BÖLÜM 26

BAŞ-BOYUN KEMİK SARKOMLARI

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GİRİŞ

Sarkomlar, mezenkimal dokudan köken alan, nadir rastlanan ve içinde çok çeşitli tümörlerin olduğu bir malignite grubunu temsil eder. Sarkomlar, pediatrik popülasyon dışında baş ve boyun bölgesinde primer maligniteler olarak genellikle karşımıza çıkmazlar. Baş ve boyun sarkomları (BBS), tüm baş-boyun kanserlerinin sadece %1'ine tekabül eder (1), ancak önemli bir morbidite ve mortalite kaynağı olması sebebiyle baş-boyun cerrahlarına zorluk oluşturmaya devam etmektedir. Çoğu çalışmada, BBS tüm yetişkin sarkom vakalarının %5-15'ini temsil etmektedir (2,3). Ancak pediatrik yaşta tüm sarkomların %35'i baş ve boyunda ortaya çıkar (4,5). BBS'ler, türetildiği mezenkimal hücrelere bağlı olarak kemik-kıkırdak veya yumuşak doku elemanlarından kaynaklanabilir ve yumuşak doku orijinli olanlar kemik-kıkırdak orijinlilere göre 4 kat daha fazla görülür (1).

Bu bölümün ilerleyen kısımlarında baş ve boyunda görülen kemik sarkomlarının semptomatolojisi, muayene bulguları, görüntüleme teknikleri ve tanı metodları ile ilgili bilgiler verilecek ardından en sık görülen baş-boyun kemik sarkomları (osteosarkom, kondrosarkom, ewing sarkom, kordoma) hakkında daha spesifik bilgiler sunulacaktır.

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görülme eğilimindedir (56). Ayırıcı tanıda; KS, meningioma, miyoepitelyoma/ miyoepitelyal karsinom, glioma, metastatik karsinomlar düşünülmelidir. Kordomalar için birincil tedavi yöntemi tipik olarak cerrahi rezeksiyon ve ardından adjuvan RT'den oluşur (56). Klasik olarak kordomalar KT'ye duyarlı değildir. EGFR inhibitörlerinin yanı sıra imatinibin etkinliğini değerlendiren çalışmalar, özellikle lokal olarak ilerlemiş ve metastatik kordomada umut vericidir (57, 58).

Baş ve boyun bölgesinde yukarıda anlatılan kemik sarkomlarının dışında fibrosarkoma, epiteloid hemanjioendotelyoma, anjiosarkoma, kemiğin malign dev hücreli tümörü, undiferansiye pleomorfik sarkoma, kemik metastazları görülebilir (59).

SONUÇ

Baş ve boyun kemik sarkomları çok nadir görülen ve kulak burun boğaz hekimleri için yönetimi zor olan tümörlerdir. En sık görülen histopatolojik alt tip osteosarkomdur. Hangi kemik sarkomu olursa olsun primer tedavi yöntemi cerrahidir bunu RT ve/veya KT takip edebilir. Baş ve boyun kemik sarkomları için tedavi başarısızlığının klasik belirleyicileri büyük tümör boyutu, yüksek gradeli histoloji ve pozitif cerrahi sınırlardır.

KAYNAKLAR

1. Sturgis EM, Potter BO. Sarcomas of the head and neck region. Curr Opin Oncol. 2003;15 (3):239-252. Doi: 10.1097/00001622-200305000-00011.
2. Kraus DH, Dubner S, Harrison LB, et al. Prognostic factors for recurrence and survival in head and neck soft tissue sarcomas. Cancer. 1994;74 (2):697-702. Doi: 10.1002/1097-0142 (19940715)74:2<697::aid-cncr2820740224>3.0.co;2-a.
3. Wanebo HJ, Koness RJ, MacFarlane JK,. Head and neck sarcoma: report of the Head and Neck Sarcoma Registry. Society of Head and Neck Surgeons Committee on Research. Head Neck. 1992;14 (1):1-7. Doi: 10.1002/hed.2880140102.
4. Lawrence W Jr, Anderson JR, Gehan EA, et al. Pretreatment TNM staging of childhood rhabdomyosarcoma: a report of the Intergroup Rhabdomyosarcoma Study Group. Children's Cancer Study Group. Pediatric Oncology Group. Cancer. 1997;80 (6):1165-1170.
5. Lyos AT, Goepfert H, Luna MA, et al. Soft tissue sarcoma of the head and neck in children and adolescents. Cancer. 1996;77 (1):193-200. Doi: 10.1002/ (SICI)1097-0142 (19960101)77:1<193::AID-CNCR31>3.0.CO;2-Z.
6. Klein MJ, Siegal GP. Osteosarcoma: anatomic and histologic variants. Am J Clin Pathol. 2006;125 (4):555-581. Doi: 10.1309/UC6K-QHLD-9LV2-KENN.
7. Mendenhall WM, Fernandes R, Werning JW, et al. Head and neck osteosarcoma. Am J Otolaryngol. 2011;32 (6):597-600. Doi: 10.1016/j.amjoto.2010.09.002.
8. Xie Y, Zhang Z, Hong J, et al. Furazolidone-containing triple and quadruple eradication therapy for initial treatment for Helicobacter pylori infection: A multicenter randomized controlled trial in China. Helicobacter. 2018;23 (5):e12496. Doi: 10.1111/hel.12496.

9. Liang L, Zhang T, You Y, et al. An individual patient data meta-analysis on the effect of chemotherapy on survival in patients with craniofacial osteosarcoma. Head Neck. 2019;41 (6):2016-2023. Doi: 10.1002/hed.25668.
10. Chen Y, Gokavarapu S, Shen Q, et al. Chemotherapy in head and neck osteosarcoma: Adjuvant chemotherapy improves overall survival. Oral Oncol. 2017;73:124-131. Doi: 10.1016/j.oraloncology.2017.08.017.
11. Guadagnolo BA, Zagars GK, Raymond AK, et al. Osteosarcoma of the jaw/craniofacial region: outcomes after multimodality treatment. Cancer. 2009;115 (14):3262-3270. Doi: 10.1002/cncr.24297.
12. O'Neill JP, Bilsky MH, Kraus D. Head and neck sarcomas: epidemiology, pathology, and management. Neurosurg Clin N Am. 2013;24 (1):67-78. Doi: 10.1016/j.nec.2012.08.010.
13. Kalavrezos N, Sinha D. Head and neck sarcomas in adulthood: current trends and evolving management concepts. Br J Oral Maxillofac Surg. 2020;58 (8):890-897. Doi: 10.1016/j.bjoms.2020.05.015.
14. Boon E, van der Graaf WT, Gelderblom H, et al. Impact of chemotherapy on the outcome of osteosarcoma of the head and neck in adults. Head Neck. 2017;39 (1):140-146. Doi: 10.1002/hed.24556.
15. Ha PK, Eisele DW, Frassica FJ, et al. Osteosarcoma of the head and neck: a review of the Johns Hopkins experience. Laryngoscope. 1999;109 (6):964-969. Doi: 10.1097/00005537-199906000-00023.
16. Mark RJ, Sercarz JA, Tran L, et al. Osteogenic sarcoma of the head and neck. The UCLA experience. Arch Otolaryngol Head Neck Surg. 1991;117 (7):761-766. Doi: 10.1001/archotol.1991.01870190073015.
17. Kneisl, J.S., Rosenberg, A.E., Anderson, P.M., et al. (2017). Bone. M.B. Amin (Ed.), In: AJCC Cancer Staging Manual, AJCC, (8th ed. p:471). Chicago: Springer.
18. Chen Y, Shen Q, Gokavarapu S, et al. Osteosarcoma of head and neck: a retrospective study on prognostic factors from a single institute database.Oral oncol, 2016; 58, 1-7. Doi:10.1016/j.oraloncology.2016.04.008
19- Whelan JS, Bielack SS, Marina N, et al. EURAMOS-1, an international randomised study for osteosarcoma: results from pre-randomisation treatment. Ann Oncol. 2015;26 (2):407-414. Doi: 10.1093/annonc/mdu526.
20. Smith RB, Apostolakis LW, Karnell LH, et al. National Cancer Data Base report on osteosarcoma of the head and neck. Cancer. 2003;98 (8):1670-1680. Doi: 10.1002/cncr.11716.
21. Anninga JK, Gelderblom H, Fiocco M, et al. Chemotherapeutic adjuvant treatment for osteosarcoma: where do we stand? Eur J Cancer. 2011;47 (16):2431-2445. Doi: 10.1016/j.ejca.2011.05.030.
22. Coca-Pelaz A, Rodrigo JP, Triantafyllou A, et al. Chondrosarcomas of the head and neck. Eur Arch Otorhinolaryngol. 2014;271 (10):2601-2609. Doi: 10.1007/s00405-013-2807-3.
23. Koch BB, Karnell LH, Hoffman HT et al. National cancer database report on chondrosarcoma of the head and neck. Head Neck. 2000;22:408–425. Doi: 10.1002/1097-0347(200007)22:4<408::aid-hed15>3.0.co;2-h.
24. Lee SY, Lim YC, Song MH et al. Chondrosarcoma of the head and neck. Yonsei Med J. 2005;46:228–232. Doi: 10.3349/ymj.2005.46.2.228.
25. Mark RJ, Tran LM, Sercarz J, et al. Chondrosarcoma of the head and neck. The UCLA experience, 1955-1988. Am J Clin Oncol. 1993;16 (3):232-237. Doi: 10.1097/00000421-199306000-00008.

26. Filiz, G., & Yerci, Ö. (2003). Baş-boyunda ender görülen tümörler. In Kayıhan, E., & Levent E. (Eds.), Baş-Boyun Kanserleri (1st ed., pp. 673-693). Istanbul: Nobel Tıp Kitabevleri.
27. Ogalai, J.S. (2005). Kafa tabanı ve temporal kemik tümörleri. In Anil, K.L. (Ed). Current Otorinolaringoloji – Baş ve Boyun Cerrahisi Tanı ve Tedavi (1st ed., pp. 845-864). Ankara: Güneş Kitabevi.
28. Picket, B.P., & Kelly, J.P. (2011). Lateral kafa tabanı ve kulak tümörleri. In Byron, J.B., & Jonas, T.J. (Eds). Baş & Boyun Cerrahisi – Otolarengoloji (4th ed. pp. 2003-2025). Ankara: Güneş Tıp Kitapevleri.
29. de Souza LL, Pontes FSC, Fonseca FP, et al. Chondrosarcoma of the jaw bones: a review of 224 cases reported to date and an analysis of prognostic factors. Int J Oral Maxillofac Surg. 2019;48 (4):452-460. Doi: 10.1016/j.ijom.2018.11.006.
30. Gawande M, Swastika N, Chaudhary M, et al. Chondrosarcoma of maxilla. J Oral Maxillofac Pathol. 2014;18 (3):423-427. Doi: 10.4103/0973-029X.151338
31. Choi J, Dharmarajan H, Victores A, et al. Chondrosarcoma of the Epiglottis: A Case Report and Literature Review. J Voice 2018;32 (4):484-487. Doi: 10.1016/j.jvoice.2017.07.014.
32. El Naggar AK, Chan JKC, Grandis JR, et al. (2017). Chapter 8: Odontogenic and maxillofacial bone tumours. WHO Classification of Head and Neck Tumours. (4th ed, pp. 205-260). Lyon: IARC.
33. Evans HL, Ayala AG, Romsdahl MM. Prognostic factors in chondrosarcoma of bone: a clinicopathologic analysis with emphasis on histologic grading. Cancer. 1977;40:818–831. Doi: 10.1002/1097-0142 (197708)40:2<818::aid-cncr2820400234>3.0.co;2-b.
34. Bloch OG, Jian BJ, Yang I, et al. A systematic review of intracranial chondrosarcoma and survival. J Clin Neurosci. 2009;16 (12):1547-1551. Doi: 10.1016/j.jocn.2009.05.003.
35. Italiano A, Mir O, Cioffi A, et al. Advanced chondrosarcomas: role of chemotherapy and survival. Ann Oncol. 2013;24 (11):2916-2922. Doi: 10.1093/annonc/mdt374.
36. Harnsberger, H.R., Hudgins, P.A., Wiggins, R.H., & Davidson, H.C. (2011). PocketRadiologist Baş ve Boyun En sık 100 Tanı (1st ed). İstanbul: Nobel Tıp Kitabevleri
37. Delattre O, Zucman J, Melot T, et al. The Ewing family of tumors--a subgroup of small-round-cell tumors defined by specific chimeric transcripts. N Engl J Med. 1994;331 (5):294-299. Doi: 10.1056/NEJM199408043310503.
38. Grier HE. The Ewing family of tumors. Ewing's sarcoma and primitive neuroectodermal tumors. Pediatr Clin North Am. 1997;44 (4):991-1004. Doi: 10.1016/s0031-3955 (05)70541-1.
39. Arora RS, Alston RD, Eden TO, et al. The contrasting age-incidence patterns of bone tumours in teenagers and young adults: Implications for aetiology. Int J Cancer. 2012;131 (7):1678-1685. doi: 10.1002/ijc.27402.
40. Duchman KR, Gao Y, Miller BJ. Prognostic factors for survival in patients with Ewing's sarcoma using the surveillance, epidemiology, and end results (SEER) program database. Cancer Epidemiol. 2015;39 (2):189-195. Doi: 10.1016/j.canep.2014.12.012.
41. van den Berg H, Kroon HM, Slaar A, et al. Incidence of biopsy-proven bone tumors in children: a report based on the Dutch pathology registration "PALGA". J Pediatr Orthop. 2008;28 (1):29-35. Doi: 10.1097/BPO.0b013e3181558cb5.
42. Cotterill SJ, Ahrens S, Paulussen M, et al. Prognostic factors in Ewing's tumor of bone: analysis of 975 patients from the European Intergroup Cooperative Ewing's Sarcoma Study Group. J Clin Oncol. 2000;18 (17):3108-3114. Doi: 10.1200/JCO.2000.18.17.3108.
43. Whaley JT, Indelicato DJ, Morris CG, et al. Ewing tumors of the head and neck. Am J Clin Oncol. 2010;33 (4):321-326. Doi: 10.1097/COC.0b013e3181aaca71.

44. Bouaoud J, Temam S, Cozic N, et al. Ewing's Sarcoma of the Head and Neck: Margins are not just for surgeons. Cancer Med. 2018;7 (12):5879-5888. Doi: 10.1002/cam4.1801.
45. Ozaki T. Diagnosis and treatment of Ewing sarcoma of the bone: a review article. J Orthop Sci. 2015;20 (2):250-263. doi: 10.1007/s00776-014-0687-z.
46. Györke T, Zajic T, Lange A, et al. Impact of FDG PET for staging of Ewing sarcomas and primitive neuroectodermal tumours. Nucl Med Commun. 2006;27 (1):17-24. Doi: 10.1097/01.mnm.0000186608.12895.69.
47. Olson MD, Van Abel KM, Wehrs RN, et al. Ewing sarcoma of the head and neck: The Mayo Clinic experience. Head Neck. 2018;40 (9):1999-2006. Doi: 10.1002/hed.25191.
48. Juergens C, Weston C, Lewis I, et al. Safety assessment of intensive induction with vincristine, ifosfamide, doxorubicin, and etoposide (VIDE) in the treatment of Ewing tumors in the EURO-E.W.I.N.G. 99 clinical trial. Pediatr Blood Cancer. 2006;47 (1):22-29. Doi: 10.1002/pbc.20820.
49. Foulon S, Brennan B, Gaspar N, et al. Can postoperative radiotherapy be omitted in localised standard-risk Ewing sarcoma? An observational study of the Euro-E.W.I.N.G group. Eur J Cancer. 2016;61:128-136. Doi: 10.1016/j.ejca.2016.03.075.
50. Le Deley MC, Paulussen M, Lewis I, et al. Cyclophosphamide compared with ifosfamide in consolidation treatment of standard-risk Ewing sarcoma: results of the randomized noninferiority Euro-EWING99-R1 trial. J Clin Oncol. 2014;32 (23):2440-2448. Doi: 10.1200/JCO.2013.54.4833.
51. Whelan JS, Jinks RC, McTiernan A, et al. Survival from high-grade localised extremity osteosarcoma: combined results and prognostic factors from three European Osteosarcoma Intergroup randomised controlled trials. Ann Oncol. 2012;23 (6):1607-1616. Doi: 10.1093/annonc/mdr491.
52. Dirksen U, Le Deley MC, Brennan B, et al. Efficacy of busulfan-melphalan high dose chemotherapy consolidation (BuMel) compared to conventional chemotherapy combined with lung irradiation in ewing sarcoma (ES) with primary lung metastases: Results of EURO-EWING 99-R2pulm randomized trial (EE99R2pul). J Clin Oncol. 2016;34:15_suppl:11001.
53. Ladenstein R, Pötschger U, Le Deley MC, et al. Primary disseminated multifocal Ewing sarcoma: results of the Euro-EWING 99 trial. J Clin Oncol. 2010;28 (20):3284-3291. Doi: 10.1200/JCO.2009.22.9864.
54. Bosma SE, Ayu O, Fiocco M, et al. Prognostic factors for survival in Ewing sarcoma: A systematic review. Surg Oncol. 2018;27 (4):603-610. Doi: 10.1016/j.suronc.2018.07.016.
55. McMaster ML, Goldstein AM, Bromley CM, et al. Chordoma: incidence and survival patterns in the United States, 1973-1995. Cancer Causes Control. 2001;12 (1):1-11. Doi: 10.1023/a:1008947301735.
56. Wasserman JK, Gravel D, Purgina B. Chordoma of the Head and Neck: A Review. Head Neck Pathol. 2018;12 (2):261-268. Doi: 10.1007/s12105-017-0860-8.
57. Hindi N, Casali PG, Morosi C, et al. Imatinib in advanced chordoma: A retrospective case series analysis. Eur J Cancer. 2015;51 (17):2609-2614. Doi: 10.1016/j.ejca.2015.07.038.
58. Launay SG, Chetaille B, Medina F, et al. Efficacy of epidermal growth factor receptor targeting in advanced chordoma: case report and literature review. BMC Cancer. 2011;11:423. Doi: 10.1186/1471-2407-11-423.
59. Choi JH, Ro JY. The 2020 WHO Classification of Tumors of Bone: An Updated Review. Adv Anat Pathol. 2021;28 (3):119-138. Doi: 10.1097/PAP.0000000000000293.