

BÖLÜM 1

YUMUŞAK DOKU TÜMÖRLERİ İLE İLGİLİ GENEL BİLGİLER

Orhan BALTA ¹

GİRİŞ

Yumuşak doku tümörleri geniş bir biyolojik davranış yelpazesine sahip mezenkimal kökenli tümörlerdir(1-7). Yumuşak doku tümörleri fibröz bağ dokudan, yağ dokusundan, iskelet kaslarından, kan ve lenf damarlarından veya periferik sinir sistemini içeren vücudun kemik dışındaki, epitelial olmayan mezanşimal doku proliferasyonudur (Tablo 1)(8). Genelde iyi huylu (bening) yumuşak doku tümörleri kötü huylu (maling) olanlardan daha sıktır. Yumuşak doku sarkomları nispeten nadir olmasına rağmen, tanınasal olarak tanı koymayı zorlayıcı bir

çok faktörü içeren bir neoplazma grubudur(7). Doğru tanı, uygun tedavi bu hastalığın prognozu için önemlidir(7).

Nadir olarak görülseden 100'den fazla heterojen histolojik alt tipi olan, mezenkimal kökenli bir tümör grubu olarak karşılaştığımız yumuşak doku (7) neoplazmı, tümörün en çok benzediği olası menşee dokusuna göre sınıflanır (Tablo 1)(9,10). Genelde benign yumuşak doku tümörleri komşu dokulara invazyon yapmazlar ve metastaz potansiyelleri yoktur ve ölüme sebebiyet vermezler. İyi huylu yumuşak

Tablo 1. Sarkom türlerinin benzer olduğu dokular.

Sarkom Türü	Benzer olduğu dokular
Fibrosarkom (doğuştan olanlar hariç)	Fibröz doku
Leiomyosarkom	Düz kas
Farklılaşmamış pleomorfik sarkom	Kötü farklılaşmış
Osteosarkom	Kemik
Kondrosarkom (iskelet dışı miksoid dışındaki tipler)	Kıkırdak
Liposarkom (miksoid dışındaki türler)	Yağ
Embriyonal rabdomiyosarkom	İskelet kası
Malign periferik sinir kılıfı tümörü	Sinir kılıfı kaynaklı
Anjiyosarkom	Endotel
Alveolar soft part sarkomu, Epiteloid sarkom, Berrak hücreli sarkom	Histogenez belirsizdir

¹ Doç. Dr., Tokat Gaziosmanpaşa Üniversitesi Tıp Fakültesi Ortopedi ve Travmatoloji AD., drorhanbalta@hotmail.com

teknikleri kullanılarak tedavi edilirler. Tedavi algoritması her zaman tıbbi öykü, fizik muayene, radyolojik incelemeler, sistemik tarama ve histopatolojik tanı modelini izlemelidir.

KAYNAKLAR

- Wilke BK, Goulding KA, Sherman CE, Houdek MT. Soft Tissue Tumors: Diagnosis, Treatment, and Follow-up from the Orthopedic Oncologist Perspective. *Radiologic Clinics*. 2022;60(2):253-62.
- Bancroft LW, Peterson JJ, Kransdorf MJ, Nomikos GC, Murphey MD. Soft tissue tumors of the lower extremities. *Radiologic Clinics*. 2002;40(5):991-1011.
- Morrison BA, editor Soft tissue sarcomas of the extremities. Baylor University Medical Center Proceedings; 2003: Taylor & Francis.
- Ryan CW, Meyer J, Maki R, Pollock RE, Savarese DM. Clinical presentation, histopathology, diagnostic evaluation, and staging of soft tissue sarcoma. UpToDate <http://www.uptodate.com/contents/clinical-presentation-histopathology-diagnostic-evaluation-and-staging-of-soft-tissue-sarcoma> (accessed 14 March 2016). 2012.
- Ardakani AHG, Woollard A, Ware H, Gikas P. Soft tissue sarcoma: Recognizing a rare disease. *Cleveland Clinic Journal of Medicine*. 2022;89(2):73-80.
- Habibollahi S, Lozano-Calderon S, Chang CY. Common Soft Tissue Mass-like Lesions that Mimic Malignancy. *Radiologic Clinics*. 2022;60(2):301-10.
- Choi JH, Ro JY. The 2020 WHO classification of tumors of soft tissue: selected changes and new entities. *Advances in Anatomic Pathology*. 2021;28(1):44-58.
- Norris JP. Soft Tissue Tumors. *Clinical Foundations of Musculoskeletal Medicine*: Springer; 2021. p. 209-26.
- Weiss SW, Goldblum JR, Folpe AL. Enzinger and Weiss's soft tissue tumors: Elsevier Health Sciences; 2007.
- Honoré C, Mééus P, Stoeckle E, Bonvalot S. Soft tissue sarcoma in France in 2015: epidemiology, classification and organization of clinical care. *Journal of visceral surgery*. 2015;152(4):223-30.
- Frassica FJ, Thompson Jr RC. Instructional Course Lectures, The American Academy of Orthopaedic Surgeons-Evaluation, Diagnosis, and Classification of Benign Soft-Tissue Tumors. *JBJS*. 1996;78(1):126-40.
- Goldblum JR, Weiss SW, Folpe AL. Enzinger and Weiss's soft tissue tumors E-book: Elsevier Health Sciences; 2013.
- Mankin HJ, Hornicek FJ. Diagnosis, classification, and management of soft tissue sarcomas. *Cancer control*. 2005;12(1):5-21.
- Wibmer C, Leithner A, Zielonke N, Sperl M, Windhager R. Increasing incidence rates of soft tissue sarcomas? A population-based epidemiologic study and literature review. *Annals of Oncology*. 2010;21(5):1106-11.
- Mastrangelo G, Coindre JM, Ducimetière F, Dei Tos AP, Fadda E, Blay JY, et al. Incidence of soft tissue sarcoma and beyond: a population-based prospective study in 3 European regions. *Cancer*. 2012;118(21):5339-48.
- Xia C, Dong X, Li H, Cao M, Sun D, He S, et al. Cancer statistics in China and United States, 2022: profiles, trends, and determinants. *Chinese medical journal*. 2022;135(05):584-90.
- Ward E, DeSantis C, Robbins A, Kohler B, Jemal A. Childhood and adolescent cancer statistics, 2014. *CA: a cancer journal for clinicians*. 2014;64(2):83-103.
- Miller KD, Fidler-Benaoudia M, Keegan TH, Hipp HS, Jemal A, Siegel RL. Cancer statistics for adolescents and young adults, 2020. *CA: a cancer journal for clinicians*. 2020;70(6):443-59.
- Cormier JN, Pollock RE. Soft tissue sarcomas. *CA: a cancer journal for clinicians*. 2004;54(2):94-109.
- Ogura K, Higashi T, Kawai A. Statistics of soft-tissue sarcoma in Japan: report from the bone and soft tissue tumor registry in Japan. *Journal of Orthopaedic Science*. 2017;22(4):755-64.
- Gilbert NF, Cannon CP, Lin PP, Lewis VO. Soft-tissue sarcoma. *JAAOS-Journal of the American Academy of Orthopaedic Surgeons*. 2009;17(1):40-7.
- Ahmed AA, Tsokos M. Sinonasal rhabdomyosarcoma in children and young adults. *International Journal of Surgical Pathology*. 2007;15(2).
- Herzog CE. Overview of sarcomas in the adolescent and young adult population. *Journal of Pediatric Hematology/Oncology*. 2005;27(4):215-8.
- Kunisada T, Nakata E, Fujiwara T, Hosono A, Takihira S, Kondo H, et al. Soft-tissue sarcoma in adolescents and young adults. *International Journal of Clinical Oncology*. 2022;1-11.
- van der Graaf WT, Orbach D, Judson IR, Ferrari A. Soft tissue sarcomas in adolescents and young adults: a comparison with their paediatric and adult counterparts. *The Lancet Oncology*. 2017;18(3):e166-e75.
- Huvos AG, Heilweil M, Bretsky SS. The pathology of malignant fibrous histiocytoma of bone. A study of 130 patients. *The American journal of surgical pathology*. 1985;9(12):853-71.
- Bancroft LW, Kransdorf MJ, Peterson JJ, O'Connor MI. Benign fatty tumors: classification, clinical course, imaging appearance, and treatment. *Skeletal radiology*. 2006;35(10):719-33.
- Walker EA, Fenton ME, Salesky JS, Murphey MD. Magnetic resonance imaging of benign soft tissue neoplasms in adults. *Radiologic Clinics*. 2011;49(6):1197-217.
- Fröhner M, Wirth MP. Etiologic factors in soft tissue sarcomas. *Oncology Research and Treatment*. 2001;24(2):139-42.
- Zahm SH, Fraumeni J, editors. The epidemiology of soft tissue sarcoma. *Seminars in oncology*; 1997: Wb Saunders Co.
- Boland P. Bone and Soft Tissue Sarcomas. *Cancer*: CRC Press; 1998. p. 240-51.
- Delaney TF, Harmon DC, YOON S, Kirsch DG, Rosenberg AE, Mankin HJ, et al. Soft tissue sarcomas. *Head and neck*. 2015;12(15):8.

33. Brennan M, Lewis J. Diagnosis and management of soft tissue sarcoma: CRC Press; 2002.
34. Bansal A, Goyal S, Goyal A, Jana M. WHO classification of soft tissue tumours 2020: An update and simplified approach for radiologists. *European Journal of Radiology*. 2021;143:109937.
35. Glass AG, Fraumeni Jr JF. Epidemiology of bone cancer in children. *Journal of the National Cancer Institute*. 1970;44(1):187-99.
36. Andersson HC, Parry DM, Mulvihill JJ. Lymphangiosarcoma in late-onset hereditary lymphedema: Case report and nosological implications. *American journal of medical genetics*. 1995;56(1):72-5.
37. Roat JW, Wald A, Mendelow H, Pataki KI. Hepatic angiosarcoma associated with short-term arsenic ingestion. *The American Journal of Medicine*. 1982;73(6):933-6.
38. Restrepo CS, Martínez S, Lemos JA, Carrillo JA, Lemos DF, Ojeda P, et al. Imaging manifestations of Kaposi sarcoma. *Radiographics*. 2006;26(4):1169-85.
39. Kuhn KJ, Cloutier JM, Boutin RD, Steffner R, Riley G. Soft tissue pathology for the radiologist: a tumor board primer with 2020 WHO classification update. *Skeletal Radiology*. 2021;50(1):29-42.
40. Clark MA, Fisher C, Judson I, Thomas JM. Soft-tissue sarcomas in adults. *New England journal of medicine*. 2005;353(7):701-11.
41. Rimondi E, Benassi MS, Bazzocchi A, Balladelli A, Facchini G, Rossi G, et al. Translational research in diagnosis and management of soft tissue tumours. *Cancer Imaging*. 2016;16(1):1-13.
42. Stramare R, Beltrame V, Gazzola M, Gerardi M, Scattolin G, Coran A, et al. Imaging of soft-tissue tumors. *Journal of magnetic resonance imaging*. 2013;37(4):791-804.
43. Gómez MPA, Errani C, Lalam R, Nikodinovska VV, Fanti S, Tagliafico AS, et al., editors. The role of ultrasound in the diagnosis of soft tissue tumors. *Seminars in Musculoskeletal Radiology*; 2020: Thieme Medical Publishers.
44. Aisen AM, Martel W, Braunstein EM, McMillin KI, Phillips WA, Kling T. MRI and CT evaluation of primary bone and soft-tissue tumors. *American journal of Roentgenology*. 1986;146(4):749-56.
45. Enneking WF, Spanier SS, Malawer MM. The effect of the anatomic setting on the results of surgical procedures for soft parts sarcoma of the thigh. *Cancer*. 1981;47(5):1005-22.
46. Shmookler B, Bickels J, Jelinek J, Sugarbaker P, Malawer M. Bone and soft-tissue sarcomas: epidemiology, radiology, pathology and fundamentals of surgical treatment. *Musculoskeletal cancer surgery*. 2004:3-35.
47. Malawer MM, Sugarbaker PH. *Musculoskeletal cancer surgery: treatment of sarcomas and allied diseases*: Springer Science & Business Media; 2001.
48. Brouns F, Stas M, De Wever I. Delay in diagnosis of soft tissue sarcomas. *European Journal of Surgical Oncology (EJSO)*. 2003;29(5):440-5.
49. Skrzynski MC, Biermann JS, Montag A, Simon MA. Diagnostic accuracy and charge-savings of outpatient core needle biopsy compared with open biopsy of musculoskeletal tumors. *JBJS*. 1996;78(5):644-9.
50. Hoeber I, Spillane AJ, Fisher C, Thomas JM. Accuracy of biopsy techniques for limb and limb girdle soft tissue tumors. *Annals of surgical oncology*. 2001;8(1):80-7.
51. Kilpatrick SE, Ward WG, Cappellari JO, Bos GD. Fine-needle aspiration biopsy of soft tissue sarcomas: a cytomorphologic analysis with emphasis on histologic subtyping, grading, and therapeutic significance. *American journal of clinical pathology*. 1999;112(2):179-88.
52. De Schepper A, De Beuckeleer L, Vandevenne J, Somville J. Magnetic resonance imaging of soft tissue tumors. *European radiology*. 2000;10(2):213-23.
53. Miwa S, Otsuka T. Practical use of imaging technique for management of bone and soft tissue tumors. *Journal of Orthopaedic Science*. 2017;22(3):391-400.
54. Bar-Shalom R, Yefremov N, Guralnik L, Gaitini D, Frenkel A, Kuten A, et al. Clinical performance of PET/CT in evaluation of cancer: additional value for diagnostic imaging and patient management. *Journal of nuclear medicine*. 2003;44(8):1200-9.
55. Jacob LA, Sreevalli A, Ninutha S, Dasappa L, Babu S, Lokesh K, et al. Soft Tissue Sarcomas with Special Reference to Molecular Aberration, Chemotherapy, and Recent Advances: A Review Article. *Indian Journal of Medical and Paediatric Oncology*. 2022.
56. Blazer III DG, Sabel MS, Sondak VK. Is there a role for sentinel lymph node biopsy in the management of sarcoma? *Surgical Oncology*. 2003;12(3):201-6.
57. Behranwala KA, A'Hern R, Omar A-M, Thomas JM. Prognosis of lymph node metastasis in soft tissue sarcoma. *Annals of surgical oncology*. 2004;11(7):714-9.
58. Siegel HJ, Dunham WH, Lopez-Ben R, Siegal GP. Intracranial metastasis from synovial sarcoma. *Orthopedics (Online)*. 2008;31(4):405.
59. Thompson MJ, Ross J, Domson G, Foster W. Screening and surveillance CT abdomen/pelvis for metastases in patients with soft-tissue sarcoma of the extremity. *Bone & Joint Research*. 2015;4(3):45-9.
60. Jones NB, Iwenofu H, Scharschmidt T, Kraybill W. Prognostic factors and staging for soft tissue sarcomas: an update. *Surgical oncology clinics of North America*. 2012;21(2):187-200.
61. Ezuddin NS, Pretell-Mazzini J, Yechieli RL, Kerr DA, Wilky BA, Subhawong TK. Local recurrence of soft-tissue sarcoma: issues in imaging surveillance strategy. *Skeletal radiology*. 2018;47(12):1595-606.
62. Hajdu SI, Shiu MH, Brennan MF. The role of the pathologist in the management of soft tissue sarcomas. *World journal of surgery*. 1988;12(3):326-31.
63. Italiano A, Di Mauro I, Rapp J, Pierron G, Auger N, Alberti L, et al. Clinical effect of molecular methods in sarcoma diagnosis (GENSARC): a prospective, multicentre, observational study. *The Lancet Oncology*. 2016;17(4):532-8.
64. Toro JR, Travis LB, Wu HJ, Zhu K, Fletcher CD, Devessa SS. Incidence patterns of soft tissue sarcomas,

- regardless of primary site, in the surveillance, epidemiology and end results program, 1978–2001: an analysis of 26,758 cases. *International journal of cancer*. 2006;119(12):2922–30.
65. Hemmings C, Miles C, Slavin J, Bonar SF, Graf N, Austen L, et al. Optimising the management of soft tissue tumours. *Pathology*. 2011;43(4):295–301.
 66. Reynoso D, Subbiah V, Trent JC, Guadagnolo BA, Lazar AJ, Benjamin R, et al. Neoadjuvant treatment of soft-tissue sarcoma: A multimodality approach. *Journal of surgical oncology*. 2010;101(4):327–33.
 67. Rektman N, Baine MK, Bishop JA. Grading (and Classification) Systems Quick Reference: Solid Tumors. *Quick Reference Handbook for Surgical Pathologists*; Springer; 2019. p. 93–111.
 68. Wei S, Henderson-Jackson E, Qian X, Bui MM. Soft tissue tumor immunohistochemistry update: illustrative examples of diagnostic pearls to avoid pitfalls. *Archives of pathology & laboratory medicine*. 2017;141(8):1072–91.
 69. Frassica FJ, Khanna JA, McCarthy EF. The role of MR imaging in soft tissue tumor evaluation: perspective of the orthopedic oncologist and musculoskeletal pathologist. *Magnetic resonance imaging clinics of North America*. 2000;8(4):915–27.
 70. Nystrom LM, Reimer NB, Reith JD, Dang L, Zlotecki RA, Scarborough MT, et al. Multidisciplinary management of soft tissue sarcoma. *The Scientific World Journal*. 2013;2013.
 71. Beaman FD, Kransdorf MJ, Andrews TR, Murphey MD, Arcara LK, Keeling JH. Superficial soft-tissue masses: analysis, diagnosis, and differential considerations. *Radiographics*. 2007;27(2):509–23.
 72. Dogan ÖY, Oksuz DÇ, Atalar B, Dincbas FO. Long-term results of extremity soft tissue sarcomas limb-sparing surgery and radiotherapy. *Acta Ortopédica Brasileira*. 2019;27:207–11.
 73. Mischianu D, Iorga L, Marcu D, Ciuca A, Bratu I, Costache RS. Renal involvement in primary retroperitoneal tumors. *RJMM*. 122(1):27.
 74. Kotilingam D, Lev DC, Lazar AJ, Pollock RE. Staging soft tissue sarcoma: evolution and change. *CA: a cancer journal for clinicians*. 2006;56(5):282–91.
 75. Hunt KK, Patel SR, Pollack A. Soft tissue sarcomas. *BASIC AND CLINICAL ONCOLOGY*. 1999;20:394–415.
 76. Woo KS, Kim SH, Kim HS, Cho PD. Clinical experience with treatment of angioleiomyoma. *Archives of plastic surgery*. 2014;41(04):374–8.
 77. Van Glabbeke M, Van Oosterom A, Oosterhuis J, Mouridsen H, Crowther D, Somers R, et al. Prognostic factors for the outcome of chemotherapy in advanced soft tissue sarcoma: an analysis of 2,185 patients treated with anthracycline-containing first-line regimens—a European Organization for Research and Treatment of Cancer Soft Tissue and Bone Sarcoma Group Study. *Journal of Clinical Oncology*. 1999;17:150–7.
 78. Levine EA, editor *Prognostic factors in soft tissue sarcoma. Seminars in surgical oncology*; 1999: Wiley Online Library.
 79. Maretty-Nielsen K, Aggerholm-Pedersen N, Safwat A, Jørgensen PH, Hansen BH, Baerentzen S, et al. Prognostic factors for local recurrence and mortality in adult soft tissue sarcoma of the extremities and trunk wall: a cohort study of 922 consecutive patients. *Acta orthopaedica*. 2014;85(3):323–32.
 80. Gutierrez JC, Perez EA, Franceschi D, Moffat Jr FL, Livingstone AS, Koniaris LG. Outcomes for soft-tissue sarcoma in 8249 cases from a large state cancer registry. *Journal of surgical research*. 2007;141(1):105–14.
 81. Lahat G, Tuvin D, Wei C, Anaya D, Bekele B, Lazar A, et al. New perspectives for staging and prognosis in soft tissue sarcoma. *Annals of surgical oncology*. 2008;15(10):2739–48.
 82. Abbas JS, Holyoke ED, Moore R, Karakousis CP. The surgical treatment and outcome of soft-tissue sarcoma. *Archives of Surgery*. 1981;116(6):765–9.
 83. Markhede G, Angervall L, Stener B. A multivariate analysis of the prognosis after surgical treatment of malignant soft-tissue tumors. *Cancer*. 1982;49(8):1721–33.
 84. Gutierrez JC, Perez EA, Moffat FL, Livingstone AS, Franceschi D, Koniaris LG. Should soft tissue sarcomas be treated at high-volume centers?: An analysis of 4205 patients. *Annals of surgery*. 2007;245(6):952.
 85. Zagars GK, Ballo MT, Pisters PW, Pollock RE, Patel SR, Benjamin RS, et al. Prognostic factors for patients with localized soft-tissue sarcoma treated with conservation surgery and radiation therapy: an analysis of 1225 patients. *Cancer: Interdisciplinary International Journal of the American Cancer Society*. 2003;97(10):2530–43.
 86. Miwa S, Yamamoto N, Tsuchiya H. Bone and soft tissue tumors: new treatment approaches. *MDPI*; 2021. p. 1832.
 87. Rosenberg SA, Tepper J, Glatstein E, Costa J, Baker A, Brennan M, 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. *Annals of surgery*. 1982;196(3):305.
 88. Yang JC, Chang AE, Baker AR, Sindelar WF, Danforth DN, Topalian SL, et al. Randomized prospective study of the benefit of adjuvant radiation therapy in the treatment of soft tissue sarcomas of the extremity. *Journal of clinical oncology*. 1998;16(1):197–203.