16. Bölüm

İDİYOPATİK PULMONER FİBROZİS TANISI, FENOTİPLERİ VE KOMORBİDİTELERİ

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İDİYOPATİK PULMONER FİBROZİS TANISI

Giriş

İdiopatik pulmoner fibrozis (İPF) nedeni bilinmeyen, akciğere sınırlı kronik fibrozan bir akciğer hastalığıdır. İdiopatik interstisyel pnömoniler grubunun en sık görülen hastalığı olup, patolojik olarak olağan interstisyel pnömoni (OİP) ile karakterizedir. Hastalık tanı sonrası 3-5 yıllık bir sürvi ile kötü bir prognoza sahiptir. İPF tanı ve tedavisi için ilk kez 2000 yılında ATS/ERS dernekleri bir rehber yayınlamışlardır. Bu rehber yeni bilgiler elde edildikçe güncellenmiş ve 2018 yılında son öneriler bildirilmiştir. Bu bölümde İPF ‹in klinik bulguları, yapılması gereken tetkikler ve tanısı tartışılacaktır.

Klinik bulgular

İPF hastaları tipik olarak 60 yaş üzerinde, erkek cinsiyetin daha baskın olduğu sigara içme hikâyesi olan kişilerdir. Hastalığın temel klinik bulguları eforla his-

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gelişimine katkıda bulunacağını düşünmektedir.[61]İPF olgularında klavuzlaragöre DM tedavisi ve sıkı kontrol yapılmalıdır.

Depresyon, Kaygı

Depresyon ve anksiyete İPF'de oldukça yaygındır ve özellikle şiddetli hastalıkla ilişkilidir.Depresyon prevalansı %21-49, anksiyete prevalansı %27- 31 civarında saptanmıştır. Depresyon ve anksiyetenin artmışsıklığı ve belirginpsikolojik etkileri nedeniyle, İPF'li tüm hastalar bu bozukluklar için taranmalıdır. Standart tedavi, bilişsel davranışçı terapi ve antidepresan ilaçları içerecektir, ancak bu tedavilerin etkinliği bu popülasyonda özel olarak doğrulanmamıştır.

Pulmoner rehabilitasyon; anksiyete ve depresyonun depresif semptomlarında ve ayrıca fonksiyonel iyileşmede sürekli bir iyileşme gösterdiğinden önerilir. Hasta destek grupları genellikle iyi bir psikolojik destek sağlarken, psikososyal destek de pulmoner rehabilitasyona katılım yoluyla elde edilebilir.[27,62,63]

SONUÇ

İPF tanısından itibaren bütüncül değerlendirilmesi gereken kompleks bir hastalıktır. Hastalığın tanısının multidisipliner bir ekiple konulması tanısal doğruluğu en yüksek düzeyde sağlayacaktır. Hastanın genel sağlık durumunun düzelmesi için eşlik eden hastalıklarının birlikte tedavi edilmesi gerekmektedir.

KAYNAKLAR

1. Tighe RM, Meltzer EB, Noble PW (2015). Idiopathic Pulmonary Fibrosis. In Grippi M, Elias J, Fishman J, Kotloff R, Pack A, Senior R, (Eds.), Fishman's pulmonary diseases and disorders. (5th edition pp. 842-58). New York: McGraw-Hill;
2. Fell CD, Martinez FJ, Liu LX, et al. Clinical determinants of the diagnosis of idiopathic pulmonary fibrosis. Am J Respir Crit Care Med 2010; 181: 832.
3. Wells AU, Hirani N, British Thoracic Society Interstitial Lung Disease Guideline Group. Interstitial Lung Disease Guideline. Thorax 2008;63(Suppl V):v1-v58.
4. Kim DS; Collard HR; King TE. Classification and natural history of the idiopathic interstitial pneumonias.Proc Am Thorac Soc. 2006; 3(4):285-92
5. King TE. Jr. (2011). İdiyopatik pulmoner fibroz. In Schwarz MI, King TE, Jr (Eds),Interstitial Lung Disease, (5th eds.p.895). People's Medical Publishing House-USA, Shelton, CT
6. Funke-Chambour M, Azzola A, Adler D, et al. Idiopathic Pulmonary Fibrosis in Switzerland: Diagnosis and Treatment Position Paper of the Swiss Working Group for Interstitial and Rare Lung Diseases of the Swiss Respiratory Society. Respiration 2017;93:363-78.
7. Fidler L, Doubelt I, Kandel S, Fisher JH, Mittoo S, Shapera S Screening for Myositis Antibodies in Idiopathic Interstitial Lung Disease. Lung. 2019;197(3):277. Epub 2019 Mar 5.
8. Drakopanagiotakis F, Wujak L, Wygrecka M, et al. Biomarkers in idiopathic pulmonary fibrosis Matrix Biol 2018; 68-69: 404-21.
9. Guiot J, Moermans C, Henket M, et al. Blood Biomarkers in Idiopathic Pulmonary Fibrosis. Lung 2017;195:273-80.

10. Raghu G, Remy-Jardin M, Myers JL, et al. Diagnosis of Idiopathic Pulmonary Fibrosis. An Official ATS/ERS/JRS/ALAT Clinical Practice Guideline. Am J Respir Crit Care Med 2018; 198: e44-e68.
11. Lynch JP, Saggar R, Weigt SS, et al. Usual interstitial pneumonia Semin Respir Crit Care Med 2006; 27: 634-51.
12. Lancaster LH. Utility of the six-minute walk test in patients with idiopathic pulmonary fibrosis. Multidiscip Respir Med 2018; 13: 45.
13. Hansell DM, Bankier AA, MacMahon H, McLoud TC, Müller NL, Remy J. Fleischner Society: glossary of terms for thoracic imaging. *Radiology* 2008;246:697–722
14. Watadani T, Sakai F, Johkoh T, *et al*. Interobserver variability in the CT assessment of honeycombing in the lungs. *Radiology* 2013;266:936–944.
15. Akira M, Kozuka T, Yamamoto S, et al. Computed tomography findings in acute exacerbation of idiopathic pulmonary fibrosis. Am J Respir Crit Care Med 2008;178:372–378.
16. Collard HR, Moore BB, Flaherty KR, et al.; Idiopathic Pulmonary Fibrosis Clinical Research Network Investigators. Acute exacerbations of idiopathic pulmonary fibrosis. Am J Respir Crit Care Med 2007;176:636–643.
17. Tcherakian C, Cottin V, Brillet et al. Progression of idiopathic pulmonary fibrosis: lessons from asymmetrical disease. Thorax 2011; 66: 226-31.
18. Oda T, Ogura T, Kitamura H, et al. Distinguishing features of usual interstitial pneumonia and pleuroparenchymal fibroelastosis compared with idiopathic pulmonary fibrosis. Chest 2014; 146: 1248.
19. Kebbe J, Abdo T. Interstitial lung disease: the diagnostic role of bronchoscopy. J Thorac Dis 2017; 9: 996-1010.
20. Ravaglia C, Wells AU, Tomassetti S, et al. Diagnostic yield and risk/ benefit analysis of trans-bronchial lung cryobiopsy in diffuse parenchymal lung diseases: a large cohort of 699 patients. BMC Pulm Med 2019; 19: 16.
21. Hetzel J, Maldonado F, Ravaglia C,et al. Transbronchial Cryobiopsies for the Diagnosis of Diffuse Parenchymal Lung Diseases: Expert Statement from the Cryobiopsy Working Group on Safety and Utility and a Call for Standardization of the Procedure. Respiration 2018; 95: 188-200.
22. Colella S, Haentschel M, Shah P, et al. Transbronchial Lung Cryobiopsy in Interstitial Lung Diseases: Best Practice. Respiration 2018; 95: 383-91.
23. Sethi J, Ali MS, Mohananey D, et al. Are Transbronchial Cryobiopsies Ready for Prime Time? : A Systematic Review and Meta-Analysis. J Bronchology Interv Pulmonol 2019; 26: 22-32.
24. Lynch DA, Sverzellati N, Travis WD, et al. Diagnostic criteria for idiopathic pulmonary fibrosis: Fleischner Community Technical Report. Lancet Respir Med 2018; 6: 138.
25. Fisher JH, Shapera S, To T, et al. Procedure volume and mortality after surgical lung biopsy in interstitial lung disease. Eur Respir J 2019; 53
26. Jaume Sauleda, Belén Núñez,Idiopathic Pulmonary Fibrosis: Epidemiology, Natural History, Phenotypes,Med Sci (Basel). 2018; 6(4): 110.
27. King CS, Nathan SD. Idiopathic pulmonary fibrosis: effects and optimal management of comorbidities. Lancet Respir Med. 2017 Jan;5(1):72-84. doi: 10.1016/S2213-2600(16)30222-3. Epub 2016 Sep 3. PMID: 27599614.
28. Brown A.W., Shlobin O.A., Weir N., et al. Dynamic patient counseling: A novel concept in idiopathic pulmonary fibrosis. Chest. 2012;142:1005–1010. doi: 10.1378/chest.12-0298
29. Buendía-Roldán I., Mejía M., Navarro C., et al. Idiopathic pulmonary fibrosis: Clinical behavior and aging associated comorbidities. Respir. Med. 2017;129:46–52. doi: 10.1016/j.rmed.2017.06.001
30. Sokai A., Tanizawa K., Handa T.et al. Importance of serial changes in biomarkers in idiopathic pulmonary fibrosis. ERJ Open Res. 2017;3 doi: 10.1183/23120541.00019-2016

31. Fell C.D. Idiopathic Pulmonary Fibrosis: Phenotypes and Comorbidities. Clin. Chest Med. 2012;33:51–57. doi: 10.1016/j.ccm.2011.12.005
32. Borie R., Kannengiesser C., Nathan N. et al. Familial pulmonary fibrosis. Rev. Des. Mal. Respir. 2015;32:413–434. doi: 10.1016/j.rmr.2014.07.017.
33. Cottin V. The impact of emphysema in pulmonary fibrosis. Eur. Respir. Rev. 2013;22:153–157. doi: 10.1183/09059180.00000813
34. Mejía M., Carrillo G., Rojas-Serrano J., et al. Idiopathic pulmonary fibrosis and emphysema: Decreased survival associated with severe pulmonary arterial hypertension. Chest. 2009;136:10–15. doi: 10.1378/chest.08-2306.
35. Ryerson CJ, Hartman T, Elicker BM. et al. Clinical features and outcomes in combined pulmonary fibrosis and emphysema in idiopathic pulmonary fibrosis. Chest 2013; 144: 234-40
36. Kimura M, Taniguchi H, Kondoh Y, et al. Pulmonary hypertension as a prognostic indicator at initial evaluation in idiopathic pulmonary fibrosis. Respiratory. 2013; 85 (6): 456--63.
37. Galiè N, Humbert M, Vachiery JL, et al. 2015 ESC / ERS Guidelines for the diagnosis and treatment of pulmonary hypertension: Joint Task Force of the European Society of Cardiology (ESC) and the European Respiratory Society (ERS) for the Diagnosis and Treatment of Pulmonary Hypertension: Approved by: European Union Pediatric and Congenital Cardiology (AEPC), International Heart and Lung Transplantation Association (ISHLT). Eur Heart J. 2016; 37 (1): 67--119.
38. Galie N., Humbert M., Vachieri JL. et al. ESC / ERS Guidelines for the diagnosis and treatment of pulmonary hypertension. EUR. Respir. J. 2015; 46: 903--975
39. Cano-Jiménez E, Hernández González F, Peloche GB. Comorbidities and Complications in Idiopathic Pulmonary Fibrosis Med Sci (Basel) 2018; 6: 71
40. Cano-Jiménez E, Hernández González F, Peloche GB. Comorbidities and Complications in Idiopathic Pulmonary Fibrosis Med Sci (Basel) 2018; 6: 71.
41. Hubbard R, Venn A, Lewis S, at al. Lung cancer and cryptogenic fibrosing alveolitis. A population-based cohort study. Am J Respir Crit Care Med 2000;161:5-8.
42. Karampitsakos T., Tzilas V., Tringidou R., et al. Lung cancer in patients with idiopathic pulmonary fibrosis. Pulm. Pharmacol. Ther. 2017; 45: 1–10.
43. Cottin V., Nunes H., Brillet PY, et al. Devouassoux G., Tillie-Leblond I., Israel-Biet D., Court-Fortune I., Valeyre D., Cordier JF, et al. Combined pulmonary fibrosis and emphysema: A prominent, poorly recognized entity. EUR. Respir. J. 2005; 26: 586--593.
44. Gao F, Hobson AR, Shang ZM, et al. The prevalence of gastro-esophageal reflux disease and esophageal dysmotility in Chinese patients with idiopathic pulmonary fibrosis.BMC Gastroenterol 2015;15:26.
45. Tcherakian C, Cottin V, Brillet PY, et al.. Progression of idiopathic pulmonary fibrosis: lessons from asymmetrical disease. *Thorax* 2011; 66: 226– 231
46. Raghu G, Yang ST, Spada C, et al. Sole treatment of acid gastroesophageal reflux in idiopathic pulmonary fibrosis: a case series. Chest 2006;129:794-800.
47. Lee JS, Collard HR, Anstrom KJ, et al. Anti-acid treatment and disease progression in idiopathic pulmonary fibrosis: an analysis of data from three randomised controlled trials. Lancet Respir Med2013;1:369-76.
48. Raghu G, Crestani B, Bailes Z, et al.Effect of anti-acid medication on reduction in FVC decline with nintedanib. Eur Respir J 2015;46:A4502.
49. Kreuter M, Wuyts W, Renzoni E, et al. Antacid therapy and disease outcomes in idiopathic pulmonary fibrosis: a pooled analysis. Lancet Respir Med. 2016 May;4(5):381-9. doi: 10.1016/S2213-2600(16)00067-9. Epub 2016 Mar 31. PMID: 27050871.
50. Raghu G. Collard HR, Egan JJ, et al. An offical ATS/ERS/JRS/ ALAT Clinical Practice Guideline. Treatment of Idiopathic pulmonary fibrosis. Am J Respir Crit Care Med 2015;192:3-19.
51. Sprunger DB, Olson AL, Huie TJ, et al. Pulmonary fibrosis is associated with an elevated risk

of thromboembolic disease. Eur Respir J. 2012;39(1):125–32.
52. Noth I, Anstrom KJ, Calvert SB, et al. A placebo-controlled randomized trial of warfarin in idiopathic pulmonary fibrosis. Am J Respir Crit Care Med. 2012;186(1):88–95.
53. Kreuter M, Wijsenbeek MS, Vasakova M, et al. Unfavourable effects of medically indicated oral anticoagulants on survival in idiopathic pulmonary fibrosis: methodological concerns. Eur Respir J. 2016;48(5):1524–6.
53. King TE Jr, Albera C, Bradford WZ, et al. All-cause mortality rate in patients with idiopathic pulmonary fibrosis. Implications for the design and execution of clinical trials. Am J Respir Crit Care Med. 2014;189(7):825–31.
54. Kim WY, Mok Y, Kim GW, et al. Association between idiopathic pulmonary fibrosis and coronary artery disease: a case-control study and cohort analysis. Sarcoidosis Vasc Diffuse Lung Dis. 2015;31(4):289–96.
55. Zisman DA, Kawut SM. Idiopathic pulmonary fibrosis: a shot through the heart? Am J Respir Crit Care Med. 2008;178(12):1192–3.
56. Troy LK, Corte TJ. Sleep disordered breathing in interstitial lung disease: a review. World J Clin Cases 2014;2:828-34.
57. Pıhtılı A, Bingol Z, Kıyan E, et al. Obstructive sleep apnea is common in patients with interstitial lung disease. Sleep Breath 2013; 17: 1281-8.
58. Corte T.J., Wort S.J., Talbot S., et al. Elevated nocturnal desaturation index predicts mortality in interstitial lung disease. Sarcoidosis Vasc. Diffus. Lung Dis. 2012;29:41–50
59. Torrisi SE, Palmucci S, Stefano A, et al. Assessment of survival in patients with idiopathic pulmonary fibrosis using quantitative HRCT indexes. Multidiscip Respir Med 2018; 13: 43.
60. Gribbin J, Hubbard R, Smith C. Role of diabetes mellitus and gastro-oesophageal reflux in the aetiology of idiopathic pulmonary fibrosis. Respir Med. 2009;103(6):927–31.
61. Kim YJ, Park JW, Kyung SY, et al. Clinical characteristics of idiopathic pulmonary fibrosis patients with diabetes mellitus: the national survey in Korea from 2003 to 2007. J Korean Med Sci. 2012;27(7):756–60.
62. Ryerson CJ, Cayou C, Topp F, et al. Pulmonary rehabilitation improves long-term outcomes in interstitial lung disease: a prospective cohort study. Respir Med. 2014;108(1):203–10.
63. Holland AE, Fiore JF Jr, Bell EC, et al. Dyspnoea and comorbidity contribute to anxiety and depression in interstitial lung disease. Respirology 2014;19:1215-21.