Bölüm
10

DESMOİD TÜMÖRLERDE CERRAHİ TEDAVİ

Haydar ÖCAL[12]

ETYOLOJİ-EPİDEMİYOLOJİ

Diğer adı ‘’Agresif fibromatözis” olan ‘’Desmoid tümörler” fasya, tendonlar ve skar dokusundan köken alan lokal invaziv karakterde tümörlerdir (1). Desmoid tümörler Dünya Sağlık Örgütü’ne göre ‘’infiltratif büyüyen, lokal rekürrens eğiliminde olan ve derin yumuşak dokudan köken alan klonal fibroblastik proliferasyonlar” olarak tanımlanır. Desmoid tümörlerin kapsülü yoktur ve derin doku infiltrasyonu yaparlar. Bu tümörler eksizyon marjinleri pozitif olduğunda lokal rekürrens eğilimlerinin yüksek olması nedeniyle sıklıkla ‘’düşük dereceli sarkomlar” olarak kategorize edildir. Nadiren metastaz yapabilirler. Gebelerde karın duvarında görülen kitlelerden intraabdominal mezenterik kitlelere ve yaşlı hastalardaki geniş ekstremite kitlelerine kadar lokalizasyonu ve neden olduğu klinik tablo çok değişken olabilen bir tümördür.

Desmoid tümörlerin (tüm yumuşak doku tümörlerinin %3’ü kadardır) görülme sıklığı %0.03’tür (2). İki tip desmoid tümör tanımlanmıştır: ‘’Sporadik” ve ‘’FAP ile ilişkili” desmoid tümörler. Kadın ve erkekte eşit görülmekle birlikte genç yaş grubunda kadınlarda daha sık görülür. İnsidansı 3. ve 4. dekatta pik yapar (3). Gövde, ekstremiteler ve abdominal kavite desmoid tümörün en sık görüldüğü lokalizasyonlardır. %5 ‘i intraabdominal yerleşimliyken bu oran ‘’FAP ilişkili” desmoidlerde %80 ‘dir (4). FAP ilişkili desmoidlerde cerrahi sonrası rekürrens oranı yüksektir.

Sıklıkla endokrin faktörler (Genç kadın hastalarda östrojen etkisi), FAP, travma ile ilişkili olmasına rağmen etyolojisi net olarak ortaya konamamıştır (5). FAP’lı hastalarda normal popülasyona göre daha sık görülür. FAP vakalarınn %7.5-16’ sında desmoid tümör görülür (6,7,8,9).

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KAYNAKÇA

1. Burke AP, Sobin LH, Shekitka KM, et al. Intra-abdominal fibromatosis. A pathologic analysis of 130 tumours with comparison of clinical subgroups. Am J Surg Pathol. 1990;14(4):335–41.
2. Merchant NB, Lewis JJ, Woodruff JM et al: Extremity and trunk desmoid tumors: a multifactorial analysis of outcome. Cancer 1999;86:2045-2052.
3. Plukker JT, van Oort I, Vermey A, et al. Aggressive fibromatosis (non-familial desmoids tumour): therapeutic problems and the role of adjuvant radiotherapy. Br J Surg. 1995;82(4):510–14
4. Kasper B, Ströbel P, Hohenberger P. DTs et al: Clinical Features and Treatment Options for Advanced Disease. Oncologist. 2011 May;16(5):682–693.
5. Reitamo JJ, Häyry P, Nykyri E et al. The DT. I. Incidence, sex-, age- and anatomical distribution in the Finnish population. Am J Clin Pathol. 1982;77(6):665–73.
6. Giardiello FM, Petersen GM, Krush AJ et al. Desmoid tumours in familial adenomatous polyposis. 1994 Mar;35(3):377-81.
7. Fallen T, Wilson M,Morlan B et al. Desmoid tumors -- a characterization of patients seen at Mayo Clinic 1976-1999.Fam cancer. 2006;5(2):191-4
8. Nieuwenhuis MH, Casparie M, Mathus-Vliegen LM et al A nation-wide study comparing sporadic and familial adenomatous polyposis-related desmoid-type fibromatoses.Int J Cancer. 2011 Jul 1;129(1):256-61. doi: 10.1002/ijc.25664. Epub 2010 Nov 3
9. Nieuwenhuis MH, Lefevre JH, Bülow S et al. Family history, surgery, and APC mutation are risk factors for desmoid tumors in familial adenomatous polyposis: an international cohort study. Dis Colon Rectum 2011 Oct;54(10):1229-34. doi: 10.1097/DCR.0b013e318227e4e8).
10. Brueckl WM, Ballhausen WG, Förtsch T,et al. Genetic testing for germline mutations of the APC gene in patients with apparently sporadic desmoid tumors but a family history of colorectal carcinoma. Dis Colon Rectum 2005;48:1275-1281.
11. Bayrakçı B, Özen E, Sever A, Özütemiz Ö. Gardner Sendromu: Olgu sunumu. Akad Gastroent Derg 2007; 6:86-89.
12. ASGE Standards of Practice Committee, Evans JA, Chandrasekhara V, Chathadi KV. The role of endoscopy in the management of premalignant and malignant conditions of the stomach. Gastrointest Endosc. 2015 Jul;82(1):1-8. doi: 10.1016/j.gie.2015.03.1967. Epub 2015 Apr 29 .
13. Schöffski P1, Reichardt P, Blay JY et al. A phase I-II study of everolimus (RAD001) in combination with imatinib in patients with imatinib-resistant gastrointestinal stromal tumors.Ann Oncol. 2010 Oct;21(10):1990-8. doi: 10.1093/annonc/mdq076. Epub 2010 May 27.
14. Mir O, Cropet C, Toulmonde M. PAZOGIST study group of the French Sarcoma Groupe-Groupe d'Etude des Tumeurs Osseuses (GSF-GETO). Pazopanib plus best supportive care versus best supportive care alone in advanced gastrointestinal stromal tumours resistant to imatinib and sunitinib (PAZOGIST): a randomised, multicentre, open-label phase 2 trial. Lancet Oncol. 2016 May;17(5):632-41. doi: 10.1016/S1470-2045(16)00075-9. Epub 2016 Apr 5.
15. Dematteo RP, Gold JS, Saran L et al. Tumor mitotic rate, size, and location independently predict recurrence after resection of primary gastrointestinal stromal tumor (GIST). Cancer. 2008 Feb 1;112(3):608-15.
16. Wang D, Zhang Q, Blanke CD et al. Phase II trial of neoadjuvant/adjuvant imatinib mesylate for advanced primary and metastatic/recurrent operable gastrointestinal stromal tumors: long-term follow-up results of Radiation Therapy Oncology Group 0132. Ann Surg Oncol. 2012 Apr;19(4):1074-80. doi: 10.1245/s10434-011-2190-5. Epub 2011 Dec 28
17. Adalı F, Turan Güner N, Bayramoğlu S. Batın Sağ Yan Duvarında Agresif Fibromatozis: Olgu Sunumu. Van Tıp Dergisi: 2008; 15 (1): 26-29.
18. Demiral G, Yılmaz A, Aksoy F. Abdominal Desmoid Tümör: Olgu Sunumu. Marmara Medical Journal: 2010; 23 (2): 297-301

19. Penel N, Coindre JM, Bonvalot S et al. Management of desmoid tumours: a nationwide survey of labelled reference centre networks in France. Eur J Cancer 2016; 58: 90–96.
20. Huss S, Nehles J, Binot E et al. b-catenin (CTNNB1) mutations and clinicopathological features of mesenteric desmoid-type fibromatosis. Histopathology 2013; 62: 294–304.
21. Mankin HJ, Hornicek FJ, Springfield DS. Extra-abdominal desmoid tumors: a report of 234 cases. J Surg Oncol 2010;102(5):380-4.
22. Rutkowski P, Nowecki Z, Nyckowski P, et al. Surgical treatment of patients with initially inoperable and/or metastatic gastrointestinal stromal tumors (GIST) during therapy with imatinib mesylate. J Surg Oncol 2006;93:304-311.
23. Andtbacka RH,Ng CS, Scaife CL, et al. Surgical resection of gastrointestinal stromal tumors after treatment with imatinib. Ann Surg Oncol 2007;14:14-24.
24. DeMatteo RP, Maki RG, Singer S, et al. Results of tyrosine kinase inhibitor therapy followed by surgical resection for metastatic gastrointestinal stromal tumor. Ann Surg 2007;245:347-352.
25. Gronchi A, Fiore M, Miselli F, et al. Surgery of residual disease following molecular-targeted therapy with imatinib mesylate in advanced/metastatic GIST. Ann Surg 2007;245:341-346.
26. Mussi C, Ronellenfitsch U, Jakob J. Post-imatinib surgery in advanced/metastatic GIST: is it worthwhile in all patients? Ann Oncol. 2010 Feb;21(2):403-8. doi: 10.1093/annonc/mdp310. Epub 2009 Jul 23.
27. Van Den AbbeeleAD, Badawi RD, Manola J, et al. Effects of cessation of imatinib mesylate (IM) therapy in patients (pts) with IM- refractory gastrointestinal stromal tumors (GIST) as visualized by FDG- PET scanning [abstract]. J Clin Oncol 2004;22(14_Suppl):Abstract 3012).
28. Yeh C-N, Chen T-W, Tseng J-H, et al. Surgical management in metastatic gastrointestinal stromal tumor (GIST) patients after imatinib mesylate treatment. J Surg Oncol 2010;102:599-603.
29. Raut CP, Wang Q, Manola J, et al. Cytoreductive surgery in patients with metastatic gastrointestinal stromal tumor treated with sunitinib malate.Ann Surg Oncol 2010;17:407-415.
30. Kang YK, RyuMH,Yoo C, et al. Resumption of imatinib to control metastatic or unresectable gastrointestinal stromal tumours after failure of imatinib and sunitinib (RIGHT): a randomised, placebo-controlled, phase 3 trial. Lancet Oncol 2013;14:1175-1182.
31. Blay JY, Le Cesne A, Ray-Coquard I, et al. Prospective multicentric randomized phase III study of imatinib in patients with advanced gastrointestinal stromal tumors comparing interruption versus continuation of treatment beyond 1 year: the French Sarcoma Group. J Clin Oncol 2007;25:1107-1113.
32. Le Cesne A, Ray-Coquard I, Bui BN, et al. Discontinuation of imatinib in patients with advanced gastrointestinal stromal tumours after 3 years of treatment: an open-label multicentre randomised phase 3 trial. Lancet Oncol 2010;11:942-949.
33. Friedl W, Caspari R, Sengteller M. Can APC mutation analysis contribute to therapeutic decisions in familial adenomatous polyposis? Experience from 680 FAP families. Gut. 2001 Apr;48(4):515-21.
34. Durno C, Monga N, Bapat B, et al. Does early colectomy increase desmoid risk in familial adenomatous polyposis? Clin Gastroenterol Hepatol 2007;5:1190-1194.
35. Colombo C, Miceli R, Lazar AJ, et al. CTNNB1 45F mutation is a molecular prognosticator of increased postoperative primary desmoid tumor recurrence: An independent, multicenter validation study. Cancer 2013;119:3696-3702.
36. Mullen JT, DeLaney TF, Rosenberg AE, et al. beta-Catenin mutation status and outcomes in sporadic desmoid tumors. Oncologist 2013;18:1043-1049.
37. Godwin Y, McCulloch TA, Sully L. Extra-abdominal desmoid tumour of the breast: review of the primary management and the implications for breast reconstruction. Br J Plast Surg 2001;54:268-271.).
38. Kasper B, Baumgarten C, Garcia J. Desmoid Working G. An update on the management of sporadic desmoid-type fibromatosis: A European Consensus Initiative between Sarco-

ma PAtients EuroNet (SPAEN) and European Organization for Research and Treatment of Cancer (EORTC)/Soft Tissue and Bone Sarcoma Group (STBSG). Ann Oncol. 2017; 28:2399-2408.

39. Otero S, Moskovic EC, Strauss DC. Desmoid-type fibromatosis. Clinical radiology. 2015; 70:1038-1045.). . A case series of IA/RPDF treated with *en bloc* resection of the tumor with adhesive organs is reported for the first time in this study. Complete macroscopic resection was achieved in all cases.
40. Bonvalot S, Eldweny H, Haddad V. Extra-abdominal primary fibromatosis: Aggressive management could be avoided in a subgroup of patients. Eur J Surg Oncol. 2008; 34:462-468.
41. Fiore M, Rimareix F, Mariani L. Desmoid-type fibromatosis: A front-line conservative approach to select patients for surgical treatment. Ann Surg Oncol. 2009; 16:2587-2593.)
42. Menegazzo M, Tonello M, Bardini R. A 13 kg intra-abdominal mass: A case of mesenteric fibromatosis. Updates Surg. 2013; 65:237-240.
43. Wang Y, Cui NY, Li L. An abdominal desmoid-type fibromatosis. Quantitative imaging in medicine and surgery. 2013; 3:228-230.
44. Venkat D, Levine E, Wise WE. Abdominal pain and colonic obstruction from an intra-abdominal desmoid tumor. Gastroenterology & hepatology. 2010; 6:662-665.
45. Kreuzberg B, Koudelova J, Ferda J. Diagnostic problems of abdominal desmoid tumors in various locations. European journal of radiology. 2007; 62:180-185. 28. Gari MK, Guraya SY, Hussein AM, Hego MM. Giant mesenteric fibromatosis: Report of a case and review of the literature. World J Gastrointest Surg. 2012; 4:79-82.
46. Kobayashi H, Sugihara K.Intra-abdominal desmoid tumor after resection for gastrointestinalstromal tumor of the small intestine: Case report. Japanese journal of clinical oncology. 2014; 44:982-985.
47. Ogawa N, Iseki H, Tsunozaki H. Intra-abdominal desmoid tumor difficult to distinguish from a gastrointestinal stromal tumor: Report of two cases. Surgery today. 2014; 44:2174-2179.
48. Zhen Wang, Jianhui Wu, Ang Lv. *En bloc* resection for intra-abdominal/retroperitoneal desmoid-type fibromatosis with adjacent organ involvement: A case series and literature review . *BioScience Trends. 2018; 12(6):620-626.* DOI: 10.5582/bst.2018.01285.
49. Kouriefs C, Leris ACA, Mokbel K, et al. Infiltrating fibromatosis of the breast: a potential pitfall. Int J Clin Pract 2002;56:401-402.
50. Neuman HB, Brogi E, Ebrahim A, et al. Desmoid tumors (fibromatoses) of the breast: a 25-year experience. Ann Surg Oncol 2008;15:274-280.
51. Lev D, Kotilingam D, Wei C, et al. Optimizing treatment of desmoid tumors. J Clin Oncol 2007;25:1785-1791.
52. Pritchard DJ, Nascimento AG, Petersen IA. Local control of extra- abdominal desmoid tumors. J Bone Joint Surg Am 1996;78:848-854.
53. Ballo MT, Zagars GK, Pollack A, et al. Desmoid tumor: prognostic factors and outcome after surgery, radiation therapy, or combined surgery and radiation therapy. J Clin Oncol 1999;17:158-167.
54. Stojadinovic A, Hoos A, KarpoffHM,et al. Soft tissue tumors of the abdominal wall: analysis of disease patterns and treatment. Arch Surg 2001;136:70-79.
55. Ma D, Li S, Fu R, Zhang Z. Long-term outcomes of 47 patients with aggressive fibromatosis of the chest treated with surgery. Eur J Surg Oncol. 2016 Nov;42(11):1693-1698. doi: 10.1016/j.ejso.2016.06.396. Epub 2016 Jun 29
56. Gronchi A, Casali PG, Mariani L, et al. Quality of surgery and outcome in extra-abdominal aggressive fibromatosis: a series of patients surgically treated at a single institution. J Clin Oncol 2003;21:1390-1397
57. Mullen JT, Delaney TF, Kobayashi WK, et al. Desmoid tumor: analysis of prognostic factors and outcomes in a surgical series. Ann Surg Oncol 2012;19:4028-4035.

58. Janssen ML, van Broekhoven DL, Cates JM, et al. Meta-analsis of the influence of surgical margin and adjuvant radiotherapy on local recurrence after resection of sporadic desmoid-type fibromatozis.Br J Surg 2017; 104: 347-357.
59. Gluck I, Griffith KA, Biermann JS, et al. Role of radiotherapy in the management of desmoid tumors. Int J Radiat Oncol Biol Phys 2011;80:787-792.
60. Crago AM, Denton B, Salas S, et al. A prognostic nomogram for prediction of recurrence in desmoid fibromatosis. Ann Surg 2013;258:347-353.
61. Baumert BG, Spahr MO, Von Hochstetter A, et al. The impact of radiotherapy in the treatment of desmoid tumours.An international survey of 110 patients. A study of the Rare Cancer Network. Radiat Oncol . 2007 Mar 7;2:12).
62. Valentin T, Fournier C, Penel N, et al. Sorafenib in patients with progressive malignant solitary fibrous tumors: a subgroup analysis from a phase II study of the French Sarcoma Group (GSF/GETO). Invest New Drugs 2013;31:1626-1627.
63. Bonvalot S, Rimareix F, Causeret S et al. Hyperthermic isolated limb perfusion in locally advanced soft tissue sarcoma and progressive desmoidtype fibromatosis with TNF 1mg and melphalan (T1-M HILP) is safe and efficient. Ann Surg Oncol 2009; 16: 3350–3357.
64. Van Broekhoven DL, Deroose JP, Bonvalot S et al. Isolated limb perfusion using tumour necrosis factor a and melphalan in patients with advanced aggressive fibromatosis. Br J Surg 2014; 101: 1674–1680.
65. Kujak JL, Liu PT, Johnson GB, Callstrom MR. Early experience with percutaneous cryoablation of extra-abdominal desmoid tumors. Skeletal Radiol 2010; 39: 175–182.
66. Zeng WG, Zhou ZX, Liang JW, et al. Prognostic factors for DT: a surgical series of 233 patients at a single institution. Tumour Biol. 2014;35:7513–21. ,
67. Stoeckle E, Coindre JM, Longy M, et al. A critical analysis of treatment strategies in DTs: a review of a series of 106 cases. Eur J Surg Oncol. 2009;35(2):129–34.
68. Huang K, Wang CM, Chen JG, et al. Prognostic factors influencing event-free survival and treatments in desmoid-type fibromatosis: analysis from a large institution. Am J Surg. 2014;207(6):847–54.