Case Report

Desmoid Tumor of the Abdominal Wall: A Case Report

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Abstract

Desmoid tumors alternatively known as aggressive fibromatosis are benign myofibroblastic neoplasms originating from the muscle aponeurosis and classified as deep fibromatoses and these tumors are non-cancerous growth that occurs in the connective tissue. These tumors constitute 3% of all soft tissue tumors and 0.03% of all neoplasms and they occur usually between the age group of 20-40 years with a strong prevalence among women with fertile age group. In female patients presenting a tumor of the lower abdominal wall especially after cesarian section, an endometriotic tumor as well as an aggressive desmoid tumor should be considered. Symptoms in correlation with the monthly period can facilitate the presurgical differentiation between endometriosis and fibromatosis. Ultrasound reveals the typical location of both tumors and its remarkable sonographic appearance. In the clinical practice, the desmoid fibromatosis of the lower abdominal wall is a very rare disease. We present a case of a 19-year-old pregnant and discuss diagnostic and therapeutic options by literature review. With the knowledge of the prognosis of the desmoid fibromatosis and the respective treatment options including wait and see, complete surgical resection with macroscopically free margins and adjuvant approaches is essential to avoid further interventions and progression of the locally destructive tumor.

Keywords: Abdominal wall mass, Benign mesenchymal tumor, Endometrioma, Desmoid tumor

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Introduction

Desmoid tumors are benign myofibroblastic neoplasms originating from the muscle aponeurosis and classified as deep fibromatoses[1]. Desmoid tumors are non-cancerous growth that occurs in the connective tissue. Another term for desmoid tumors is aggressive fibromatosis. They occur usually between the age group of 20-40 years with a strong prevalence among women with fertile age group. Despite their aggressive local infiltration, desmoid tumors lack a metastatic potential[2]. However, because of this local infiltration and compression of surrounding structures, a high recurrence rate exists and in anatomic locations with restricted access to surgical resection desmoid tumors can lead to death[3]. Especially in patients with familial adenomatous polyposis (FAP) undergoing colectomy desmoid tumors are the leading cause of morbidity and mortality[4].

Molecular studies demonstrated desmoid tumors in FAP as clonal neoplasms arising from germ line mutation or changes in the APC alleles[5]. Cytogenic data verified clonal chromosome aberrations in deep-seated sporadic extra-abdominal fibromatoses and lesions of the abdominal wall and therefore provide additional evidence for the neoplastic nature of these lesions[6]. In sporadic cases, they occur in localisations of trauma, and scars or irradiation. The therapeutic management of these tumors is still controversial. Treatment for desmoid tumor include:1) Monitoring the growth of the tumor, if no sign and symptom 2) Surgery 3) Radiotherapy 4) Chemotheraphy and other medication. This case report present a case of abdominal wall desmoids tumor mimicking endometrioma.

Case report

A 19 years old women, who gave birth by caesarean section(c/s), admitted in Eastern Medical College and Hospital with the complaints of gradually increasing painful abdominal mass under her caesarean section(c/s) incision scar, suprapubic region. She had a c/s operation 2 years ago without known post-operative complication. She had severe pain particularly during menstruation. A mass about (6x6) cm in diameter, solid, painful and semi mobile mass below the c/s scar on suprapubic region was detected by abdominal physical examination. Clinically the case diagnosed as endometrioma.

USG examination revealed mixed echogenic area (6.23x4.31) cm at the anterior to urinary bladder wall, it was below the parietal wall of hypogastrium. Diagnosis: complex pelvic subcutaneous mass, due

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to haematoma/ endometrioma. For the further evaluation FNAC of swelling (mass) was done and send for cytopathology. Surprisingly, fibromatosis (desmoids) was found on cytological examination.

So, we performed wide surgical excision of (6.2x4.5) cm solid mass with 1 cm tumor free margin. There is no need to repair of fascia and use of mash. The tumor was a dense, bright beige yellow in color and located in rectus sheath. When sectioned it was homogenous and firm in consistency. Microscopic examination of the resected specimen in the cases revealed the spindle-cellular tumors immigrated through muscle-tissues consisting of partially intact muscle fibers surrounded by spindle like elements. Tumor-cells had a pale eosinophilic cytoplasm and chromatin structures and were embedded in collagen network interrupted by fibrotic sections. The pathological diagnosis was desmoid type fibromatosis, classic desmoid tumor (abdominal fibromatosis).

The post-operative course was uneventful and recovered without any complications or functional defect. We recommended clinical and ultrasonological control of tumors location. For 8 months after surgical intervention, the patient is recurrence free.

Figure-1: Cut section of desmoid tumor

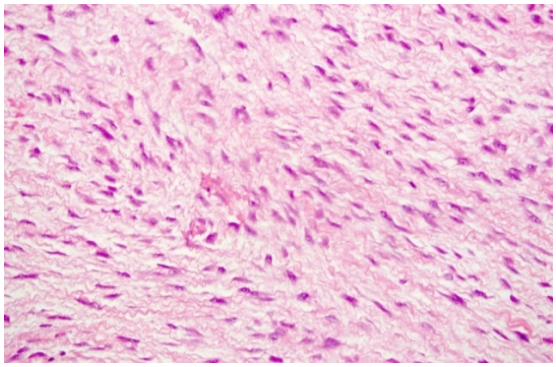

Figure-2: Histopathological view of desmoid tumor

Discussion

The desmoid fibromatosis is characterized by local aggressive growth without any tendency of metastasis. These very rare tumors can develop in any musculo-aponeurotic structure, and they can be found in all regions of the human body. The commonest site of presentation in case of abdominal wall desmoids seems to be the infra umbilical rectus sheath. The clinical behaviour and the prognosis of the desmoids is very diverse and depends on the anatomic location and the proximity to vitally important organ[7].

They can be divided into five subgroups: extra abdominal, intra-abdominal, multiple, multiple familial and as part of Gardner's syndrome. Extra abdominal desmoid tumors have a wide distribution; the shoulder girdle, trunk and lower extremities are most involved. Abdominal desmoids, which may occur in the abdominal wall, mesentery, or retro peritoneum, have an increased incidence in individuals with Gardner syndrome. The histologic findings in these lesions are identical[8,9]. In our case desmoid occur in infra umbilical rectus sheath. As the location of the tumor is not proximity with vital organs no sever symptom, only local swelling and pain.

A correlation with the familiar intestinal polyposis could be shown. Approximately 10–25% of the patients with polyposis present intra- or extra abdominal desmoid tumors[10]. But in our case, there is no correlation with the familial intestinal polyposis.

The commonest groups associated with these tumors are young women during or after pregnancy. The fibroblast has been shown to exhibit a proliferative response to oestrogen. In this case the patient is young, age 19 years and swelling appear 2 years after pregnancy.

There is a well-known association in patients with a history of abdominal or pelvic surgery. This tumor is also associated with trauma, oestrogen therapy, FAP and Gardner syndrome[11]. In the presented case, the local tissue trauma of the caesarean section in the clinical history of the patient is one possible risk factor of the disease.

Most of the abdominal wall desmoids measure 5 cm by 15 cm in diameter. These masses have a firm, gritty texture. These tumors have no distinct capsule, and their margins are ill defined even when they appear well circumscribed on imaging[12]. Our patient presented with a painful mass measuring 6 cm in maximal diameter.

The desmoid tumor of the lower abdominal wall, especially around a caesarean section scar, can be

confound with a scar endometriosis. A clear clinical diagnosis of the tumors origin and a differentiation to endometriosis by palpation and ultrasound are not possible. The intramuscular location of the tumor in ultrasound is an indicator for a desmoid tumor. While desmoids in almost all cases are located close to a muscular aponeurosis, the endometriotic tumors of the abdominal wall can be found most frequently in the subcutaneous region. In the actual literature, only 18 case reports about endometriosis exclusively located in the rectus muscle sheath can be found[13]. In this case USG examination revealed mixed echogenic area (6.23x4.31) cm at the anterior to urinary bladder wall, it was below the parietal wall of hypogastrium. So, initially it was diagnosed as a case of endometrioma.

Wide local excision followed by reconstruction of the defect is the treatment of choice. Full-thickness resection of the tumor-containing abdominal wall with a grossly negative margin must be performed when the lesion closely approximates or involves the peritoneum. Intra peritoneal organs or adjacent bony structures involved by tumor must be resected as well. Incomplete tumor removal or involved excision margins may lead to local recurrence[14]. In our case wide local excision of the tumor with 1 cm tumor free margin was done, no intra-abdominal organ was involved. In our case no recurrence occurs after 8 months of surgery.

Cormio et al. reported a recurrence-free period of 30 month after surgical resection of a pelvic fibromatosis without any adjuvant treatment[15]. In a retrospective review of 151 patients who underwent a macroscopically complete resection of desmoid tumors, Huang et al. showed a local recurrence rate of 20.5%.

Salas et al. showed three unfavourable prognostic factors: age less than 37 years, tumor size >7cm, and extra-abdominal tumor location[16]. The most important factor for recurrence was a tumor size of >5cm 17. In our case the patient was 19 years age and the tumor size was less than 7 cm. Bertani et al. described a recurrence-free follow-up period of 55 months in 14 patients with disease-free margins of >1cm in intraoperative frozen section[18]. Chao et al. also described the complete regression of a tumor recurrence by tamoxifen[19].

Maseelall et al. reported the successful postoperative therapy of a pelvic fibromatosis with Toremifene. Constantinidou et al. published the successful use of chemotherapy in advanced aggressive fibromatosis with no response to hormonal treatment[20]. In another actual publication the possibility of a therapy with tyrosine kinase inhibition in inoperable tumors is described[21]. Radiation therapy is used in patients with inoperable tumors, local recurrences, or incompletely excised lesions. Chemotherapy and endocrine therapy have also been used to treat desmoid tumors in patients in whom resection is technically impossible because of a widespread tumor infiltration[22].

Conclusion

The combination of features, such as the history of previous surgery, the age and sex of the patient, the location of the mass within the anterior abdominal wall and the imaging features, make desmoid tumor a strong primary diagnostic consideration even if it is a rare entity. The treatment approach remains aggressive and includes complete surgical resection. Repair of abdominal wall defects can be sufficiently achieved with prosthetic mesh reconstruction with excellent functional results.

References

1. Fletcher CDM. Myofibroblastic tumours: an update. Verh Dtsch Ges Path. 1998; 82: 75-82.
2. Lewis JJ, Boland PJ, Leung DH, Woodruff JM, Brennan MF. The enigma of desmoid tumors. Ann Surg. 1999; 229 (6): 866-72.
3. Merchant NB, Lewis JJ, Woodruff JM, Leung DH, Brennan MF. Extremity and trunk desmoid tumors: a multifactorial analysis of outcome. Cancer. 1999; 86 (10): 2045-52.
4. Nugent KP, Spigelman AD, Phillips RK. Life expectancy after colectomy and ileorectal anastomosis for familial adenomatous polyposis. Dis Colon Rectum. 1993; 36 (11): 1059-62.
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: 94-8.
6. de Wever I, Dal Cin P, Fletcher CD, Mandahl N, Mertens F, Mitelman F, et al. Cytogenetic, clinical, and morphologic correlations in 78 cases of fibromatosis: a report from the CHAMP Study Group. CHromosomes And Morphology. Mod Pathol. 2000; 13: 1080-5.
7. Mulik V, Griffiths AN, Beattie RB. Desmoid tumours with familial adenomatous polyposis in pregnancy. J Obstet Gynaecol. 2003; 23 (3): 307-8.
8. Teo HEL, Peh WCG, Shek TWH. Case 84: desmoid tumor of the abdominal wall. Radiology. 2005; 236: 81-4.
9. Cormio G, Cormio L, Marzullo A, et al. Fibromatosis of the female pelvis. Annales Chirurgiae et Gynaecologiae. 1997; 86 (1): 84-6.
10. Sinha A, Gibbons DC, Phillips RK, Clark S. Surgical prophylaxis in familial adenomatous polyposis: do pre-existing desmoids outside the abdominal cavity matter? Familial Cancer. 2010; 9 (3): 407-11.

11. Kumar V, Khanna S, Khanna AK, Khanna R. Desmoid tumors: experience of 32 cases and review of the literature. Indian J Cancer. 2009; 46: 3.
12. Casillas J, Sais GJ, Greve JL, Iparraguirre MC, Morillo G. Imaging of intraand extra abdominal desmoid tumors. Radiographics. 1991; 11: 959-68.
13. Giannella L, La Marca A, Ternelli G, Menozzi G. Rectus abdominis muscle endometriosis: case report and review of the literature. J Obstet Gynaecol Res. 2010; 36 (4): 902-6.
14. Lahat G, Nachmany I, Itzkowitz E, Abu-Abeid S, Barazovsky E, Merimsky O, et al. Surgery for sporadic abdominal desmoid tumor: is low/no recurrence an achievable goal? IMAJ 2009; 11: 398-402.
15. Galeotti F, Facci E, Bianchini E. Desmoid tumour involving the abdominal rectus muscle: report of a case. Hernia. 2006; 10 (3): 278-81.
16. Granese R, Cucinella G, Barresi V, Navarra G, Candiani M, Triolo O. Isolated endometriosis on the rectus abdominis muscle in women without a history of abdominal surgery: a rare and intriguing finding. J Minimally Invasive Gynecol. 2009; 16 (6): 798-801.
17. Gourgiotis S, Veloudis G, Pallas N, Lagos P, Salemis NS, Villias C. Abdominal wall endometriosis: report of two cases. Romanian J Morphol Embryol. 2008; 49 (4): 553-55.
18. Bertani E, Chiappa A, Testori A, et al. Desmoid tumors of the anterior abdominal wall: results from a monocentric surgical experience and review of the literature. Annals Surg Oncol. 2009; 16 (6): 1642-9.
19. Chao AS, Lai CH, Hsueh S, Chen CS, Yang YC, Soong YK. Successful treatment of recurrent pelvic desmoid tumour with tamoxifen. Human Reproduction. 2000; 15 (2): 311-3.
20. Constantinidou A, Jones RL, Scurr M, Al-Muderis O, Judson I. Advanced aggressive fibromatosis: effective palliation with chemotherapy. Acta Oncologica. 2011; 50 (3): 455-61.
21. Chugh R, Wathen JK, Patel SR, et al. Efficacy of imatinib in aggressive fibromatosis: results of a phase II multicenter Sarcoma Alliance for Research through Collaboration (SARC) trial. Clinical Cancer Research. 2010; 16 (19): 4884-91.
22. Overhaus M, Decker P, Fischer HP, Textor HJ, Hirner A. Desmoid tumors of the abdominal wall: a case report. World J Surg Oncol. 2003; 1: 11.

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