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GİRİŞ

Yumuşak doku tümörleri maligniteler içerisinde ender görülen tümörlerdendir. Yumuşak doku sarkomları embriyolojik gelişim sırasında mezenkimal hücrelerden gelişen fibröz doku, yağ dokusu, çizgili kas, düz kas, vasküler ve sinir dokularından kaynaklanırlar. Erişkin kanserlerinin yaklaşık %0,7 sini, çocukluk çağının kanserlerinin ise yaklaşık %6,5' ini oluşturan nadir görülen kanserlerdendir. Sarkomlar, yumuşak doku sarkomları ve primer kemik sarkomları olarak iki temel gruba ayrırlırlar. Tümör alt gruplarının türüne, yerleşim yerine ve histopatolojik özelliklerine göre birbirinden farklı klinik semptomatoloji, tedavi ve doğal seyir gösterirler (1-4).

Sarkomlarda semptomatolojiyi belirleyen faktörler, tümörün orijin aldığı bölge, komşu organ tutulumu, tümör boyut ve derinliği ve paraneoplastik sendroma sebep olması gibi birçok faktöre bağlı olarak değişebilmektedir. Bazı tümörler ise asemptomatik seyir gösterebilir (5-7).

KLİNİK SEMPTOMATOLOJİ

Periton, primitif embriyoda vücut boşluklarını kaplayan mezodermden köken alır. Abdominal vücut boşluğununda "gut tube" oluşur ve karın ön duvarını kaplayan paryetal yaprak ile organların üzerini saran visseral tabakanın oluşumu tamamlanır (8). Parietal periton ile karın duvarı arasındaki boşluğa retroperitoneum denir ve bu bölgedeki organlara retroperitoneal organlar; periton organı tamamen sarıyorsa bu organlar da intraperitoneal organlar olarak tanımlanır.

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KAYNAKÇA

1. Grimer R, Judson I, Peake D et al . Guidelines for the management of soft tissue sarcomas. Sarcoma. 2010;2010:506182. doi:10.1155/2010/506182.
2. Dinçbaş FÖ. Yumuşak doku tümörlerinin tedavisine güncel yaklaşım, Yumuşak Doku Tümörleri Ek Sayısı , Türk Onkoloji Dergisi .2015;30
3. Suit HD, Spiro I. Role of radiation in the management of adult patients with sarcoma of soft tissue. Semin Surg Oncol . 1994;10(5):347-56.
4. Rosenberg SA, Tepper J, Glatstein E et al. The treatment of soft-tissue sarcomas of the extremities: prospective randomized evaluations of (1) limb sparing surgery plus radiation therapy compared with amputation and (2) the role of adjuvant chemotherapy. Ann Surg. 1982;196(3):305-15.
5. Borden EC, Baker LH, Bell RS et al. Soft tissue sarcomas of adults: state of the translational science. Clin Cancer Res. 2003;9:1941-1956.
6. Clark MA, Fisher C, Judson I et al . Soft-tissue sarcomas in adults. N Engl J Med. 2005;353:701-711.
7. Dileo P, Demetri GD. Update on new diagnostic and therapeutic approaches for sarcomas. Clin Adv Hematol Oncol. 2005;3:781-791.
8. Ellis H. Clinical Anatomy. 9th ed. London:Blackwell ScienceLtd;1997.
9. Burkitt GJC, Healy JC. Anatomy of retroperitoneum, Imaging. 2000, 12(1):10–20.
10. Sanyal R, Remer EM, Radiology of the retroperitoneum: case-based review, AJR Am J Roentgenol. 2009, 192(6 Suppl):S112–S117 (Quiz S118–S121).
11. Weiss SW, Goldblum JR. Weiss's Soft Tissue Tumors. St. Louis, MO: Mosby; 2001.
12. Brennan M, Alektiar KM, Maki R. Sarcomas of soft tissue and bone: soft tissue sarcoma, in Cancer: Principles and Practice of Oncology. Philadelphia, PA: Williams and Wilkins; 2001:1841-1891.
13. Skubitz KM, Skubitz AP. Differential gene expression in leiomyosarcoma. Cancer. 2003;98:1029-1038.
14. Skubitz KM, Skubitz AP. Characterization of sarcomas by means of gene expression. J Lab Clin Med. 2004;144:78-91.
15. Segal NH, Pavlidis P, Antonescu CR et al. Classification and subtype prediction of adult soft tissue sarcoma by functional genomics. Am J Pathol. 2003;163:691-700.
16. Nielsen TO, West RB, Linn SC et al. Molecular characterisation of soft tissue tumours: a gene expression study. Lancet. 2002;359:1301-1307.
17. West RB, van de Rijn M. The role of microarray technologies in the study of soft tissue tumours. Histopathology. 2006;48:22-31.
18. Baird K, Davis S, Antonescu CR et al. Gene expression profiling of human sarcomas: insights into sarcoma biology. Cancer Res. 2005;65:92269235.
19. Pisters PW, Leung DH, Woodruff J et al. Analysis of prognostic factors in 1,041 patients with localized soft tissue sarcomas of the extremities. J Clin Oncol. 1996;14:1679-1689.
20. Jemal A, Siegel R, Ward E et al . Cancer statistics,2007. CA Cancer J Clin. 2007;57(1):43–66.
21. Rasmussen SA, Friedman JM. NF1 gene and neurofibromatosis 1. Am J Epidemiol. 2000;151(1):33–40.
22. Uusitalo E, Rantanen M, Kallionpää RA et al. Distinctive Cancer Associations in patients with neurofibromatosis type 1. J Clin Oncol. 2016;34(17):1978–86
23. Chen CS, Suthers G, Carroll J et al . Sarcoma and familial retinoblastoma. Clin Exp Ophthal. 2003;31(5):392–6.
24. Bell DW, Varley JM, Szydlo TE et al. Heterozygous germ line hCHK2 mutations in Li-Fraumeni syndrome. Science. 1999;286(5449):2528–31.
25. Albritton K, Bleyer WA. The management of cancer in the older adolescent. Eur J Cancer. 2003;39(18):2584–99.
26. Birch JM, Alston RD, Quinn M et al. Incidence of malignant disease by morphological type, in young persons aged 12–24 years in England, 1979–1997. Eur J Cancer. 2003;39(18):2622–31.

27. Geraci M, Birch JM, Alston RD et al. Cancer mortality in 13 to 29-year-olds in England and Wales, 1981–2005. *Br J Cancer*. 2007;97(11):1588–94.
28. Johnson CJD, Pynsent PB, Grimer RJ. Clinical features of soft tissue sarcomas. *Ann R Coll Surg Engl*. 2001;83(3):203–5.
29. McGinn CJ, Lawrence TS. Soft tissue sarcomas (excluding retroperitoneum). In: Perez CA, Brady LW, editors. *Principles and practice of radiation oncology*. 3rd ed. Philadelphia: Lippincott-Raven Publishers; 1998. p. 2051–72.
30. de Saint Aubain Somerhausen N, Fletcher CD. Soft-tissue sarcomas: an update. *Eur J Surg Oncol* 1999;25:215–220.
31. Baş A, Gülsen F, Kantarci F. Yumuşak doku sarkomlarında görüntü eşliğinde biyopsi. *Türk Onkoloji Dergisi*. 2015;30(Ek 1):29–31
32. Cohan RH, Baker ME, Cooper C et al. Computed tomography of primary retroperitoneal malignancies *J Comput Assist Tomogr*. 1988; 12: 804-810.
33. Neville A, Herts BR. CT characteristics of primary retroperitoneal neoplasms. *Crit Rev Comput Tomogr*. 2004; 45: 247-270.
34. Mullinax JE, Zager JS, Gonzalez RJ. Current diagnosis and management of retroperitoneal sarcoma. *Cancer Control*. 2011; 18: 177–87.
35. Lehnert T, Cardona S, Hinz U et al. Primary and locally recurrent retroperitoneal soft-tissue sarcoma: local control and survival. *Eur J Surg Oncol*. 2009; 35: 986–93.
36. Yeh JJ, Singer S, Brennang MF et al., Effectiveness of palliative procedures for intra-abdominal sarcomas. *Ann Surg Oncol*. 2005; 12:1084–1089.
37. Jaime P, Nomonde M. Uterine sarcomas. *Int J Gynecol Obstet [Internet]*. 2015 Sep 30; 131(S2): S105–10. Available from: <https://doi.org/10.1016/j.ijgo.2015.06.006>
38. Siegel RL, Miller KD, Jemal A. Cancer statistics 2016. *CA Cancer J Clin* . 2016;66(1):7–30.
39. Cormier JN, Pollock RE. Soft tissue sarcomas. *CA Cancer J Clin* . 2004;54(2):94–109.
40. Levy AD, Manning MA, Al-Refaie WB et al . Soft-tissue sarcomas of the abdomen and pelvis: radiologicpathologic features, part 1—common sarcomas. *RadioGraphics* . 2017;37(2):462–483.
41. Goldblum JR, Folpe AL, Weiss SW. Undifferentiated pleomorphic sarcoma. In: Goldblum JR, Folpe AL, Weiss SW, eds. *Enzinger and Weiss's soft tissue tumors*. 6th ed. Philadelphia, Pa: Saunders/Elsevier. 2013; 421–442.
42. Angela DL , Maria AM , Markku MM . Soft-Tissue Sarcomas of the Abdomen and Pelvis: RadiologicPathologic Features, Part 2— Uncommon Sarcomas. *RadioGraphics*. 2017; 37:797–812. 10.1148/rg.2017160201
43. Strickland L, Letson GD, Muro-Cacho CA. Gastrointestinal stromal tumors. *Cancer Control* . 2001; 8:252–261.
44. Sturgeon C, Chejfec G, Espat N. Gastrointestinal stromal tumors: a spectrum of diseases. *Surg Oncol*. 2003; 12:21–6.
45. Vijay A , Ram L. Retroperitoneal liposarcoma: A comprehensive review. *Am J Clin Oncol*.2015-38: 213-219.
46. Staguchi S, Kume H , Fukuhara H et al. Symptoms at diagnosis as independent prognostic factors in retroperitoneal liposarcoma. *Molecular and Clinical Oncology* . 2016- 4: 255-260.
47. Lazar A, Evans HL, Shipley J. Leiomyosarcoma. In: Fletcher CDM, Bridge JA, Hogendoorn PCW, Mertens F, eds. *WHO classification of tumours of soft tissue and bone*. 4th ed. Lyon, France: IARC. 2013; 111–113.
48. Hartman DS, Hayes WS, Choyke PL et al . Leiomyosarcoma of the retroperitoneum and inferior vena cava: radiologic-pathologic correlation. *RadioGraphics*.1992;12 (6):1203–1220.
49. Howe JR, Karnell LH, Scott-Conner C . Small bowel sarcoma: Analysis of survival from the National Cancer Data Base. *Ann Surg Oncol*. 2001;8:496–508.
50. Plaat BE, Hollema H, Molenaar WM et al. Soft tissue leiomyosarcomas and malignant gastrointestinal stromal tumors: Differences in clinical outcome and expression of multidrug resistance proteins. *J Clin Oncol*. 2000;18:3211–3220.

51. Agaimy A, Wunsch P. True smooth muscle neoplasms of the gastrointestinal tract: morphological spectrum and classification in a series of 85 cases from a single institute. *Langenbecks Arch Surg.* 2007;392:75-81.
52. Miettinen M, Sarlomo-Rikala M, Sabin LH et al. Gastrointestinal stromal tumors and leiomyosarcomas in the colon. A clinicopathologic, immunohistochemical, and molecular genetic study of 44 cases. *Am J Surg Pathol.* 2000;24:1339-52.
53. Rajani B, Smith TA, Reith JD et al . Retroperitoneal leiomyosarcomas unassociated with the gastrointestinal tract: a clinicopathologic analysis of 17 cases. *Mod Pathol.* 1999;12(1):21-28.
54. Kempson RL, Fletcher CDM , Evans HL et al . *Atlas of tumor pathology: tumors of the soft tissues.* Washington, DC: Armed Forces Institute of Pathology, 2001.
55. Lane RH, Stephens DH, Reiman HM. Primary retroperitoneal neoplasms: CT findings in 90 cases with clinical and pathologic correlation. *AJR Am J Roentgenol.* 1989;152(1):83-89
56. Cooley CL, Jagannathan JP, Kurra V et al. Imaging features and metastatic pattern of non-IVC retroperitoneal leiomyosarcomas: are they different from IVC leiomyosarcomas? *J Comput Assist Tomogr.* 2014;38(5):687-692.
57. Suster S. Gastrointestinal stromal tumors. *Semin Diagn Pathol.* 1996;13:297-313
58. Mazur MT, Clark HB. Gastric stromal tumors. Reappraisal of histogenesis. *Am J Surg Pathol.* 1983;7:507-519.
59. Kindblom LG, Remotti HE, Aldenborg F et al. Gastrointestinal pacemaker cell tumor (GIPAC-CT): Gastrointestinal stromal tumors show phenotypic characteristics of the interstitial cells of Cajal. *Am J Pathol* 1998;152:1259-1269.
60. Sanders KM. A case for interstitial cells of Cajal as pacemakers and mediators of neurotransmission in the gastrointestinal tract. *Gastroenterology.* 1996;111:492-515.
61. Prakash S, Sarran L, Socci N et al. Gastrointestinal stromal tumors in children and young adults: A clinicopathologic, molecular, and genomic study of 15 cases and review of the literature. *J Pediatr Hematol Oncol.* 2005;27:179-187.
62. Miettinen M, Sarlomo-Rikala M, Sabin LH, et al.: Esophageal stromal tumors: A clinicopathologic, immunohistochemical, and molecular genetic study of 17 cases and comparison with esophageal leiomyomas and leiomyosarcomas. *Am J Surg Pathol.* 2000;24:211-222.
63. DeMatteo RP, Lewis JJ, Leung D, et al. Two hundred gastrointestinal stromal tumors: Recurrence patterns and prognostic factors for survival. *Ann Surg* 2000; 231:51-58
64. Katz SC, Ronald PD . *Gastrointestinal Stromal Tumors and Leiomyosarcoma.* Journal of Surgical Oncology. 2008;97:350-35
65. Nilsson B, Bunning P, Meis-Kindblom JM et al. Gastrointestinal stromal tumors: The incidence, prevalence, clinical course, and prognostication in the preimatinib mesylate era—A population-based study in Western Sweden. *Cancer.* 2005;103: 821-829.
66. Dematteo RP, Maki RG, Antonescu C et al. Targeted molecular therapy for cancer: The application of ST1571 to gastrointestinal stromal tumor. *Curr Probl Surg.* 2003;40:144- 193.
67. Ng EH, Pollock RE, Munsell MF et al. Prognostic factors influencing survival in gastrointestinal leiomyosarcomas. Implications for surgical management and staging. *Ann Surg.* 1992; 215:68-77.
68. Antonescu CR, Besmer P, Guo T et al. Acquired resistance to imatinib in gastrointestinal stromal tumor occurs through secondary gene mutation. *Clin Cancer Res.* 2005;11:4182- 4190.
69. Debiec-Rychter M, Cools J, Dumez H et al. Mechanisms of resistance to imatinib mesylate in gastrointestinal stromal tumors and activity of the PKC412 inhibitor against imatinib-resistant mutants. *Gastroenterology.* 2005;128:270-279.
70. Prenen H ,Cools J, Mentens N et al. Efficacy of the kinase inhibitor SU11248 against gastrointestinal stromal tumor mutants refractory to imatinib mesylate. *Clin Cancer Res.* 2006;12:2622-2627.