Current Pulmonology Reports (2020) 9:143–150
https://doi.org/10.1007/s13665-020-00263-x

PULMONARY RADIOLOGY (M STEPHENS AND S KAPUR, SECTION EDITORS)

Pulmonary Imaging Findings of Vasculitis

Matthew J. Stephens[1] · Ayaz Aghayev[2]

Accepted: 30 October 2020 / Published online: 27 November 2020

Abstract
Purpose of Review The purpose of this article is to review the various pulmonary imaging findings that may be present in patients with various types of vasculitis. The review will also go over the potential clinical significance of the various findings.
Recent Findings The majority of pulmonary findings related to vasculitis revolve around granulomatosis with polyangiitis (GPA) and eosinophilic granulomatosis with polyangiitis (EGPA). Additional etiologies are rare and mostly present as diffuse alveolar hemorrhage when small vessels are involved and aneurysms when medium and large vessels are involved.
Summary Pulmonary imaging findings related to vasculitis vary widely across etiologies. While many of these manifestations are rare, it is important to be familiar with them as they can cause severe complications.

Keywords Vasculitis · Pulmonary · Computed Tomography · Radiology · Imaging, ANCA-associated vasculitis · Antiglomerular basement membrane disease · Antiphospholipid syndrome · Behçet syndrome · Diffuse alveolar hemorrhage · Granulomatosis with polyangiitis (Wegener) · Microscopic polyangiitis · Takayasu arteritis

Introduction

Cellular inflammation of the vessel walls, also called vasculitis, is a multisystem disease with a variety of causes. It is often difficult to diagnose and manage given signs and symptoms often overlap with infection, malignancy, connective tissue disease, and medication reactions. There are several types of vasculitis that involve the vessels of the lungs. Depending on the vessels involved and the underlying cause of the vasculitis, this can give various presentations on imaging. Understanding these common patterns on imaging can help in the diagnosis and classification of the different types of vasculitis.

Vasculitis can be classified by the size of vessel involved and the underlying etiology [1]. When vasculitis is classified by size, it is usually categorized as small, medium, or large vessel vasculitis. The different-sized vasculotides are further categorized by etiology. In this review, we will go through each class of vasculitis and the pulmonary imaging manifestations that may present with each type.

This article is part of the Topical Collection on *Pulmonary Radiology*

✉ Matthew J. Stephens
mattjstephens@gmail.com

[1] Department of Radiology, University of Cincinnati School of Medicine, Cincinnati, OH, USA

[2] Cardiovascular Imaging Program, Department of Radiology, Brigham and Women's Hospital, Harvard Medical School, Boston, MA 02115, USA

Small Vessel

Small vessel vasculitis affects capillaries, venules, and arterioles. Most commonly, they affect patients 50–69 years old but can occur in patients of all ages. Small vessel vasculitis can be divided into four major categories including idiopathic, immune complex–mediated vasculitis, vasculitis from underlying systemic disease, and vasculitis from underlying etiology such as infection or drug reaction. Small vessel vasculitis is more commonly associated with pulmonary findings which can vary depending on the underlying etiology.

ANCA-Associated Vasculitis

Idiopathic causes of small vessel vasculitis are often associated with antineutrophil cytoplasmic antibodies (ANCAs). These antibodies are often associated with response to microbiocidal components used by neutrophils in host defense and cause significant proinflammatory effects with activation of neutrophils, monocytes, and endothelial cells. Idiopathic ANCA-associated small vessel vasculitis more frequently involves the lungs. The

most common types include granulomatosis with polyangiitis (GPA), eosinophilic granulomatosis with polyangiitis (EGPA) also known as Churg-Strauss Syndrome, and microscopic polyangiitis. GPA and EGPA have associated necrotizing granulomatous inflammation. Microscopic polyangiitis has necrotizing inflammation without granulomatosis.

Granulomatosis with Polyangiitis

GPA is the most common of ANCA-associated small vessel vasculitides. This type of vasculitis usually involves both the airways and the lung parenchyma. Renal involvement is common and can be present in up to 85% of cases [2].

On presentation, GPA is often acute causing sudden onset of shortness of breath. Classically GPA is associated with pulmonary nodules on imaging. Studies have shown that the most common finding on CT is multiple bilateral lung nodules [3] (Fig. 1). The presence of groundglass opacities around a nodule is common and represents associated alveolar hemorrhage. The nodules often cavitate with wall thickness thinning out as they resolve following treatment (Fig. 2). Cavitary lesions are associated with a higher rate of recurrence but no increased mortality [4].

Less commonly peribronchovascular consolidation is sometimes the dominant finding which can have a dramatic appearance (Fig. 3). Up to 8% of patients with GPA present with diffuse alveolar hemorrhage (DAH) [5]. DAH presents with multifocal groundglass opacities and consolidation usually without septal thickening (Fig. 4). Occasionally, GPA can evolve into another small vessel vasculitis such as microscopic polyangiitis (MPA) as a late manifestation of disease [6]. Bronchiectasis is an additional finding in patients with GPA and is usually associated with the presence of anti-MPO ANCA [7]. Occasionally, pneumothorax can occur and is associated with higher mortality when occurring in active disease [8].

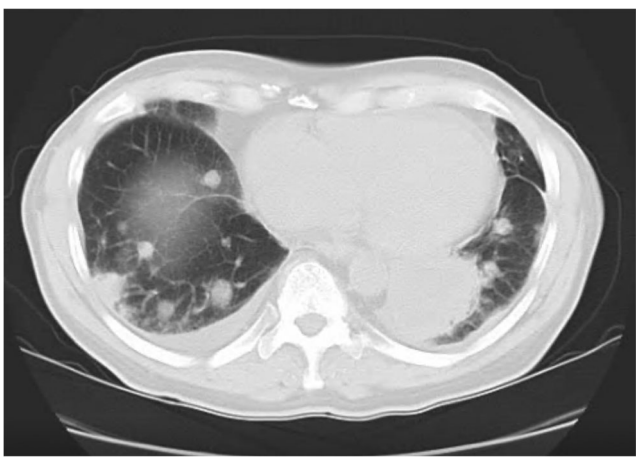

Fig. 1 Multiple bilateral pulmonary nodules in patient with GPA

Airway involvement often affects the trachea and lobar bronchi causing diffuse symmetrical thickening of the airway wall (Fig. 5). Patients also commonly demonstrate ENT involvement which can lead to septal perforation (Fig. 6). CT is usually the most common way to monitor the airways, but MRI is also a useful tool that can be used to monitor subglottic stenosis [9].

Eosinophilic Granulomatosis with Polyangiitis

EGPA also known as Churg-Strauss Syndrome (CSS) is associated with asthma and hypereosinophilia. Kidney involvement is less common occurring in approximately 25% of patients. It usually presents in three phases: the prodromal phase which is primarily asthma and allergic rhinitis, the second phase of marked peripheral blood eosinophilia, and finally the life-threatening vasculitic phase [10].

The most common imaging findings are migratory peripheral predominant groundglass opacities [11] which often overlap with chronic eosinophilic pneumonia (Fig. 6). This more common pulmonary appearance is more related to eosinophilic infiltrates and less to do with underlying vasculitis. Pulmonary hemorrhage related to vasculitis occurs less compared with GPA and MPA. In addition to groundglass opacities, EGPA may also demonstrate bronchial wall thickening, bronchiectasis, and uncommonly pleural effusions. Interlobular septal thickening can also sometimes be seen which is thought to represent eosinophilic infiltrate or edema from cardiac involvement. Lymphadenopathy is also occasionally demonstrated [12]. DAH is more common with MPO-ANCAs or ANCA-positive phenotypes occurring between 8 and 20% of ANCA-positive patients [13, 14]. Cardiac involvement is the leading cause of morbidity and mortality in patients with EGPA [15]. This often presents on cardiac MRI as patchy subendocardial late gadolidium enhancement with no definite vascular distribution (Figs. 7 and 8). Biopsy is usually only recommended if ANCA is negative as clinical and imaging findings are usually diagnostic when ANCA is positive.

Microscopic Polyangiitis

MPA is a nongranulomatous and noneosinophilic small vessel vasculitis. It is usually associated with MPO-ANCAs. MPA almost always has renal involvement. Pulmonary involvement presents as alveolar hemorrhage secondary to capillaritis. This presents as bilateral areas of ground-glass opacities and consolidation related to DAH. Pulmonary involvement is demonstrated in the minority of cases representing between 10 and 30% of total cases [16–20]. Pulmonary fibrosis is uncommon but can occur after the acute phase. It usually evolves from a mild reticular pattern to fibrosis with recurrent events. When present, fibrosis is usually associated with a higher mortality rate. Unlike GPA and EGPA, MPA does not have granulomatous inflammation. Onset of DAH in MPA is usually rapid and can be fatal if not promptly treated due to rapid

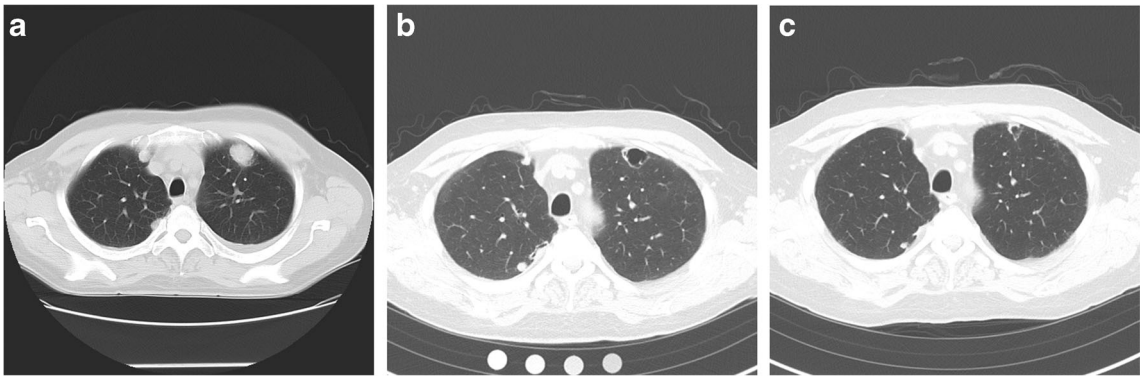

Fig. 2 Evolution of pulmonary nodule in patient with GPA. The initial nodule demonstrated in the left upper lobe is solid (**a**). The nodule later evolves into a cavitary nodule (**b**) and subsequently decreases in size and wall thickness following treatment (**c**)

progression and blood loss. Approximately 25% have more insidious onset with patients often severely anemic due to more intermittent DAH. ENT involvement is seen in 31%, and unlike GPA, there is no associated nasal perforation [21]. Progressive obstructive airway disease can occasionally be seen in nonsmokers with MPA [22, 23].

Immune Complex–Mediated Vasculitis

Immune complex–mediated vasculotides are usually not associated with ANCA. These include Goodpasture's Syndrome, Henoch-Schonlein purpura, and Behcet's disease. The predominant imaging finding involving the lung parenchyma is diffuse alveolar hemorrhage.

Goodpasture's Syndrome

Goodpasture's Syndrome (GP) is a nonvascular disorder which is caused by immunoglobulin G (IgG) deposition. This deposition can lead to pulmonary renal syndrome with DAH (Fig. 9). The deposition is usually along the glomerular basement membrane which initially causes a mild vasculitis in the kidney. This can then lead to a secondary antibody-mediated nephritis [24, 25]. Eventually, this can lead to an associated pulmonary capillartis and DAH [26–29]. Coexistence or successive development of Goodpasture syndrome and ANCA-associated vasculitis has been reported. A positive ANCA can be seen in up to one-third of patients with antiglomerular basement membrane disease with half of these patients demonstrating DAH [30].

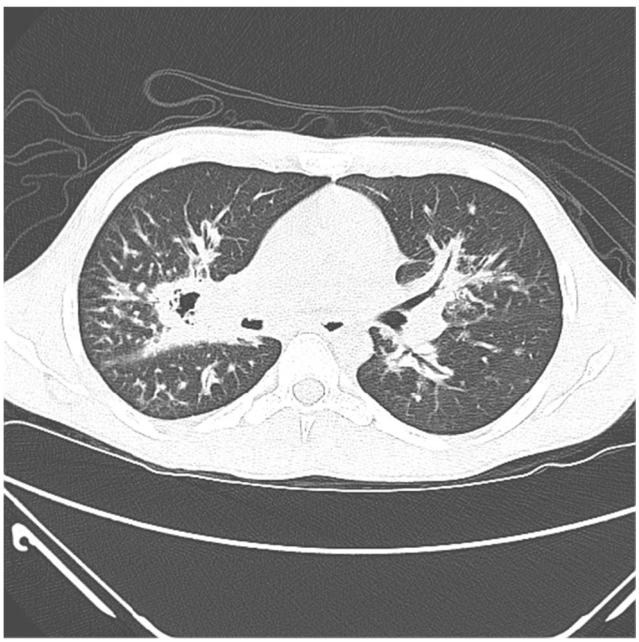

Fig. 3 Extensive peribronchial consolidation in patient with GPA. There is also an area of cavitation in the left perihilar region

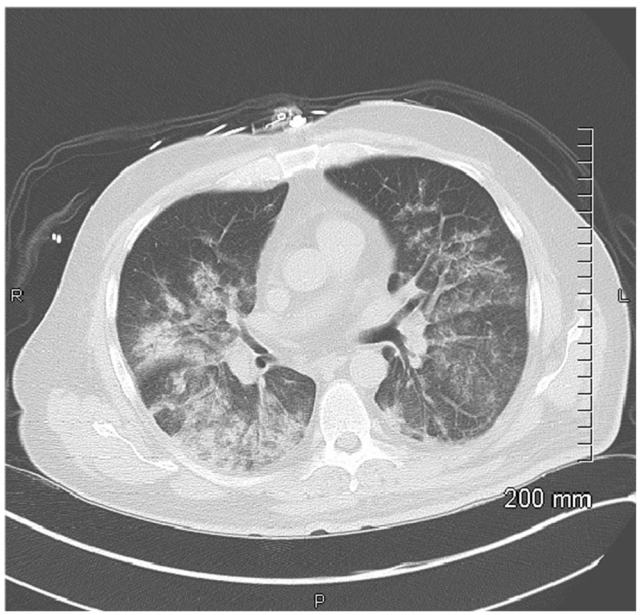

Fig. 4 Example of diffuse alveolar hemorrhage. Often this presents as patchy areas of consolidation and groundglass opacities

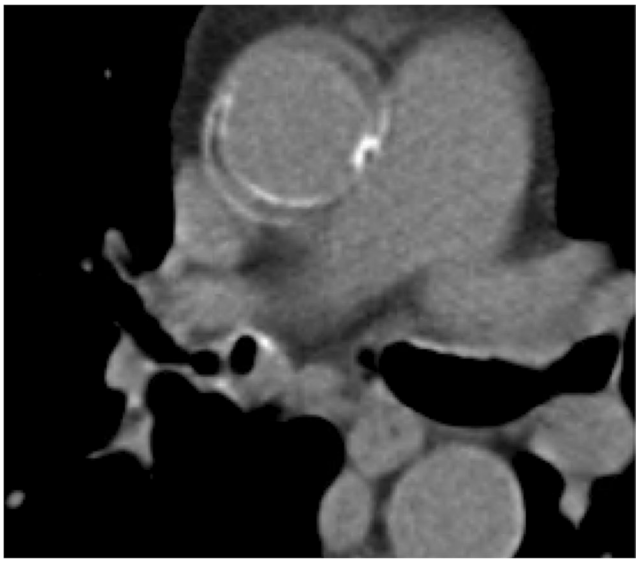

Fig. 5 Bronchial wall thickening of the right mainstem bronchus in patient with GPA

Henoch-Schonlein Purpura

Henoch-Schonlein Purpura (HSP) is a small vessel vasculitis due to immune complexes with immunoglobulin A (IgA). It most commonly is in children 4–7 years old but can occur in adults. Although uncommon, DAH has been reported in children [31, 32] and adults [33–36]. IgA lining alveolar septal vessels throughout hemorrhagic area has been demonstrated [35].

Behcet's Disease

Bechet's disease is a rare chronic multisystemic vasculitis usually in patients in their 20s or 30s. Pulmonary involvement can occur in up to 10% of cases [37]. Bechet's disease can involve small, medium, and large vessels with small vessel involvement triggering DAH appearance (Fig. 10). Pulmonary arteries are the second most common site of involvement with the aorta being the most common. Vasculitis causes dilation of the medium and large vessel lumen due to destruction of the elastic fibers. Associated aneurysms are usually multiple and bilateral with associated thrombosis. The most common pulmonary parenchymal abnormality are ill-defined areas of groundglass opacities usually from infarction, hemorrhage, or atelectasis [38, 39].

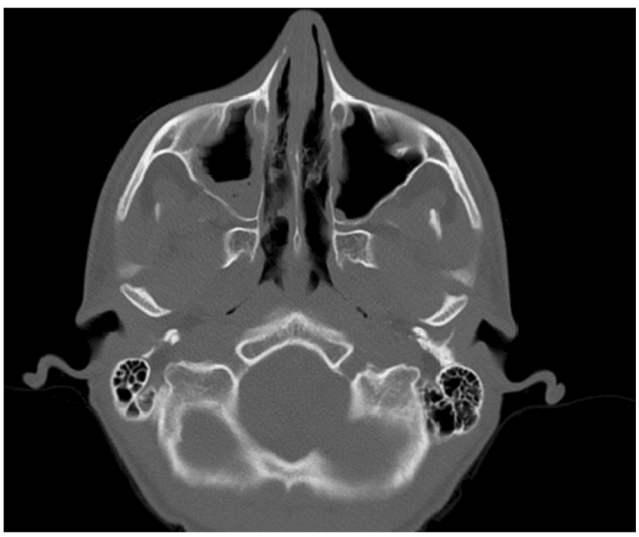

Fig. 6 Mucosal thickening in patient with GPA

Vasculitis Associated with Systemic Disease

Vasculitis can rarely be a manifestation of systemic autoimmune diseases usually presenting as DAH in the lungs. Systemic lupus erythematous (SLE) most commonly presents with pleural effusions but can also present with DAH as the first manifestation in up to 20% of patients [40]. This DAH is most commonly bland hemorrhage; however, vasculitis-induced DAH can present in up to 5% of patients making it the most common autoimmune disease with vasculitis-induced DAH [40, 41]. Prognosis of SLE with DAH has markedly improved in recent years due to improved treatment [42].

Scleroderma can rarely demonstrate DAH. The DAH may manifest as pulmonary-renal syndrome with small vessel vasculitis [43–45]. The underlying mechanism of DAH associated with scleroderma may be linked with p-ANCA [46] and more commonly MPO-ANCA [47, 48]. DAH has also been demonstrated in scleroderma patients with pulmonary pleuroparenchymal fibroelastosis (PPFE) although there is no clear link between these findings [49].

Additional systemic autoimmune disease has also shown DAH with capillaritis including mixed connective tissue disease [50], polymyositis [51], and rheumatoid arthritis [52].

Vasculitis Associated with Probable Etiology

Vasculitis can also occur due to infection, paraneoplastic syndromes, and medications. These are often caused with cryoglobulinemia or aberrant antibody production. Rarely, this can induce DAH in the lungs. For example, hepetatis C and cytomegalovirus can cause a virus-related cryoglobulinemias that can then trigger DAH [53, 54]. Lymphoprolipherative disease comprised usually of IgM monoclonal antibody can also cause DAH [55].

In drug-induced vasculitis, MPO titers are often elevated [56]. Hydralazine is the most common medication associated with drug-induced vasculitis followed by propylthiouracil (PTU). Additional medications most commonly associated with vasculitis include penicillamine, alopurinol, and sulfasalazine. Antibodies to elastase and lactoferrin are common with patients treated with hydralazine [57] while MPO-ANCAs with

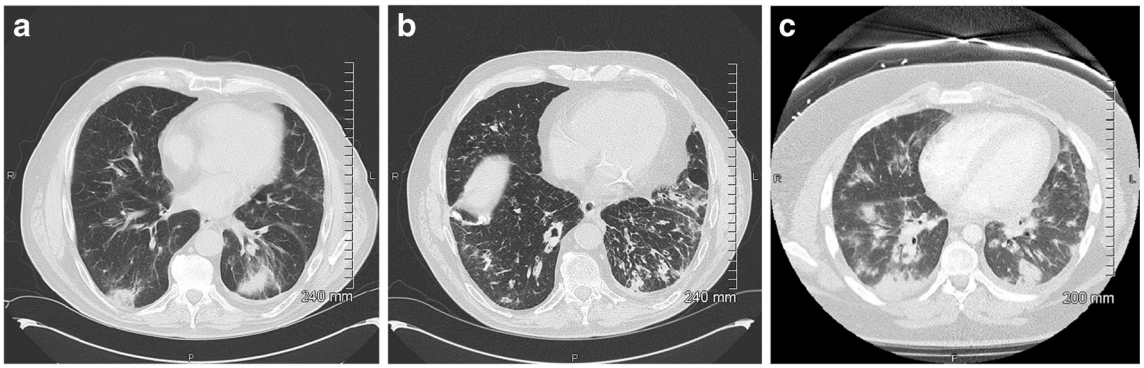

Fig. 7 Several examples of EGPA demonstrating peripheral predominant areas of consolidation and groundglass opacities (**a**, **c**) as well as extensive bronchial wall thickening (**b**)

anielastase and/or antilactoferin antibodies are characteristic of vasculitis associated with hydralazine or PTU [58]. Levimasol-induced vasculitis with DAH is demonstrated in Fig. 11.

Medium Vessel

Medium vessel vasculitis primarily affects the arteries and arterioles. Pulmonary involvement is uncommon. Aneurysm is the primary finding of vascular involvement and opposed to DAH seen predominantly with small vessel vasculotides.

Polyarteritis nodosa (PAN) is a medium vessel vasculitis most commonly affecting the renal arteries. Occasionally, this can present as a pulmonary artery aneurysm [59]. Pulmonary hemorrhage would be uncommon unless the aneurysm ruptured.

Kawasaki's disease is a medium vessel vasculitis primarily affecting children with the dominant pulmonary finding of aneurysms. Pulmonary involvement is rare [60] with only 1.8% of patients having predominantly pulmonary manifestations [61].

Large Vessel

Large vessel vasculitis primarily affects the aorta, pulmonary artery, and proximal branches. These will primarily present as aneurysms or vessel wall thickening of the involved vessels (Fig. 12). There are two large cell vasculitis that represent the

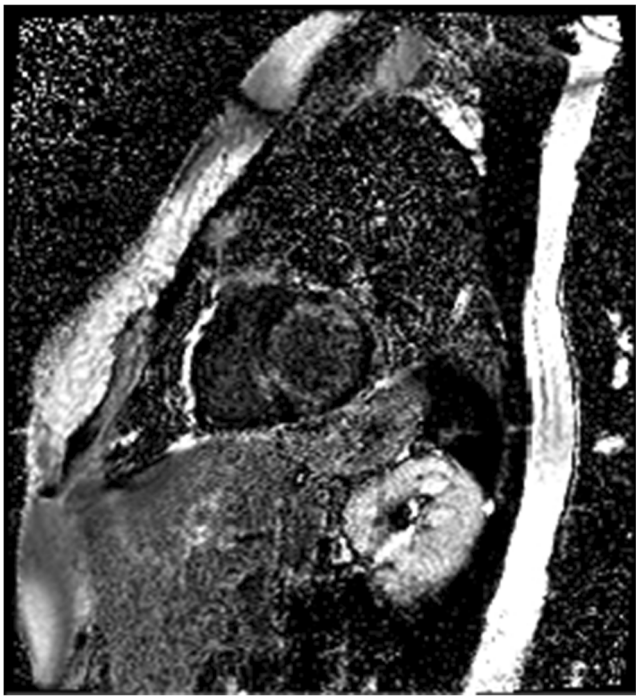

Fig. 8 Delayed contrast enhanced cardiac image of patient with EGPA demonstrating patchy subendocardial enhancement

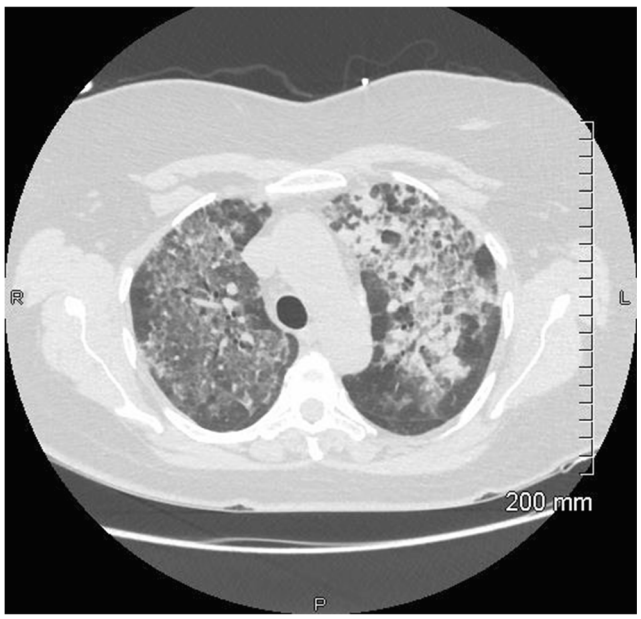

Fig. 9 Patient with Goopasture's Syndrome demonstrating diffuse alveolar hemorrhage secondary to vasculitis

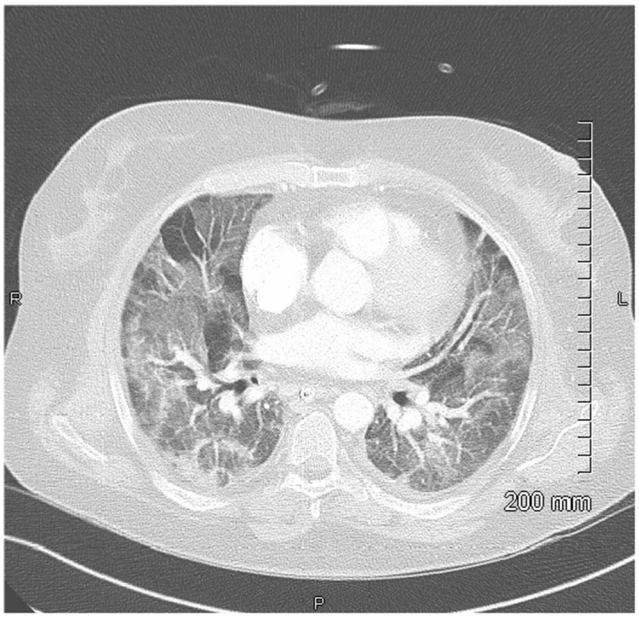

Fig. 10 Patient with Bechet's syndrome demonstrating multifocal areas of groundglass opacities that are consistent with DAH

vast majority of cases. These include Takayasu's arteritis and giant cell arteritis (GCA).

Takyasu's arteritis is one of the large vessel vasculotides primarily affecting the aorta and its branches. Pulmonary artery involvement is uncommon occurring in less than 10% of patients. When present, pulmonary artery stenosis or occlusion can occur. Lung parenchymal involvement can also be present primarily consisting of subpleural wedge-shaped opacities. Pulmonary hypertension has also been shown to

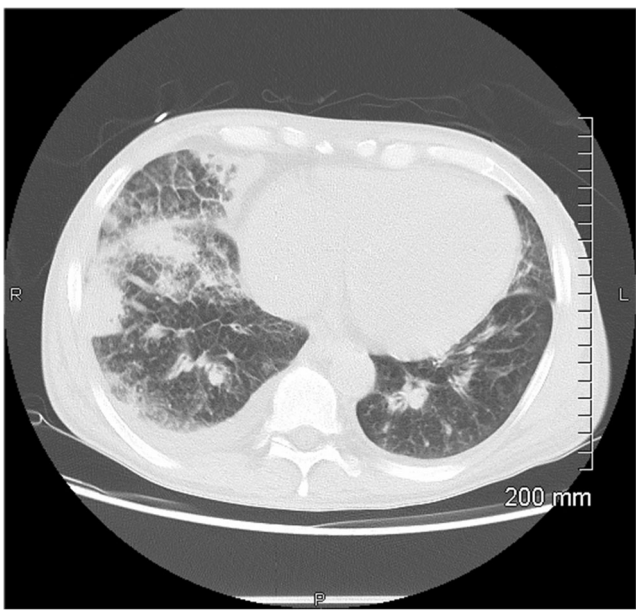

Fig. 11 Levimasol induced vascultitis causing multifocal areas of pulmonary hemorrhage and bronchial wall thickening

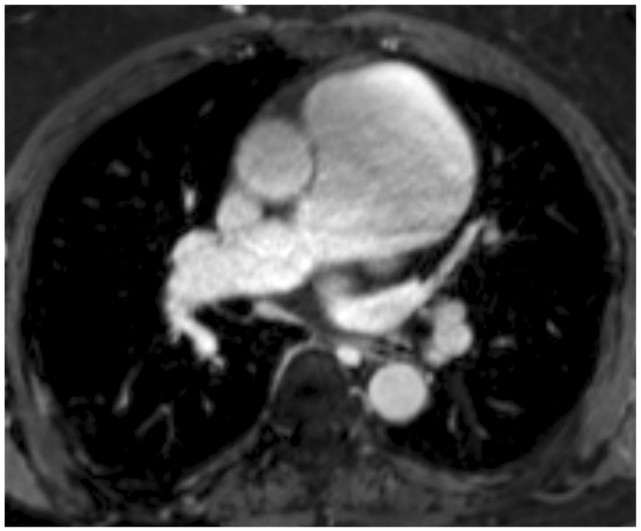

Fig. 12 Pulmonary artery aneurysm in patient with giant cell arteritis

associate with pulmonary involvement characterized by a longer disease duration, more symptoms such as fever, chest pain, and hemoptysis, and an increased erythrocyte sedimentation rate [62]. In another study, pulmonary artery involvement had increased incidence of pulmonary hypertension (62% vs. 8%) and had a higher mortality rate [63].

GCA can also rarely involve the lung parenchyma primarily presenting with pulmonary nodules or a diffuse interstitial pattern [64]. PPFE has also been shown to present in patients with GCA [65]. Additional examples of GCA-associated interstitial lung disease have also been reported [66].

Conclusions

Pulmonary imaging findings related to vasculitis vary widely in frequency and presentation depending on the underlying etiology. While small vessel vasculotides most commonly present with diffuse alveolar hemorrhage, medium and large vessel vasculotides present with pulmonary artery aneurysms. GPA and EGPA are idiopathic vasculotides that most commonly present with pulmonary findings where other small vessel vasculotides demonstrate pulmonary involvement in the minority of cases. Medium and large vessel vasculotides rarely demonstrate pulmonary imaging findings.

Compliance with Ethical Standards

Conflict of Interest The authors have no conflict of interest.

Human and Animal Rights and Informed Consent This article does not contain any studies with humans or animals performed by any of the authors.

References

Papers of particular interest, published recently, have been highlighted as:
- • Of importance
- •• Of major importance

1. 2012 revised International Chapel Hill Consensus Conference Nomenclature of Vasculitides. - PubMed - NCBI. https://www.ncbi.nlm.nih.gov/pubmed/23045170. Accessed Mar 30, 2020.
2. Frankel SK, Cosgrove GP, Fischer A, Meehan RT, Brown KK. Update in the diagnosis and management of pulmonary vasculitis. Chest. 2006;129(2):452–65.
3. Gómez-Gómez A, Martínez-Martínez MU, Cuevas-Orta E, Bernal-Blanco JM, Cervantes-Ramírez D, Martínez-Martínez R, et al. Pulmonary manifestations of granulomatosis with polyangiitis. Reumatol Clin. 2014;10(5):288–93.
4. Russell B, Mohan S, Chahal R, Carette S, Pagnoux C. Prognostic significance of cavitary lung nodules in granulomatosis with polyangiitis (Wegener's): a clinical imaging study of 225 patients. Arthritis Care Res. 2018;70(7):1082–9.
5. Cordier JF, Valeyre D, Guillevin L, Loire R, Brechot JM. Pulmonary Wegener's granulomatosis. A clinical and imaging study of 77 cases. Chest. 1990;97(4):906–12.
6. Bosch X. Microscopic polyangiitis (microscopic polyarteritis) with late emergence of generalised Wegener's granulomatosis. Ann Rheum Dis. 1999;58(10):644–7.
7. Néel A, Espitia-Thibault A, Arrigoni P-P, Volteau C, Rimbert M, Masseau A, et al. Bronchiectasis is highly prevalent in anti-MPO ANCA-associated vasculitis and is associated with a distinct disease presentation. Semin Arthritis Rheum. 2018;48(1):70–6.
8. Shi X, Zhang Y, Lu Y. Risk factors and treatment of pneumothorax secondary to granulomatosis with polyangiitis: a clinical analysis of 25 cases. J Cardiothorac Surg. 2018;13(1):7.
9. Henes FO, Laudien M, Linsenhoff L, Bremer JP, Oqueka T, Adam G, et al. Accuracy of magnetic resonance imaging for grading of subglottic stenosis in patients with granulomatosis with polyangiitis: correlation with pulmonary function tests and laryngoscopy. Arthritis Care Res. 2018;70(5):777–84.
10. Kim YK, Lee KS, Chung MP, Han J, Chong S, Chung MJ, et al. Pulmonary involvement in Churg-Strauss syndrome: an analysis of CT, clinical, and pathologic findings. Eur Radiol. 2007;17(12):3157–65.
11. Silva CIS, Müller NL, Fujimoto K, Johkoh T, Ajzen SA, Churg A. Churg-Strauss syndrome: high resolution CT and pathologic findings. J Thorac Imaging. 2005;20(2):74–80.
12. Nakamoto K, Saraya T, Ogawa Y, Ishii H, Takizawa H. Comparison of findings on thoracic computed tomography with the severity and duration of bronchial asthma in patients with eosinophilic granulomatosis with polyangiitis. Respir Med. 2018;139:101–5.
13. Sinico RA, Di Toma L, Maggiore U, et al. Prevalence and clinical significance of antineutrophil cytoplasmic antibodies in Churg-Strauss syndrome. Arthritis Rheum. 2005;52(9):2926–35.
14. 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;143(9):632–8.
15. Ríos Blanco JJ, Gómez Cerezo J, Suárez I, Gutiérrez M, Vázquez JJ, Barbado FJ. Churg-Strauss syndrome: outcome and long-term follow-up of 32 patients. Comment on the article by Solans et al. Rheumatol Oxf Engl. 2002;41(8):946–7 author reply 947.
16. Savage CO, Winearls CG, Evans DJ, Rees AJ, Lockwood CM. Microscopic polyarteritis: presentation, pathology and prognosis. Q J Med. 1985;56(220):467–83.
17. Guillevin L, Durand-Gasselin B, Cevallos R, et al. Microscopic polyangiitis: clinical and laboratory findings in eighty-five patients. Arthritis Rheum. 1999;42(3):421–30.
18. Haworth SJ, Savage CO, Carr D, Hughes JM, Rees AJ. Pulmonary haemorrhage complicating Wegener's granulomatosis and microscopic polyarteritis. Br Med J (Clin Res Ed). 1985;290(6484):1775–8.
19. Takahashi K, Hayashi S, Ushiyama O, Sueoka N, Fukuoka M, Nagasawa K. Development of microscopic polyangiitis in patients with chronic airway disease. Lung. 2005;183(4):273–81.
20. Adu D, Howie AJ, Scott DG, Bacon PA, McGonigle RJ, Micheal J. Polyarteritis and the kidney. Q J Med. 1987;62(239):221–37.
21. Lauque D, Cadranel J, Lazor R, Pourrat J, Ronco P, Guillevin L, et al. Microscopic polyangiitis with alveolar hemorrhage. A study of 29 cases and review of the literature. Groupe d'Etudes et de Recherche sur les Maladies "Orphelines" Pulmonaires (GERM"O"P). Medicine (Baltimore). 2000;79(4):222–33.
22. Schwarz MI, Mortenson RL, Colby TV, Waldron JA, Lynch DA, Hutt MP, et al. Pulmonary capillaritis. The association with progressive irreversible airflow limitation and hyperinflation. Am Rev Respir Dis. 1993;148(2):507–11.
23. Brugiere O, Raffy O, Sleiman C, Groussard O, Rothchild E, Mellot F, et al. Progressive obstructive lung disease associated with microscopic polyangiitis. Am J Respir Crit Care Med. 1997;155(2):739–42.
24. Verburgh CA, Bruijn JA, Daha MR, van Es LA. Sequential development of anti-GBM nephritis and ANCA-associated Pauci-immune glomerulonephritis. Am J Kidney Dis Off J Natl Kidney Found. 1999;34(2):344–8.
25. Beechler CR, Enquist RW, Hunt KK, Ward GW, Knieser MR. Immunofluorescence of transbronchial biopsies in Goodpasture's syndrome. Am Rev Respir Dis. 1980;121(5):869–72.
26. Case records of the Massachusetts General Hospital. Weekly clinicopathological exercises. Case 52–1993. A 17-year-old girl with massive hemoptysis and acute oliguric renal failure. N Engl J Med. 1993;329(27):2019–26.
27. García-Rostan y Pérez GM, García Bragado F, Puras Gil AM. Pulmonary hemorrhage and antiglomerular basement membrane antibody-mediated glomerulonephritis after exposure to smoked cocaine (crack): a case report and review of the literature. Pathol Int. 1997;47(10):692–7.
28. Lazor R, Bigay-Gamé L, Cottin V, Cadranel J, Decaux O, Fellrath JM, et al. Alveolar hemorrhage in anti-basement membrane antibody disease: a series of 28 cases. Medicine (Baltimore). 2007;86(3):181–93.
29. Lombard CM, Colby TV, Elliott CG. Surgical pathology of the lung in anti-basement membrane antibody-associated Goodpasture's syndrome. Hum Pathol. 1989;20(5):445–51.
30. Niles JL, Böttinger EP, Saurina GR, Kelly KJ, Pan G, Collins AB, et al. The syndrome of lung hemorrhage and nephritis is usually an ANCA-associated condition. Arch Intern Med. 1996;156(4):440–5.
31. Olson JC, Kelly KJ, Pan CG, Wortmann DW. Pulmonary disease with hemorrhage in Henoch-Schöenlein purpura. Pediatrics. 1992;89(6 Pt 2):1177–81.
32. Vats KR, Vats A, Kim Y, Dassenko D, Sinaiko AR. Henoch-Schönlein purpura and pulmonary hemorrhage: a report and literature review. Pediatr Nephrol Berl Ger. 1999;13(6):530–4.
33. Markus HS, Clark JV. Pulmonary haemorrhage in Henoch-Schönlein purpura. Thorax. 1989;44(6):525–6.
34. Shichiri M, Tsutsumi K, Yamamoto I, Ida T, Iwamoto H. Diffuse intrapulmonary hemorrhage and renal failure in adult Henoch-Schönlein purpura. Am J Nephrol. 1987;7(2):140–2.
35. Kathuria S, Cheifec G. Fatal pulmonary Henöch-Schonlein syndrome. Chest. 1982;82(5):654–6.
36. Yokose T, Aida J, Ito Y, Ogura M, Nakagawa S, Nagai T. A case of pulmonary hemorrhage in Henoch-Schönlein purpura accompanied

by polyarteritis nodosa in an elderly man. Respir Int Rev Thorac Dis. 1993;60(5):307–10.
37. Erkan F, Gül A, Tasali E. Pulmonary manifestations of Behçet's disease. Thorax. 2001;56(7):572–8.
38. Hiller N, Lieberman S, Chajek-Shaul T, Bar-Ziv J, Shaham D. Thoracic manifestations of Behçet disease at CT. Radiogr Rev Publ Radiol Soc N Am Inc. 2004;24(3):801–8.
39. Tunaci M, Ozkorkmaz B, Tunaci A, Gül A, Engin G, Acunaş B. CT findings of pulmonary artery aneurysms during treatment for Behçet's disease. AJR Am J Roentgenol. 1999;172(3):729–33.
40. Zamora MR, Warner ML, Tuder R, Schwarz MI. Diffuse alveolar hemorrhage and systemic lupus erythematosus. Clinical presentation, histology, survival, and outcome. Medicine (Baltimore). 1997;76(3):192–202.
41. Barile LA, Jara LJ, Medina-Rodriguez F, García-Figueroa JL, Miranda-Limón JM. Pulmonary hemorrhage in systemic lupus erythematosus. Lupus. 1997;6(5):445–8.
42. Santos-Ocampo AS, Mandell BF, Fessler BJ. Alveolar hemorrhage in systemic lupus erythematosus: presentation and management. Chest. 2000;118(4):1083–90.
43. Naniwa T, Banno S, Sugiura Y, Yokota K, Oosawa T, Maeda S, et al. Pulmonary-renal syndrome in systemic sclerosis: a report of three cases and review of the literature. Mod Rheumatol. 2007;17(1):37–44.
44. Griffin MT, Robb JD, Martin JR. Diffuse alveolar haemorrhage associated with progressive systemic sclerosis. Thorax. 1990;45(11):903–4.
45. Bar J, Ehrenfeld M, Rozenman J, Perelman M, Sidi Y, Gur H. Pulmonary-renal syndrome in systemic sclerosis. Semin Arthritis Rheum. 2001;30(6):403–10.
46. Vázquez-Del Mercado M, Mendoza-Topete A, Best-Aguilera CR, Garcia-De La Torre I. Diffuse alveolar hemorrhage in limited cutaneous systemic sclerosis with positive perinuclear antineutrophil cytoplasmic antibodies. J Rheumatol. 1996;23(10):1821–3.
47. Endo H, Hosono T, Kondo H. Antineutrophil cytoplasmic autoantibodies in 6 patients with renal failure and systemic sclerosis. J Rheumatol. 1994;21(5):864–70.
48. Casari S, Haeney M, Farrand S, Herrick A. Antineutrophil cytoplasmic antibodies a "red flag" in patients with systemic sclerosis. J Rheumatol. 2002;29(12):2666–7.
49. Wutzl AL, Foley RN, O'Driscoll BR, Reeve RS, Chisholm R, Herrick AL. Microscopic polyangiitis presenting as pulmonary-renal syndrome in a patient with long-standing diffuse cutaneous systemic sclerosis and antibodies to myeloperoxidase. Arthritis Rheum. 2001;45(6):533–6.
50. Schwarz MI, Zamora MR, Hodges TN, Chan ED, Bowler RP, Tuder RM. Isolated pulmonary capillaritis and diffuse alveolar hemorrhage in rheumatoid arthritis and mixed connective tissue disease. Chest. 1998;113(6):1609–15.
51. Schwarz MI, Sutarik JM, Nick JA, Leff JA, Emlen JW, Tuder RM. Pulmonary capillaritis and diffuse alveolar hemorrhage. A primary manifestation of polymyositis. Am J Respir Crit Care Med. 1995;151(6):2037–40.
52. Genta MS, Genta RM, Gabay C. Systemic rheumatoid vasculitis: a review. Semin Arthritis Rheum. 2006;36(2):88–98.
53. Gómez-Tello V, Oñoro-Cañaveral JJ, de la Casa Monje RM, et al. Diffuse recidivant alveolar hemorrhage in a patient with hepatitis C virus-related mixed cryoglobulinemia. Intensive Care Med. 1999;25(3):319–22.
54. Kramer J, Hennig H, Lensing C, Krüger S, Helmchen U, Steinhoff J, et al. Multi-organ affecting CMV-associated cryoglobulinemic vasculitis. Clin Nephrol. 2006;66(4):284–90.
55. Silva F, Pinto C, Barbosa A, Borges T, Dias C, Almeida J. New insights in cryoglobulinemic vasculitis. J Autoimmun. 2019;105:102313.
56. Choi HK, Merkel PA, Walker AM, Niles JL. Drug-associated antineutrophil cytoplasmic antibody-positive vasculitis: prevalence among patients with high titers of antimyeloperoxidase antibodies. Arthritis Rheum. 2000;43(2):405–13.
57. Kudoh Y, Kuroda S, Shimamoto K, Iimura O. Propylthiouracil-induced rapidly progressive glomerulonephritis associated with antineutrophil cytoplasmic autoantibodies. Clin Nephrol. 1997;48(1):41–3.
58. Short AK, Lockwood CM. Antigen specificity in hydralazine associated ANCA positive systemic vasculitis. QJM Mon J Assoc Phys. 1995;88(11):775–83.
59. Lee YJ, Park SS, Kim SY, Lee JY, Koo HK, Yoon HI. A case of systemic polyarteritis nodosa involving bronchial artery. Sarcoidosis Vasc Diffuse Lung Dis Off J WASOG. 2010;27(2):164–8.
60. Escalon JG, Wu X, Drexler IR, Lief L, Plataki M, Bender M, et al. Rare case of pulmonary involvement in an adult with Kawasaki disease. Clin Imaging. 2018;47:1–3.
61. Singh S, Gupta A, Jindal AK, Gupta A, Suri D, Rawat A, et al. Pulmonary presentation of Kawasaki disease—a diagnostic challenge. Pediatr Pulmonol. 2018;53(1):103–7.
62. Yang J, Peng M, Shi J, Zheng W, Yu X. Pulmonary artery involvement in Takayasu's arteritis: diagnosis before pulmonary hypertension. BMC Pulm Med. 2019;19(1):225.
63. He Y, Lv N, Dang A, Cheng N. Pulmonary artery involvement in patients with Takayasu arteritis. J Rheumatol. 2020;47(2):264–72.
64. Kramer MR, Melzer E, Nesher G, Sonnenblick M. Pulmonary manifestations of temporal arteritis. Eur J Respir Dis. 1987;71(5):430–3.
65. Perruzza M, Fusha E, Cameli P, Capecchi PL, Selvi E, Gentili F, et al. Pleuroparenchymal fibroelastosis (PPFE) associated with giant cell arteritis: a coincidence or a novel phenotype? Respir Med Case Rep. 2019;27:100843.
66. Konishi C, Nakagawa K, Nakai E, et al. Interstitial lung disease as an initial manifestation of giant cell arteritis. Intern Med. 2017;56(19):2633–2637. https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5658531/. Accessed Mar 30, 2020.