

Large Desmoid Tumor Of The Anterior Abdominal Wall: A Case Report Of A 4.6kg Desmoid Tumor

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Abstract

Background: A desmoid tumor is a slowly growing neoplasm with no metastatic potential; occasionally it may attain large size.

Case presentation: We report a 4.6 kilogram anterior abdominal wall desmoid tumor in a female patient with no previous history of surgery. Preoperative ultrasound demonstrated a large tumor possibly arising from the ovary or broad ligament. Subsequent histology revealed a desmoid tumor.

Conclusion : Surgery with negative resection margins with or without radiotherapy remains the treatment of choice. Alternative treatment can be done for unresectable tumours.

BACKGROUND

Desmoid tumor, referred to as aggressive fibromatosis, is an uncommon neoplasm. It constitutes about 3% of all soft tissue neoplasms [1]. In the normal population the incidence of desmoid tumors is about 2.4 to 4.3 cases per million; this risk increases about 1000 fold in patients with familial adenomatous polyposis. This neoplasm can occur sporadically or as part of an inherited syndrome, familial adenomatous polyposis [36]. The tumor origin may be from fascia (superficial) or musculoaponeurotic (deep). The superficial disease is slowly growing, small and rarely involves deeper structures [36]. The deep variant has a relatively rapid growth, attains larger size and has a high local recurrence rate. Most spontaneous desmoids occur in the shoulder girdle and the anterior abdominal wall, whereas intra-abdominal desmoids, especially mesenteric desmoids are more common in patients with familial adenomatous polyposis. Abdominal wall desmoids typically occur in young females during gestation or, more frequently, within a year of childbirth. These associations are correlated with detection of estrogen receptors in the substance of these tumors. Molecular studies demonstrated desmoid tumors in FAP as clonal neoplasms arising from germ line mutation or changes in the APC alleles [5,6,7]. There is often a temporal association with history of abdominal trauma or operation. The typical presentation is that of a painless enlarging mass. Local symptoms may arise from compression of

neurovascular bundles [36]. Despite their benign histological appearance they are diffusely infiltrative. Treatment of these tumors is complete resection with a tumor free margin. Even with local resection with tumor free margins the local recurrence rate approaches 40%. Fortunately, systemic metastases are rare. Nuyttens and colleagues reported local control rates of 61% for surgery alone and 75% for surgery plus radiation. Antiproliferative agents and cytotoxic chemotherapy has been used to palliate the aggressive nature of desmoid tumors with variable results. Partial response has been observed after treatment with doxorubicin, dactinomycin, dacarbazine or carboplatin.

CASE PRESENTATION

A 29-year-old female presented with a slowly growing mass in the abdomen, initially infraumbilical, gradually increasing to involve the anterior abdominal wall up to the epigastrium superiorly and the pubic symphysis inferiorly as well as the bilateral flanks. There was no history of trauma or operation for any cause in the past. The mass was painless.

On examination, the patient had a lobulated firm mass extending from the epigastrium superiorly to the pubic bones inferiorly and to the bilateral lumbar region laterally. The mass was attached to the pubic bones inferiorly and became prominent on straight leg raising test. Overlying skin was stretched with prominent veins over the swelling.

A standing abdominal film showed a soft tissue mass over the whole abdomen.

Intraoperatively, the anterior rectus sheath adhered to the mass tightly and the fibrous septae in the rectus muscle caused the lobulated appearance of the tumor. The tumor was also adherent to parietal peritoneum and bladder on its posterior surface with strong adhesions to the pubic bones inferiorly as well as to the external iliac vein on the right side which needed vascular repair after excision of the tumor. The excised tumor weighed 4.6 kg. Subsequent histopathology revealed features of a desmoid tumor.

Figure 1

Operative specimen of the tumor

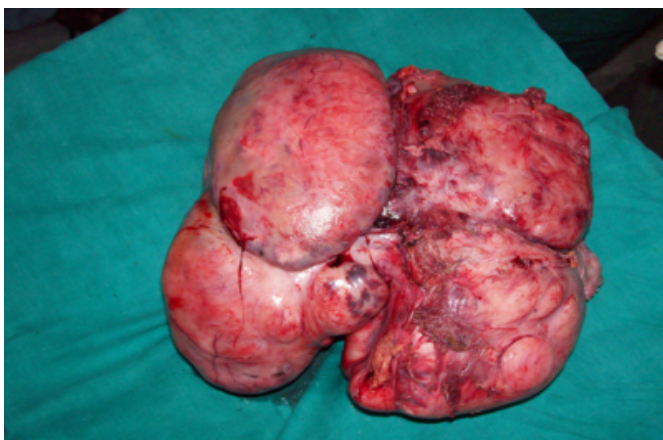
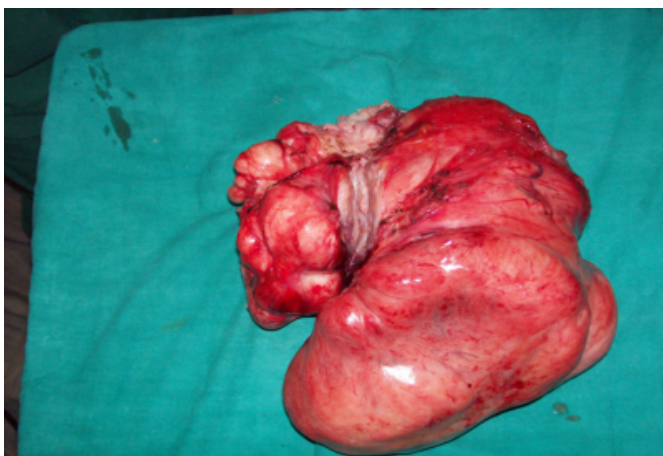


Figure 2



DISCUSSION

Desmoid tumors are benign deep fibromatoses, originating from fascia and muscle aponeurosis with an infiltrating growth [10]. Only sporadic cases localized within the thorax wall [11] or retroperitoneally [12] are described. Desmoid tumor is often associated with female gender, FAP [13] and

occasionally with surgical trauma [14]. It has a higher prevalence in women who experienced pregnancy. Depending on tumor size, therapy and negative resection margins, recurrence occurs in up to 45% [16].

CT scan localizes the tumor and excludes metastasis. MRI reveals its homogenous and intense appearance to muscles on T1 weighted images and demonstrates heterogeneous signal intensity which is slightly lower than that of fat on T2 weighted imaging, depending on the accumulation of mucoid structures [36]. Therefore, a differentiation from other solid tumors is impossible using morphological criteria [17]. Histology is the only evidentiary method which demonstrates long fascicles of spindle cells of variable cell-density with few mitoses and absence of atypical nucleus-separations. Characteristically, there is a diffuse cell infiltration of adjacent tissue structures.

Immunohistochemical response for actin can be partially positive and immunohistochemical muscle cell markers delimit desmoid tumors from fibrosarcomas [18].

The therapy of choice is still controversial [19]. Anti-inflammatory treatment [20], hormone-therapy [21] and chemotherapy [22] were not shown to be effective or were only partially effective [36]. These therapies are limited to patients, in whom resection is technically impossible because of a widespread tumor infiltration.

The effectiveness and indication of initial and adjuvant radiation is not proven yet. In a comparative analysis, a significantly better local recurrence control was described with radiation and combined surgical resection in comparison to resection only [16]. However, these results of radiation therapy can only be achieved with a higher complication rate [23].

Due to the germ line mutations and chromosomal aberrations of the APC alleles Bright-Thomas et al. performed a pre-clinical study of gene transfer for the treatment of desmoid disease in FAP [26]. Despite the success of transgene expression, further work is needed in animal models of desmoid disease to assess the clinical effects of gene therapy.

Therefore, the most effective treatment of accessible and smaller desmoid tumors is still the resection with negative margins, although it may not prevent local recurrence [3]. However, massive mesenteric lesions often occur in FAP patients after colectomy and non surgical treatment has variable and unpredictable objective efficacy. In conclusion,

there remains a need for a wider range of therapeutic options, especially in the management of these massive tumors.

Surgery always aims at radical tumor resection with free margins, which, depending on the localization of surgery, may leave major soft tissue defects behind [27,29,30]. Although abdominal wall integrity after full-thickness surgery can be restored with direct sutures [3,31], reconstruction with synthetic materials is a common technique in major abdominal wall defects. Although in our case the reconstruction of the abdominal wall was not performed, recent literature data recommend distant or free muscle flaps for greater abdominal wall defect coverage that is not accessible to local flaps. Additionally, prosthetic material is more susceptible to bacterial infection and other complications, although newly developed material showed encouraging experimental results.

In conclusion, the treatment of desmoid tumors remains controversial. Non-surgical treatment resulted in diverse and unpredictable outcome and is considered to be an opportunity in patients with unresectable lesions or for adjuvant therapy. Radical resection with clear margins remains the principal determinant of outcome with the risk of local recurrence. The abdominal wall defect often needs reconstruction using flaps or prosthetic material.

References

1. Kiel KD, Suit HD. Radiation therapy in the treatment of aggressive fibromatoses (desmoid tumors). *Cancer*. 1984;54:2051-2055. [PubMed]
2. Lewis JJ, Boland PJ, Leung DH, Woodruff JM, Brennan MF. The enigma of desmoid tumors. *Ann Surg*. 1999;229:866-872. doi: 10.1097/00000658-199906000-00014. [PubMed]
3. Merchant NB, Lewis JJ, Woodruff JM, Leung DH, Brennan MF. Extremity and trunk desmoid tumors: a multifactorial analysis of outcome. *Cancer*. 1999;86:2045-2052. doi: 10.1002/(SICI)1097-0142(19991115)86:10<2045::AID-CNCR23>3.3.CO;2-6. [PubMed]
4. Nugent KP, Spigelman AD, Phillips RK. Life expectancy after colectomy and ileorectal anastomosis for familial adenomatous polyposis. *Dis Colon Rectum*. 1993;36:1059-1062. [PubMed]
5. Dangel A, Meloni AM, Lynch HT, Sandberg AA. Deletion (5q) in a desmoid tumor of a patient with Gardner's syndrome. *Cancer Genet Cytogenet*. 1994;78:94-98. doi: 10.1016/0165-4608(94)90053-1. [PubMed]
6. Miyaki M, Konishi M, Kikuchi-Yanoshita R, Enomoto M, Tanaka K, Takahashi H, Muraoka M, Mori T, Konishi F, Iwama T. Coexistence of somatic and germ-line mutations of APC gene in desmoid tumors from patients with familial adenomatous polyposis. *Cancer Res*. 1993;53:5079-5082. [PubMed]
7. Palmirota R, Curia MC, Esposito DL, Valanzano R, Messerini L, Ficari F, Brandi ML, Tonelli F, Mariani-Costantini R, Battista P. Novel mutations and inactivation of both alleles of the APC gene in desmoid tumors. *Hum Mol Genet*. 1995;4:1979-1981. [PubMed]
8. de Wever I, Dal Cin P, Fletcher CD, Mandahl N, Mertens F, Mitelman F, Rosai J, Rydholm A, Sciort R, Tallini G, Van den Berghe H, Vanni R, Willen H. Cytogenetic, clinical, and morphologic correlations in 78 cases of fibromatosis: a report from the CHAMP Study Group. *CHromosomes And Morphology. Mod Pathol*. 2000;13:1080-1085. [PubMed]
9. Lynch HT, Fitzgibbons R, Jr. Surgery, desmoid tumors, and familial adenomatous polyposis: case report and literature review. *Am J Gastroenterol*. 1996;91:2598-2601. [PubMed]
10. Hasegawa SL, Fletcher CDM. Fibromatosis in the adult. *Adv Pathol*. 1996;9:259-275.
11. Gacouin A, Desrues B, Lecoq A, Quinquenel ML, Ramee MP, Delaval P. [Desmoid tumor of the thoracic wall]. *Rev Mal Respir*. 1993;10:554-556. [PubMed]
12. Budzynski A, Wysocki A. [Retroperitoneal desmoid tumor]. *Przeegl Lek*. 1996;53:506-507. [PubMed]
13. Soravia C, Berk T, McLeod RS, Cohen Z. Desmoid disease in patients with familial adenomatous polyposis. *Dis Colon Rectum*. 2000;43:363-369. [PubMed]
14. De Cian F, Delay E, Rudigoz RC, Ranchere D, Rivoire M. Desmoid tumor arising in a cesarean section scar during pregnancy: monitoring and management. *Gynecol Oncol*. 1999;75:145-148. doi: 10.1006/gyno.1999.5539. [PubMed]
15. Gansar GF, Markowitz IP, Cerise EJ. Thirty years of experience with desmoid tumors at Charity Hospital. *Am Surg*. 1987;53:318-319. [PubMed]
16. Nuyttens JJ, Rust PF, Thomas CR, Jr, Turrisi AT, III. Surgery versus radiation therapy for patients with aggressive fibromatosis or desmoid tumors: A comparative review of 22 articles. *Cancer*. 2000;88:1517-1523. doi: 10.1002/(SICI)1097-0142(20000401)88:7<1517::AID-CNCR3>3.3.CO;2-0. [PubMed]
17. Casillas J, Sais GJ, Greve JL, Iparraguirre MC, Morillo G. Imaging of intra- and extraabdominal desmoid tumors. *Radiographics*. 1991;11:959-968. [PubMed]
18. Mentzel T, Katenkamp D. [Myofibroblastic tumors. Brief review of clinical aspects, diagnosis and differential diagnosis]. *Pathologe*. 1998;19:176-186. doi: 10.1007/s002920050271. [PubMed]
19. Shields CJ, Winter DC, Kirwan WO, Redmond HP. Desmoid tumours. *Eur J Surg Oncol*. 2001;27:701-706. doi: 10.1053/ejso.2001.1169. [PubMed]
20. Waddell WR, Kirsch WM. Testolactone, sulindac, warfarin, and vitamin K1 for unresectable desmoid tumors. *Am J Surg*. 1991;161:416-421. [PubMed]
21. Wilcken N, Tattersall MH. Endocrine therapy for desmoid tumors. *Cancer*. 1991;68:1384-1388. [PubMed]
22. Patel SR, Evans HL, Benjamin RS. Combination chemotherapy in adult desmoid tumors. *Cancer*. 1993;72:3244-3247. [PubMed]
23. Goy BW, Lee SP, Eilber F, Dorey F, Eckardt J, Fu YS, Juillard GJ, Selch MT. The role of adjuvant radiotherapy in the treatment of resectable desmoid tumors. *Int J Radiat Oncol Biol Phys*. 1997;39:659-665. doi: 10.1016/S0360-3016(97)00334-9. [PubMed]
24. Rock MG, Pritchard DJ, Reiman HM, Soule EH, Brewster RC. Extra-abdominal desmoid tumors. *J Bone Joint Surg Am*. 1984;66:1369-1374. [PubMed]
25. Pritchard DJ, Nascimento AG, Petersen IA. Local control of extra-abdominal desmoid tumors. *J Bone Joint Surg Am*. 1996;78:848-854. [PubMed]
26. Bright-Thomas RM, Agrawal A, Hargest R. Preclinical studies of gene transfer for the treatment of desmoid disease in familial adenomatous polyposis. *Br J Surg*.

- 2002;89:1563-1569. doi: 10.1046/j.1365-2168.2002.02277.x. [PubMed]
27. Middleton SB, Phillips RK. Surgery for large intra-abdominal desmoid tumors: report of four cases. *Dis Colon Rectum*. 2000;43:1759-1762. [PubMed]
28. Okuno SH, Edmonson JH. Combination chemotherapy for desmoid tumors. *Cancer*. 2003;97:1134-1135. doi: 10.1002/cncr.11189. [PubMed]
29. Brenner P, Rammelt S. Abdominal wall and foot reconstruction after extensive desmoid tumor resection with free tissue transfer. *Langenbecks Arch Surg*. 2002;386:592-597. doi: 10.1007/s00423-002-0277-y. [PubMed]
30. Sutton RJ, Thomas JM. Desmoid tumours of the anterior abdominal wall. *Eur J Surg Oncol*. 1999;25:398-400. doi: 10.1053/ejso.1999.0664. [PubMed]
31. Rohrich RJ, Lowe JB, Hackney FL, Bowman JL, Hobar PC. An algorithm for abdominal wall reconstruction. *Plast Reconstr Surg*. 2000;105:202-216. [PubMed]
32. Bauer JJ, Salky BA, Gelernt IM, Kreel I. Repair of large abdominal wall defects with expanded polytetrafluoroethylene (PTFE). *Ann Surg*. 1987;206:765-769. [PubMed]
33. Disa JJ, Klein MH, Goldberg NH. Advantages of autologous fascia versus synthetic patch abdominal reconstruction in experimental animal defects. *Plast Reconstr Surg*. 1996;97:801-806. [PubMed]
34. Leber GE, Garb JL, Alexander AI, Reed WP. Long-term complications associated with prosthetic repair of incisional hernias. *Arch Surg*. 1998;133:378-382. doi: 10.1001/archsurg.133.4.378. [PubMed]
35. Disa JJ, Chiaramonte MF, Girotto JA, Klein MH, Goldberg NH. Advantages of autologous fascia versus synthetic patch abdominal reconstruction in experimental animal defects. *Plast Reconstr Surg*. 2001;108:2086-2087. [PubMed]
36. Sabiston textbook of surgery, 17th edition.

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