

BÖLÜM 21

SİSTEMİK HASTALIK VE SENDROMLARIN OTOLOJİK BULGULARI

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GİRİŞ

Granüloamatöz ve enfeksiyöz hastalıklar, neoplaziler, kemik bozuklukları, metabolik ve depo hastalıkları, otoimmün hastalıklar ve immün yetmezlikler dahil olmak üzere pek çok çeşitli sistemik hastalık otolojik semptom ve bulgulara yol açabilir (1). Otolojik semptom ve bulgular bu hastalıklarda az görülebilir veya hastalığın ilk ve tek özelliği olarak ortaya çıkabilir. Tutulan otolojik bölgenin yeri ve yaygınlığına göre farklı semptomlar ortaya çıkabilmektedir. Akut veya kronik otitis media, iletim tipi, sensörinöral ve miks tip işitme kaybı, vestibüler belirtiler, otalji, ani işitme kaybı, fasiyal sinir paralizisi görülebilir (1).

Otolojik belirtiler üzerinden sistemik veya sendromik hastalık tanısı koymak zor olabilir, bu nedenle öncelikle ön tanı açısından şüphelenmek, laboratuvar ve radyolojik tetkik ile biyopsi gibi tanısal yaklaşım gerekir. Bu hastalık grubu hasta bazlı olarak ve multidisipliner yaklaşımla değerlendirilmelidir. Bu bölümde otolojik semptom ve bulguları olan sistemik hastalıklar (Tablo 1) ve sendromlar (Tablo 2) tartışılmıştır.

GRANÜLOMATÖZ HASTALIKLAR

Langerhans Hücreli Histiositoz

Eozinofilik granülom olarak da bilinen bu hastalığın, immünolojik disfonksiyon sonucu patolojik Langerhans hücrelerinin çoğalmasına bağlı geliştiği düşünülmektedir. Çocuk ve genç erişkinlerde iskelet sisteminin birçok lokasyonunda görülebilir. Mandibula, maksilla ve temporal kemiklerde de osteolitik lezyonlara neden olabilir. Bu lezyonlar asemptomatik olabilir veya ağrı, şişlik ve patolojik kırıklara yol açabilir. Temporal kemikte kötü prognozlu litik lezyonlara neden olur (1).

Otolojik bulgular: Hastalığın ilk işareti olabilir. En sık görülen semptomlar otore, postauriküler şişlik, işitme kaybı ve vertigodur. En sık bulgu olarak dış kulak yolunda granülasyon dokusu veya polipler izlenir. Ayrıca timpanik zar perforasyonu, otitis media, otitis eksterna, mastoid kemik ile dış kulak arasında fistül ve postauriküler şişlik görülebilir. Mastoid kemikte düzensiz litik lezyonlar izlenir. Nadiren fasiyal paralizisi, kafa tabanı ve juguler fossa tutulumu görülebilir. Bu hastalık sıklıkla kronik otitis

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KAYNAKLAR

1. Nadol JB, Quesnel AM, (2021), *Otologic Manifestations of Systemic Disease*, Paul W. Flint, Howard W. Francis, Burce H. Haughey, Marci M. Lesperance, Valerie J. Lund, K. Thomas Robbins, J. Regan Thomas (Eds.), Cummings Otolaryngology Head and Neck Surgery (7th ed, Chapter.151, pp.2293-2310). Philadelphia: Elsevier.
2. Pasha R, Golub JS, (2018), *Otolaryngology Head and Neck Surgery Clinical Referans Guide*, 5th edition, Plural Publishing.
3. Costa CF, Polanski JF, Wegener Granulomatosis: Otologic Manifestation as First Symptom, *Int Arch Otorhinolaryngol* 2015; 19: 266–268.
4. Naples JG, Brant JA, Ruckenstein MJ, (2021), *Autoimmune Inner Ear Disease*, Paul W. Flint, Howard W. Francis, Burce H. Haughey, Marci M. Lesperance, Valerie J. Lund, K. Thomas Robbins, J. Regan Thomas (Eds.), Cummings Otolaryngology Head and Neck Surgery (7th ed, Chapter.156, pp.2365-2369). Philadelphia: Elsevier.
5. Yoon TH, Paparella MM, Schachren PA, Systemic Vasculitis: A Temporal Bone Histopathologic Study, *Laryngoscope*, 99: June 1989.
6. Lang EE, Zaruk JE, Colreavy MP, et.al, An unusual case of external ear inflammation caused by sarcoidosis, *ENT·Ear, Nose & Throat Journal*· December 2003.
7. Ralli M, D'Aguzzo V, Di Stadio A, et.al. Audiovestibular Symptoms in Systemic Autoimmune Diseases, *Journal of Immunology Research*, Volume: 2018.
8. Salvinelli F, Cancilleri F, Casale M, et.al. Hearing thresholds in patients affected by rheumatoid arthritis, *Clin. Otolaryngol.* 29. 2004.
9. Rubin F, Khai Hoan NT, Bonfils P, Sudden bilateral hearing loss revealing polyarteritis nodosa, *European Annals of Otorhinolaryngology, Head and Neck diseases*, 131, (2014) 265-266.
10. Crane BT, Minor LB, (2021) *Peripheral Vestibular Disorders*, Paul W. Flint, Howard W. Francis, Burce H. Haughey, Marci M. Lesperance, Valerie J. Lund, K. Thomas Robbins, J. Regan Thomas (Eds.), Cummings Otolaryngology Head and Neck Surgery, (7th ed, Chapter.167, pp.2517-2535). Philadelphia: Elsevier.
11. Gurrola JG, Pletcher SD, Goldberg AN, Head and Neck Manifestations in the Immunocompromised Host, Paul W. Flint, Howard W. Francis, Burce H. Haughey, Marci M. Lesperance, Valerie J. Lund, K. Thomas Robbins, J. Regan Thomas (Eds.), Cummings Otolaryngology Head and Neck Surgery, (7th ed, Chapter.10, pp.155-179). Philadelphia: Elsevier.
12. Woolf NK, Experimental Congenital Cytomegalovirus Labyrinthitis and Sensorineural Hearing Loss, *Am. J. Otolaryngol.* 11:299-303, 1990.
13. Li W, Schachern PA, Morizono T, Paparella MM, The Temporal Bone in Multiple Myeloma, *Laryngoscope* 104: June 1994.
14. Masuda A, Fisher LM, Oppenheimer ML, Hearing Changes after Diagnosis in Neurofibromatosis Type 2, *Otology & Neurotology*, 25:150–154, 2004.
15. Applebaum EL, Clemis JD, Temporal Bone Histopathology of Paget's Disease With Sensorineural Hearing Loss and Narrowing of The Internal Auditory Canal, *The Laryngoscope* 87:1977.
16. Monsell EM, The Mechanism of Hearing Loss in Paget's Disease of Bone, *Laryngoscope* 114: April 2004.
17. Santos F, McCall AA, Chien W, Merchant SN, Otopathology in Osteogenesis Imperfecta, *Otol Neurotol.* 2012, December; 33(9): 1562–1566.
18. Zhai X, Duan L, Yao Y, et. al. Clinical Characteristics and Management of Patients With McCune-Albright Syndrome With GH Excess and Precocious Puberty: A Case Series and Literature Review, *frontiers in Endocrinology*, Volume 12, October 2021.
19. Boyce AM, Brewer C, DeKlotz TR, et.al. Association of Hearing Loss and Otologic Outcomes With Fibrous Dysplasia, *JAMA Otolaryngol Head Neck Surg*, 2018, Feb; 144 (2): 102-107.
20. Stocks RM, Wang WC, Thompson JW, et.al. Malignant Infantile Osteopetrosis Otolaryngological Complications and Management, *Arch Otolaryngol Head and Neck Surg.* 1998; 124:689-694.
21. Foster HE, Rabinovich CE, Musculoskeletal Manifestations of Systemic Disease, Elizabeth D. Mellins, Lucy Wedderburn, Rose E Petty, *Textbook of Pediatric Rheumatology*, (8th ed, Chapter.52, pp.691-701). Elsevier.
22. Chen YC, Huang CY, Lee YT, et.al. Audiological and otologic manifestations of glutaric aciduria type I, *Orphanet J Rare Dis* (2020) 15:337.
23. Yoshida N, Iino Y, Pathogenesis and Diagnosis of Otitis Media with ANCA-Associated Vasculitis *Allergology International*, 2014, 63: 523-532.
24. Maiolino L, Cocuzza S, Conti A, et.al. Autoimmune ear disease: clinical and diagnostic relevance in Cogan's syndrome, *Audiology*

- Research, vol: 7: 162, 2017.
25. Schuknecht HF, Nadol JB, Temporal Bone Pathology in a Case of Cogan's Syndrome, *Laryngoscope* 104: September 1994.
 26. West SG, (2019), Systemic Diseases in Which Arthritis Is a Feature, Lee Goldman, Andrew I. Schaffer, *Goldman-Cecil Medicine*, (26th ed, Chapter.259, pp.1778-1783). Elsevier.
 27. Di Stadio A, Dipietro L, Ralli M, Sudden Hearing Loss As An Early Detector of Multiple Sclerosis: A Systematic Review, *European Review for Medical and Pharmacological Sciences*, 2018;22:4611-4624.
 28. Di Stadio A, Ralli M, Systemic Lupus Erythematosus and hearing disorders: Literature review and meta-analysis of clinical and temporal bone findings, *Journal of International Medical Research* 2017, Vol. 45(5) 1470-1480.
 29. González JLT, Torres JR, Ríos YH, et.al. Extended high-frequency audiometry as early detection of hearing loss in primary Sjögren syndrome, *Clin Rheumatol*, DOI 10.1007/s10067-017-3750-2.
 30. Patel VA, Dunklebarger M, Zacharia TT, et.al, Otolgic manifestations of Susac syndrome, *Acta Otorhinolaryngol Ital*, 2018; 38:544-553.
 31. Choung YH, Cho MJ, Park K, et.al. Audio-Vestibular Disturbance in Patients with Behçet's Disease, *Laryngoscope*, 116: November 2006.
 32. Ren Q, Su J, Zhang D, Ding X, Otolgical IgG4-Related Disease With Inner Ear Involvement: A Case Report and Review of Literature *Ear, Nose & Throat Journal*, 1-4, 2020.
 33. Cho HK, Lee YJ, Chung JH, Koo JW, Otolgic Manifestation in IgG4-Related Systemic Disease Clinical and Experimental Otorhinolaryngology Vol. 4, No. 1: 52-54, March 2011.
 34. Haddad J, Dodhia SN, Spitzer JB, (2019), Hearing Loss, Robert M. Kliegman, Bonita M.D. Stanton, Joseph St. Geme, Nina F Schor, *Nelson Textbook of Pediatrics*, (21st ed, Chapter.655, pp.3400-3411). Elsevier.
 35. Bansal M, (2013), Diseases of Ear Nose and Throat Head and Neck Surgery, First Edition, Chapter 14:166-168, Jaypee.
 36. Boisa E, Nassara M, Zenatyb D, et.al. Otolgic disorders in Turner syndrome, *European Annals of Otorhinolaryngology, Head and Neck diseases* 135 (2018) 21-24.
 37. Roberts AE, Allanson JE, Tartaglia M, Gelb BD, Noonan Syndrome, *Lancet*. 2013 January 26; 381(9863): 333-342.
 38. Verheij E, Kist AL, Molen M, et.al. Otolgic and audiolgic findings in 22q11.2 deletion syndrome, *Eur Arch Otorhinolaryngol* (2017) 274:765-771.
 39. Wenger TL, Hing AV, Evans KN, Apert Syndrome, *GeneReviews*, 2019.
 40. Guardiani E, Zalewski C, Brewer C, et.al, Otolgic and Audiolgic Manifestations of Hutchinson-Gilford Progeria Syndrome, *Laryngoscope*, 2011 October; 121(10): 2250-2255.
 41. You YS, Park SW, Yun SK, Lee EJ, Bilateral sudden sensorineural hearing loss with Sweet syndrome, *Medicine*, 2020, 99:36.
 42. Lindsey S, Brewer C, Stakhovskaya O, et.al. Auditory and Otolgic Profile of Alström Syndrome: Comprehensive Single Center Data on 38 Patients, *Am J Med Genet A*. 2017 August; 173(8): 2210-2218.