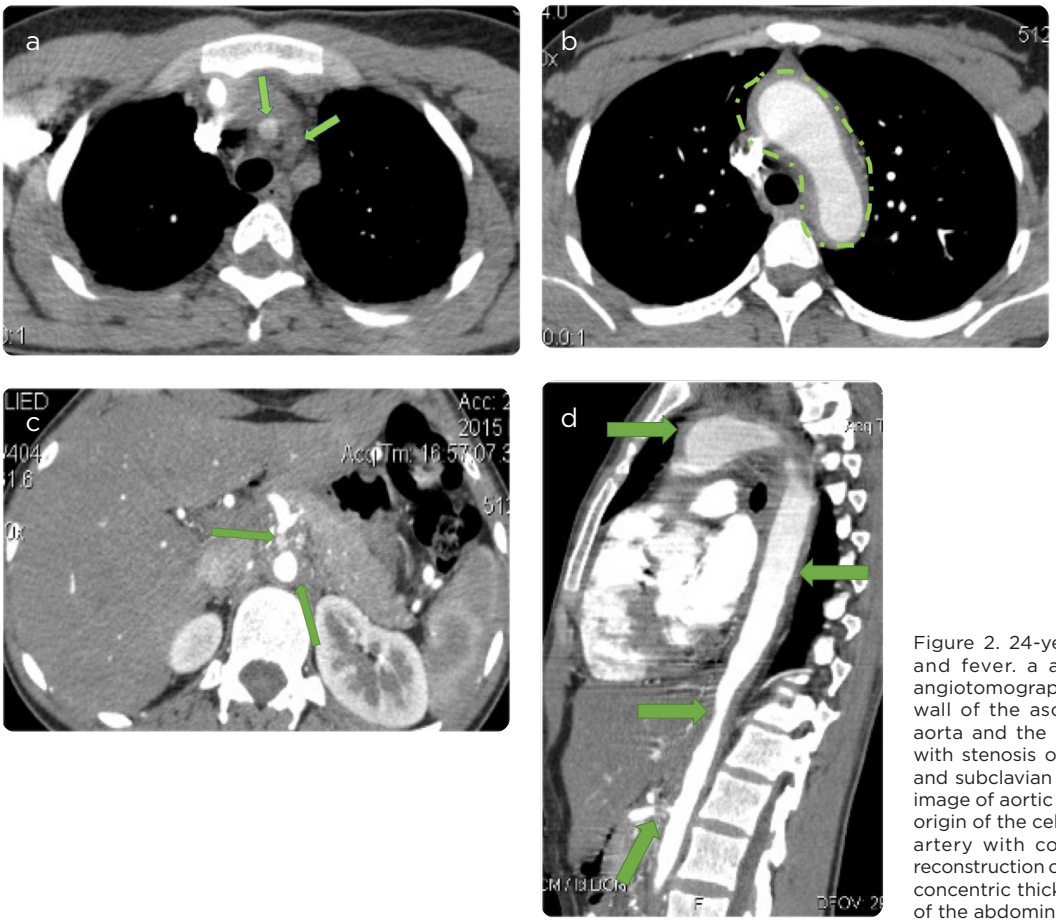

Figure 2. 24-year-old woman with weight loss and fever. a and b). Axial images of aortic angiotomography. Concentric thickening of the wall of the ascending aorta, arch, descending aorta and the origin of the supra-aortic trunks with stenosis of the left common carotid artery and subclavian artery on the same side. c). Axial image of aortic angiotomography. Stenosis of the origin of the celiac trunk and superior mesenteric artery with collateral circulation. d). Sagittal reconstruction of aortic angiotomography showing concentric thickening and the described stenosis of the abdominal vessels.

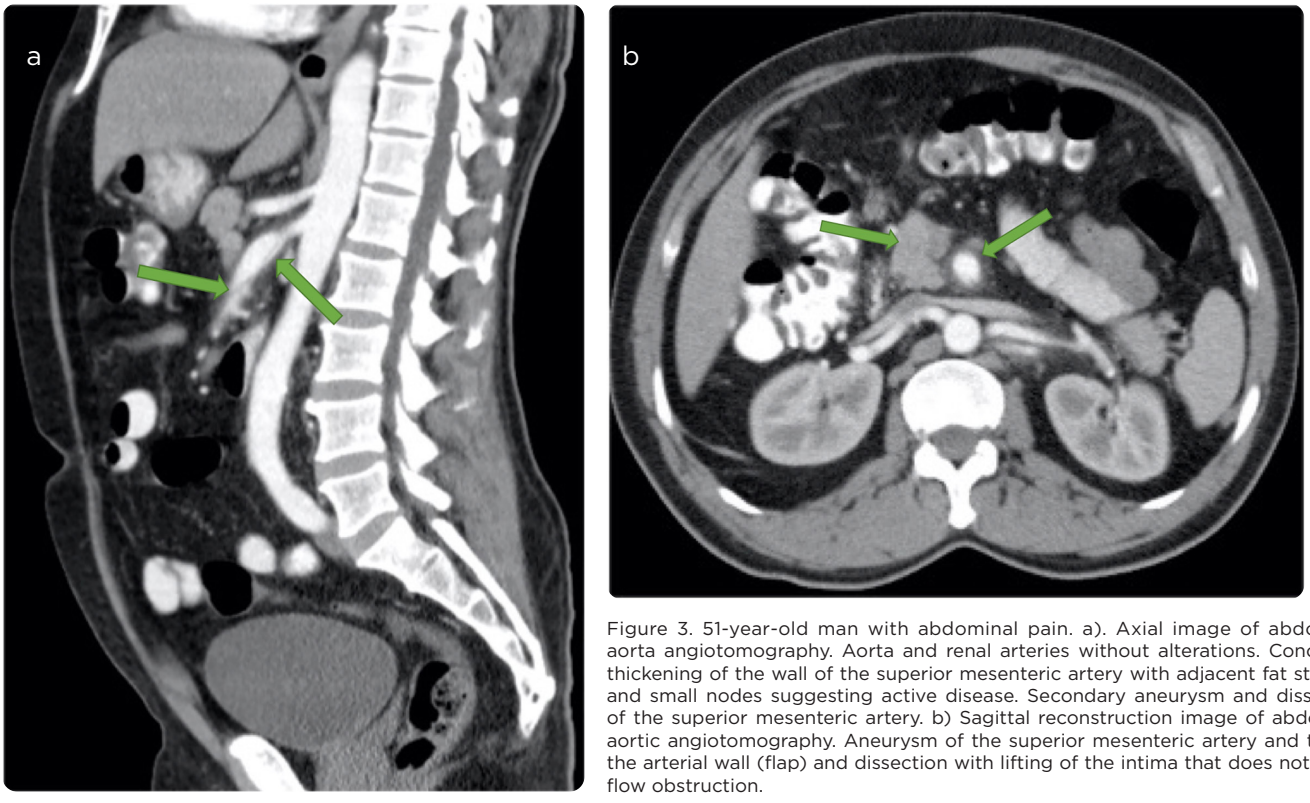

Figure 3. 51-year-old man with abdominal pain. a). Axial image of abdominal aorta angiotomography. Aorta and renal arteries without alterations. Concentric thickening of the wall of the superior mesenteric artery with adjacent fat striation and small nodes suggesting active disease. Secondary aneurysm and dissection of the superior mesenteric artery. b) Sagittal reconstruction image of abdominal aortic angiotomography. Aneurysm of the superior mesenteric artery and tear of the arterial wall (flap) and dissection with lifting of the intima that does not cause flow obstruction.

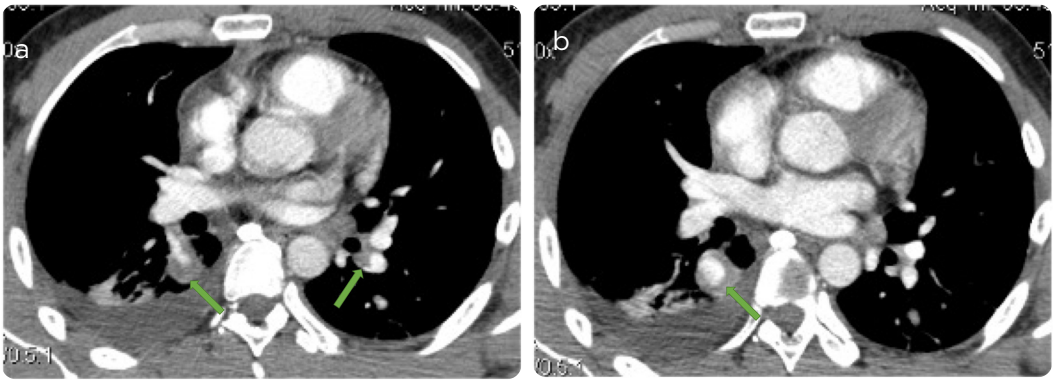

Figure 4. 29-year-old man with deep vein thrombosis (DVT) and chest pain, history of oral ulcers. a and b). Axial images of thorax tomography. Recanalized thrombosis of venous trunks, partially thrombosed focal aneurysm of segmental branch to the right lower lobe of the pulmonary artery.

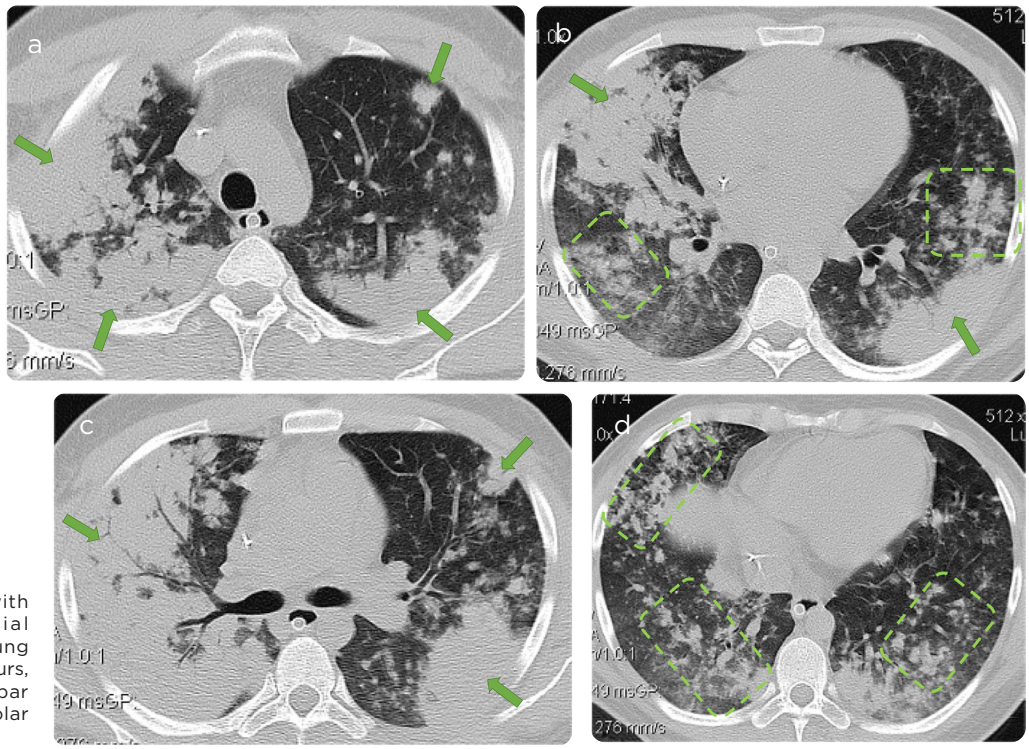

Figure 5. a-d) 19-year-old man with hemoptysis and dyspnea. Axial images of chest tomography in lung window. Nodules of irregular contours, ground-glass opacity and multilobar consolidation. BAL (bronchioalveolar lavage): alveolar hemorrhage.

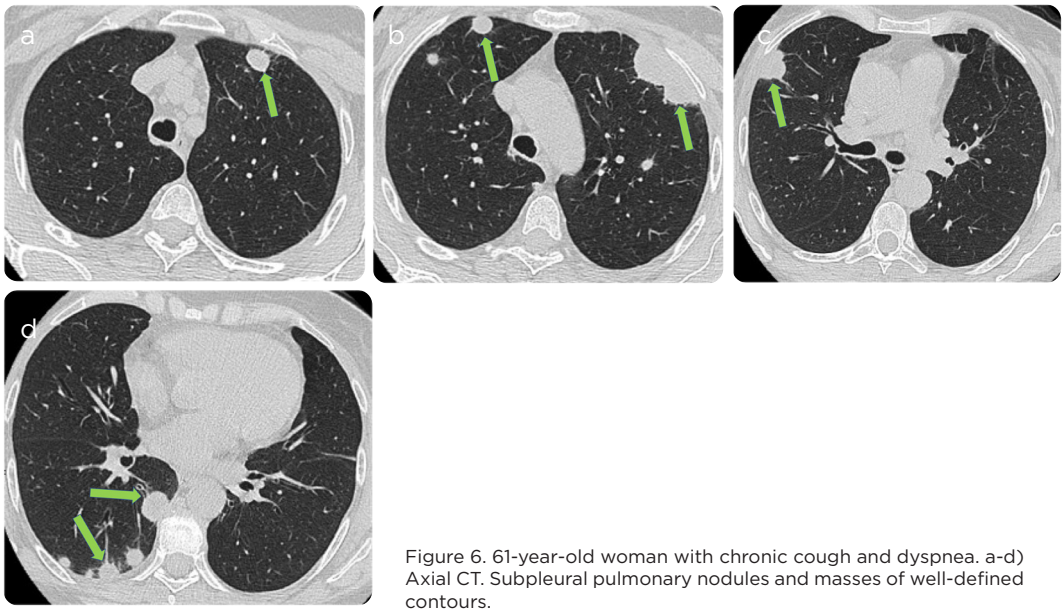

Figure 6. 61-year-old woman with chronic cough and dyspnea. a-d) Axial CT. Subpleural pulmonary nodules and masses of well-defined contours.

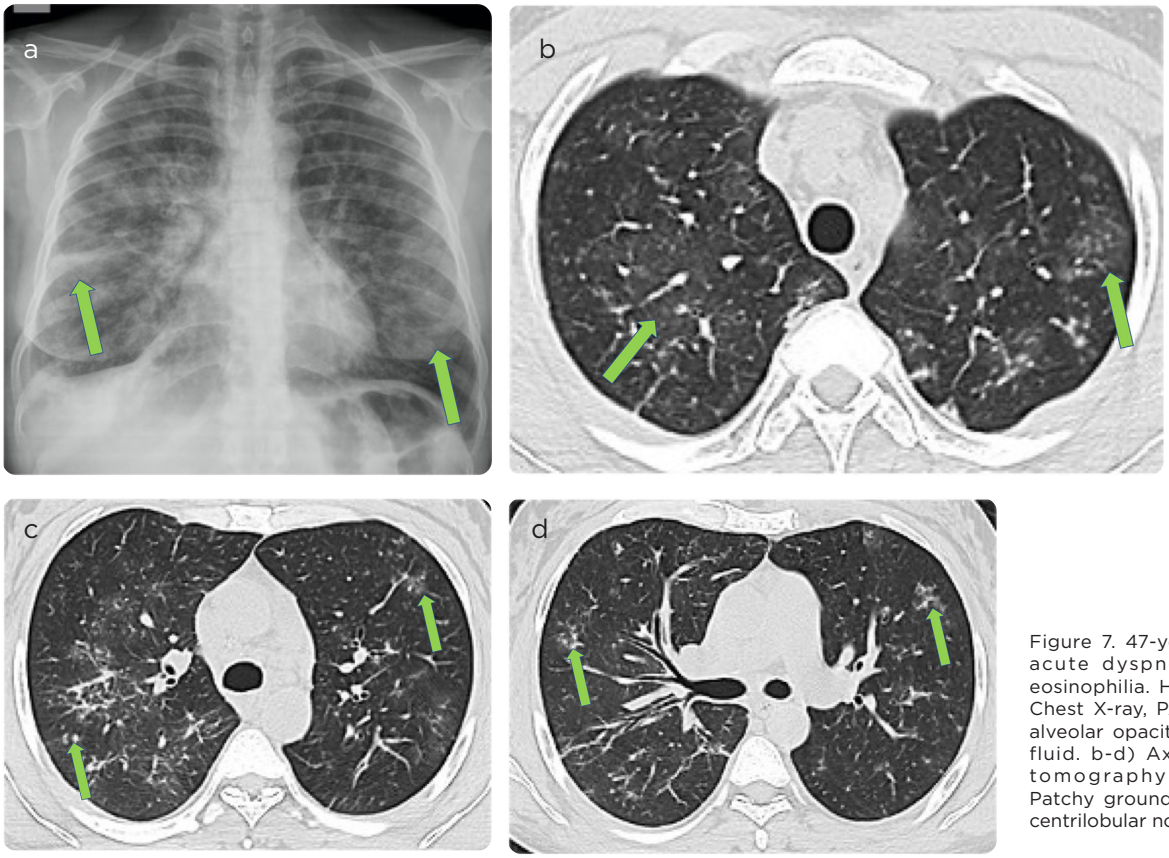

Figure 7. 47-year-old woman with acute dyspnea and peripheral eosinophilia. History of asthma. a) Chest X-ray, PA projection. Patchy alveolar opacities and right pleural fluid. b-d) Axial mages of chest tomography in lung window. Patchy ground-glass opacities and centrilobular nodules.

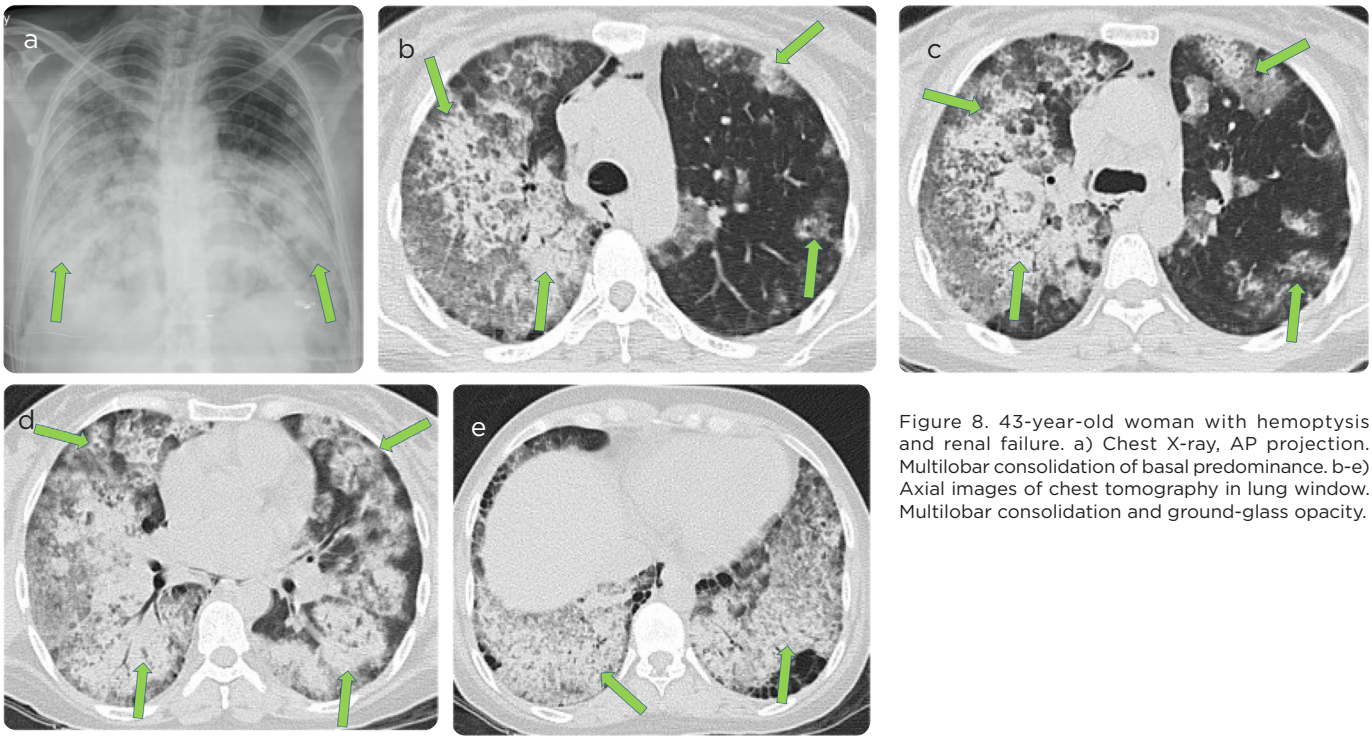

Figure 8. 43-year-old woman with hemoptysis and renal failure. a) Chest X-ray, AP projection. Multilobar consolidation of basal predominance. b-e) Axial images of chest tomography in lung window. Multilobar consolidation and ground-glass opacity.

Pulmonary findings are fusiform or multiple saccular pulmonary aneurysm, bilateral, and frequently located in the lower lobes and main pulmonary arteries. The aneurysms may be partially or totally thrombosed (33, 34) (Figure 4).

3. Small vessel vasculitis

3.1 Granulomatous polyangiitis (Wegener's granulomatosis)

It is a granulomatous vasculitis of unknown etiology and is associated with positive ANCA autoantibodies (35). Its prevalence is around 1.5 to 3 cases per 100,000 inhabitants and it is more frequent between 30 and 50 years of age (23).

It is clinically characterized by the triad of upper airway disease (nasal, oral or sinus inflammation), lower respiratory tract disease (airway or lung) and glomerulonephritis (13, 36). This triad may or may not be present from the onset.

Granulomatous polyangiitis can involve any part of the digestive tract. Ten percent of patients have gastrointestinal symptoms, such as abdominal pain (9).

Pulmonary signs and symptoms include hemoptysis, cough, chest pain and dyspnea; tracheobronchial signs and symptoms include stridor, dyspnea; the most common manifestations of upper airway involvement are rhinorrhea, epistaxis, sinusitis, otitis and sometimes destructive bone lesions (13, 37).

The scanographic findings can be pulmonary nodules and multiple masses, bilateral in subpleural regions and, less common, in the peribronchovascular region, with predilection for the lower lobes of the lung. Consolidation and "ground-glass" opacity. The nodules can be up to 10 cm in size and are usually cavitated, with thick walls and irregular margins or have the halo sign (edge of opacity in "ground glass" surrounding the pulmonary lesion) (16, 38). In the airway, thickening of the wall of the segmental and subsegmental bronchi will be visualized, as well as of the tracheal wall with narrowing of the airways (16) (figures 5 and 6).

3.2 Eosinophilic granulomatosis with polyangiitis (Churg-Strauss syndrome)

This is a necrotizing vasculitis characterized by a clinical triad (asthma, hypereosinophilia and necrotizing systemic vasculitis) (13,39). Four or more of the following six findings are required for diagnosis: asthma, > 10% eosinophilia in a differential white blood cell count, mononeuropathy or polyneuropathy due to systemic vasculitis, sinus abnormalities, migratory or transient pulmonary opacities, and histologic evidence of extravascular eosinophils in a biopsy specimen (23, 40, 41).

This disease is rare, with an incidence of one to three cases per 100,000 adults per year. It mainly affects men with a mean age of 50 years at the time of diagnosis (42). It has three phases: prodromal phase, which persists for many years and consists of asthma and previous allergic rhinitis; eosinophilic phase, characterized by marked eosinophilia in peripheral blood; and vasculitic phase (42, 43).

Extrapulmonary lesions are rare and are most commonly found in the gastrointestinal tract (20% of cases), spleen, heart and kidney. Gastrointestinal involvement typically manifests as obstructive symptoms, diarrhea, bleeding, ulceration and perforation (9).

Pulmonary findings on chest radiography are transient, bilateral areas of consolidation in any lung area. In scanography, areas of bilateral "ground-glass" opacity or randomly distributed consolidation and septal lines due to eosinophilic infiltration will be identified. When the airway is affected, centrilobular nodules and gemmation tree are observed. There may be bronchial dilatation and thickening of the bronchial wall (23, 44).

Abdominal findings are mesenteric vasculitis (thickening of the arterial wall) and infiltration of eosinophils in the intestinal wall (thickening of the wall that can produce obstruction) (9) (figure 7).

3.3 Microscopic polyangiitis

Microscopic polyangiitis is a non-granulomatous necrotizing systemic vasculitis affecting small vessels (45, 46). Its annual incidence is 0.36 cases per 100,000 (47) and affects middle-aged individuals. This pathology is the most frequent cause of lung-kidney syndrome, which is characterized by alveolar hemorrhage, alveolar capillaritis and necrotizing glomerulonephritis (23, 48).

Pulmonary symptoms include hemoptysis and respiratory distress; other relatively common manifestations include skin lesions, peripheral neuritis, and gastrointestinal hemorrhage (13, 49).

Radiologic pulmonary findings consistent with diffuse pulmonary hemorrhage are areas of "ground-glass" opacity, cobblestoning, or areas of consolidation (13, 50) (Figure 8).

4. Conclusions

Primary vasculitides are rare diseases and difficult to diagnose, because they can be confused with many other entities. For this reason it is important to know well their clinical and radiological manifestations, in order to suspect them and thus make an early diagnosis.

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