

26. Bölüm

KİSTİK AKCİĞER HASTALIKLARINA YAKLAŞIM

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Kistik akciğer hastalıkları, klinik uygulamalarda az sıklıkta karşılaşılan ve çoğunlukla radyolojik bulguların benzer özellikte olması nedeniyle tanı konulması zor hastalıklar grubudur. Kistler genellikle akciğerlerin tomografik taramaları sonucu saptanmaktadır. Multidisipliner bir yaklaşımla birleştirilen klinik ve radyografik özellikler doğru tanı konulmasına olanak sağlayarak, oldukça geniş spektrumda yer alan bu hastalıklar arasında ayırım yapmaya yardımcı olabilmektedir.

Kist; epiteliyal veya fibröz bir dış duvar ile çevrili yuvarlak hava boşluğu olarak tanımlanır. Radyolojik görünüm olarak, ince (<2mm) fakat net olarak fark edilebilen bir duvara sahip parankim içindeki hava dansitesindeki veya azalmış atenüasyon alanlarıdır ¹.

Akciğer kistleri ile ortaya çıkan hastalık grubu geniş bir yelpazede yer alır ve kistik akciğer hastalıkları, ayırıcı tanının karmaşık olabileceği heterojen bir patoloji grubudur. Kistik akciğer hastalıklarının radyolojik değerlendirmesi için, tanı sürecinin ilk adımında gerçek akciğer kistlerini diğer hava dolu akciğer lezyonlarından ayırmak önemlidir. Akciğer kistlerinin boyut, duvar kalınlığı, sayısı, konumu ve dağılımı dahil radyolojik özellikleri ve ilgili radyolojik bulgular, spesifik kistik akciğer hastalıklarının teşhisi için en yararlı tanısal ipuçlarını sağlar. Kesin tanı, klinik korelasyon ve bazen de biyopsi gerektirebilir. Bununla birlikte, doğru

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dahil olmak üzere ilişkili radyolojik bulgular dikkate alınarak sağlanabilir. Radyografik bulgulardaki benzerlikler göz önüne alındığında, bazı DKAH vakalarında doku veya genetik analiz yoluyla doğrulama testleri de gerekli olabilmektedir.

KAYNAKLAR

1. Ryu JH, Swensen SJ. Cystic and cavitory lung diseases: focal and diffuse. *Mayo Clin Proc.* 2003;78(6):744-752.
2. Kikuchi E, Kinoshita I, Yamazaki K, et al. Epithelioid sarcoma presenting as pulmonary cysts with cancer antigen 125 expression. *Respirology* 2006;11(6):826- 829.
3. Ohdama S, Akagawa S, Matsubara O, et al. Primary diffuse alveolar septal amyloidosis with multiple cysts and calcification. *Eur Respir J* 1996;9(7):1569-1571.
4. Colombat M, Caudroy S, Lagonotte E, et al. Pathomechanisms of cyst formation in pulmonary light chain deposition disease. *Eur Respir J* 2008;32(5):1399-1403.
5. Copley SJ, Wells AU, Hawtin KE, et al. Lung morphology in the elderly: comparative CT study of subjects over 75 years old versus those under 55 years old. *Radiology* 2009; 251:566-573.
6. Araki T, Nishino M, Gao W, et al. Pulmonary cysts identified on chest CT: are they part of aging change or of clinical significance? *Thorax* 2015; 70:1156-1162.
7. Raof S, Bondalapati P, Vydyula R, et al. Cystic lung diseases: algorithmic approach. *Chest* 2016; 150:945-965.
8. Biyyam DR, Chapman T, Ferguson MR, et al. Congenital lung abnormalities: embryologic features, prenatal diagnosis, and postnatal radiologic-pathologic correlation. *Radiographics* 2010; 30:1721-1738.
9. Gupta N, Vassallo R, Wikenheiser-Brokamp KA, et al. Diffuse cystic lung disease. Part II. *Am J Respir Crit Care Med* 2015;192:17-29.
10. Odev K, Guler I, Altinok T, et al. Cystic and cavitory lung lesions in children: radiologic findings with pathologic correlation. *J Clin Imaging Sci* 2013; 3:60.
11. Yoon YC, Lee KS, Kim TS, et al. Intrapulmonary bronchogenic cyst: CT and pathologic findings in five adult patients. *AJR Am J Roentgenol* 2002; 179:167- 170.
12. Meraj R, Wikenheiser-Brokamp KA, Young LR, et al. Lymphangioliomyomatosis: new concepts in pathogenesis, diagnosis, and treatment. *Semin Respir Crit Care Med* 2012; 33:486-497.
13. Samuelli S, Abraham K, Dressler A, et al. Tuberos sclerosi complex: new criteria for diagnostic work-up and management. *Wien Klin Wochenschr* 2015; 127:619-630.
14. Ryu JH, Moss J, Beck GJ, et al. The NHLBI lymphangioliomyomatosis registry: characteristics of 230 patients at enrollment. *Am J Respir Crit Care Med.* 2006;173(1):105. Epub 2005 Oct 6.
15. Vassallo R, Ryu JH, Schroeder DR, et al. Clinical outcomes of pulmonary Langerhans' cell histiocytosis in adults. *N Engl J Med.* 2002;346(7):484.
16. Schmidt LS, Linehan WM. Molecular genetics and clinical features of Birt-Hogg-Dubésyndrome. *Nat Rev Urol.* 2015 Oct;12(10):558-69. Epub 2015 Sep 01.
17. Marciniak SJ, Johnson SR. Pneumothorax and the biology of Birt-Hogg-Dubésyndrome. *Thorax.* 2020;75(6):442.
18. Chu L, Luo Y, Chen H, et al. Mesenchymal folliculin is required for alveolar development: implications for cystic lung disease in Birt-Hogg-Dubésyndrome. *Thorax.* 2020;75(6):486. Epub 2020 Apr 1.
19. Toro JR, Pautler SE, Stewart L, et al. Lung cysts, spontaneous pneumothorax, and genetic associations in 89 families with Birt-Hogg-Dubésyndrome. *Am J Respir Crit Care Med.* 2007;175(10):1044. Epub 2007 Feb 22.

20. Toro JR, Wei MH, Glenn GM, et al. BHD mutations, clinical and molecular genetic investigations of Birt-Hogg-Dubésyndrome: a new series of 50 families and a review of published reports. *J Med Genet.* 2008 Jun;45(6):321-31. Epub 2008 Jan 30.
21. Johnson SR, Tattersfield AE. Clinical experience of lymphangioleiomyomatosis in the UK. *Thorax.* 2000;55(12):1052.
22. Cha SI, Fessler MB, Cool CD, et al. Lymphoid interstitial pneumonia: clinical features, associations and prognosis. *Eur Respir J.* 2006;28(2):364-369. Epub 2006 Mar 29.
23. Mendez JL, Nadrous HF, Vassallo R, et al. Pneumothorax in pulmonary Langerhans cell histiocytosis. *Chest.* 2004;125(3):1028.
24. Strimlan CV, Rosenow EC 3rd, Weiland LH, et al. Lymphocytic interstitial pneumonitis. Review of 13 cases. *Ann Intern Med.* 1978;88(5):616.
25. Gupta N, Meraj R, Tanase D, et al. Accuracy of chest high-resolution computed tomography in diagnosing diffuse cystic lung diseases. *Eur Respir J.* 2015 Oct;46(4):1196-9. Epub 2015 Jul 9.
26. Tobino K, Gunji Y, Kurihara M, et al. Characteristics of pulmonary cysts in Birt-Hogg-Dubésyndrome: thin-section CT findings of the chest in 12 patients. *Eur J Radiol.* 2011;77(3):403.
27. Abbott GF, Rosado-de-Christenson ML, Franks TJ. From the archives of the AFIP: pulmonary Langerhans cell histiocytosis. *Radiographics.* 2004;24(3):821.
28. Escalon JG, Richards JC, Koelsch T, et al. Isolated Cystic Lung Disease: An Algorithmic Approach to Distinguishing Birt-Hogg-Dubésyndrome, Lymphangioleiomyomatosis, and Lymphocytic Interstitial Pneumonia. *AJR Am J Roentgenol.* 2019:1.
29. Johkoh T, Müller NL, Pickford HA, et al. Lymphocytic interstitial pneumonia: thin-section CT findings in 22 patients. *Radiology.* 1999;212(2):567.
30. Silva CI, Flint JD, Levy RD, et al. Diffuse lung cysts in lymphoid interstitial pneumonia: high-resolution CT and pathologic findings. *J Thorac Imaging.* 2006;21(3):241.
31. Yousem SA, Colby TV, Chen YY, et al. Pulmonary Langerhans' cell histiocytosis: molecular analysis of clonality. *Am J Surg Pathol.* 2001;25(5):630.
32. Brauner MW, Grenier P, Mouelhi MM, et al. Pulmonary histiocytosis X: evaluation with high-resolution CT. *Radiology.* 1989;172(1):255.
33. Kim HJ, Lee KS, Johkoh T, et al. Pulmonary Langerhans cell histiocytosis in adults: high-resolution CT-pathology comparisons and evolutionary changes at CT. *Eur Radiol.* 2011 Jul;21(7):1406-15. Epub 2011 Feb 11.
34. Johkoh T, Ichikado K, Akira M, et al. Lymphocytic interstitial pneumonia: follow-up CT findings in 14 patients. *J Thorac Imaging.* 2000;15(3):162.
35. Baqir M, Kluka EM, Aubry MC, et al. Amyloid-associated cystic lung disease in primary Sjögren's syndrome. *Respir Med.* 2013 Apr;107(4):616-21. Epub 2013 Feb 10.
36. Muzykewicz DA, Black ME, Muse V, et al. Multifocal micronodular pneumocyte hyperplasia: computed tomographic appearance and follow-up in tuberous sclerosis complex. *J Comput Assist Tomogr.* 2012;36(5):518.
37. Vassallo R, Jensen EA, Colby TV, et al. The overlap between respiratory bronchiolitis and desquamative interstitial pneumonia in pulmonary Langerhans cell histiocytosis: high-resolution CT, histologic, and functional correlations. *Chest.* 2003;124(4):1199.
38. Sirajuddin A, Raparia K, Lewis VA, et al. Primary Pulmonary Lymphoid Lesions: Radiologic and Pathologic Findings. *Radiographics.* 2016;36(1):53.
39. Avila NA, Kelly JA, Chu SC, et al. Lymphangioleiomyomatosis: abdominopelvic CT and US findings. *Radiology.* 2000 Jul;216(1):147-53.
40. Pavlovich CP, Grubb RL 3rd, Hurley K, et al. Evaluation and management of renal tumors in the Birt-Hogg-Dubésyndrome. *J Urol.* 2005;173(5):1482.
41. Young LR, Vandyke R, Gulleman PM, et al. Serum vascular endothelial growth factor-D prospectively distinguishes lymphangioleiomyomatosis from other diseases. *Chest.* 2010

- Sep;138(3):674-81. Epub 2010 Apr 9.
42. McCormack FX, Gupta N, Finlay GR, et al. Official American Thoracic Society/Japanese Respiratory Society Clinical Practice Guidelines: Lymphangioleiomyomatosis Diagnosis and Management. *Am J Respir Crit Care Med*. 2016 Sep;194(6):748-761.
 43. Hirose M, Matsumuro A, Arai T, et al. Serum vascular endothelial growth factor-D as a diagnostic and therapeutic biomarker for lymphangioleiomyomatosis. *PLoS One*. 2019;14(2):e0212776. Epub 2019 Feb 28.
 44. Gupta N, Finlay GA, Kotloff RM, et al. Lymphangioleiomyomatosis Diagnosis and Management: High-Resolution Chest Computed Tomography, Transbronchial Lung Biopsy, and Pleural Disease Management. An Official American Thoracic Society/Japanese Respiratory Society Clinical Practice Guideline. *Am J Respir Crit Care Med*. 2017;196(10):1337.
 45. Harari S, Torre O, Cassandro R, et al. Bronchoscopic diagnosis of Langerhans cell histiocytosis and lymphangioleiomyomatosis. *Respir Med*. 2012 Sep;106(9):1286-92. Epub 2012 Jul 7.
 46. Meraj R, Wikenheiser-Brokamp KA, Young LR, et al. Utility of transbronchial biopsy in the diagnosis of lymphangioleiomyomatosis. *Front Med*. 2012 Dec;6(4):395-405. Epub 2012 Dec 7.
 47. Koba T, Arai T, Kitaichi M, et al. Efficacy and safety of transbronchial lung biopsy for the diagnosis of lymphangioleiomyomatosis: A report of 24 consecutive patients. *Respirology*. 2018;23(3):331. Epub 2017 Sep 28.
 48. Taylor JR, Ryu J, Colby TV, et al. Lymphangioleiomyomatosis. Clinical course in 32 patients. *N Engl J Med* 1990; 323: 1254–1260.
 49. Urban T, Lazor R, Lacronique J, et al. Pulmonary lymphangioleiomyomatosis. A study of 69 patients. Groupe d'Etudes et de Recherche sur les Maladies "Orphelines" Pulmonaires (GERM" O" P). *Medicine* 1999; 78: 321–337.
 50. Taveira-DaSilva AM, Steagall WK, Moss J. Lymphangioleiomyomatosis. *Cancer Control* 2006; 13: 276–285.
 51. Radzikowska E. Lymphangioleiomyomatosis: new treatment perspectives. *Lung* 2015; 193: 467–475.
 52. Johnson SR, Cordier JF, Lazor R, et al. European Respiratory Society guidelines for the diagnosis and management of lymphangioleiomyomatosis. *Eur Respir J* 2010; 35: 14–26
 53. Taveira-DaSilva AM, Moss J. Management of lymphangioleiomyomatosis. *F1000Prime Rep* 2014; 6: 116.
 54. Maurer JR, Ryu J, Beck G, et al. Lung transplantation in the management of patients with lymphangioleiomyomatosis: baseline data from the NHLBI LAM Registry. *J Heart Lung Transplant* 2007; 26: 1293–1299
 55. Benden C, Rea F, Behr J, et al. Lung transplantation for lymphangioleiomyomatosis: the European experience. *J Heart Lung Transplant* 2009; 28: 1–7.
 56. Schönfeld N, Dirks K, Costabel U, et al. A prospective clinical multicentre study on adult pulmonary Langerhans' cell histiocytosis. *Sarcoidosis Vasc Diffuse Lung Dis* 2012; 29:132–138.
 57. Mogulkoc N, Veral A, Bishop PW, et al. Pulmonary Langerhans' cell histiocytosis: radiologic resolution following smoking cessation. *Chest* 1999; 115:1452–1455.
 58. Tazi A, Marc K, Dominique S, et al. Serial computed tomography and lung function testing in pulmonary Langerhans' cell histiocytosis. *Eur Respir J* 2012; 40:905–912.
 59. Lorillon G, Bergeron A, Detournignies L, et al. Cladribine is effective against cystic pulmonary Langerhans cell histiocytosis. *Am J Respir Crit Care Med* 2012; 186: 930–932.
 60. Lazor R, Etienne-Mastroianni B, Khouatra C, et al. Progressive diffuse pulmonary Langerhans cell histiocytosis improved by cladribine chemotherapy. *Thorax* 2009; 64: 274–275.
 61. Lorillon G, Jouenne F, Baroudjian B, et al. Response to trametinib of a pulmonary Langerhans cell histiocytosis harboring a MAP2K1 deletion. *Am J Respir Crit Care Med* 2018;198(5):675-678.

62. Boehler A, Speich R, Russi EW, et al. Lung transplantation for lymphangiomyomatosis. *N Engl J Med* 1996; 335: 1275–1280.
63. Tazi A, Montcelly L, Bergeron A, et al. Relapsing nodular lesions in the course of adult pulmonary Langerhans cell histiocytosis. *Am J Respir Crit Care Med* 1998; 157: 2007–2010.
64. Dal Sasso AA, Belem LC, Zanetti G, et al. Birt-Hogg-Dube syndrome: state-of-the-art review with emphasis on pulmonary involvement. *Respir Med* 2015; 109:289-296.
65. Gupta N, Seyama K, McCormack FX. Pulmonary manifestations of Birt-Hogg-Dube syndrome. *Fam Cancer* 2013; 12:387-396.
66. Ferreira Francisco FA, Soares Souza A Jr, Zanetti G, et al. Multiple cystic lung disease. *Eur Respir Rev* 2015; 24:552-564.
67. Stamatakis L, Metwalli AR, Middleton LA, et al. Diagnosis and management of BHD-associated kidney cancer. *Fam Cancer* 2013; 12:397-402.
68. Nicholson AG. Lymphocytic interstitial pneumonia and other lymphoproliferative disorders in the lung. *Semin Respir Crit Care Med* 2001; 22:409–422.
69. Guinee DG Jr. Update on nonneoplastic pulmonary lymphoproliferative disorders and related entities. *Arch Pathol Lab Med* 2010;134: 691–701
70. Gupta N, Wikenheiser-Brokamp KA, Fischer A, et al. Diffuse cystic lung disease as the presenting manifestation of Sjögren syndrome. *Ann Am Thorac Soc* 2016;13(3):371-375.
71. Dufour V, Wislez M, Bergot E, et al. Improvement of symptomatic human immunodeficiency virus-related lymphoid interstitial pneumonia in patients receiving highly active antiretroviral therapy. *Clin Infect Dis* 2003;36(10): e127-e130.