Misdiagnosis of aggressive fibromatosis of the abdominal wall

A case report and literature review

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Abstract

Rationale: Aggressive fibromatosis (AF) of abdominal wall is also called desmoid tumor, ligament tumor, fibrous tissue tumor hyperplasia, tendon membrane fibroma or soft tissue ligament fibroma, etc. Aggressive fibromatosis of abdominal wall was first described by MacFarlane in 1832, and it was named for the first time by Muller according to its general appearance and texture in 1838. This disease has been mistaken for a benign lesion for a long time because when the cells were examined by pathology often show normal mitosis, and distant metastases are not found clinically, but actually the disease is locally invasive and shows a local invasive growth. So it is a rare low-grade malignant soft tissue tumor. At present, the main treatment for the disease is operation, and radiotherapy and hormone therapy have a certain effect, but these therapies are not ideal.

Patient concerns: A 32-year-old woman, who underwent cesarean section three years ago came to the hospital for finding a mass on abdominal wall for half a month.

Diagnoses: Mass of abdominal wall.

Interventions: Underwent surgery.

Outcomes: Pathology: The lesion is aggressive fibromatosis of abdominal wall (ligament tumor of abdominal wall).

Lessons: We discussed the particularity of its clinical characteristics, treatment strategies and prognosis combined with literature review, and we think the surgeons need to pay high attention to this disease and make more patients get timely, correct and reasonable treatment, so as to improve the quality of life.

Abbreviation: AF = aggressive fibromatosis.

Keywords: aggressive fibromatosis, low-grade malignant, radiotherapy, surgery

Aggressive fibromatosis (AF) of abdominal wall is also called desmoid tumor, ligament tumor, fibrous tissue tumor hyperplasia, tendon membrane fibroma or soft tissue ligament fibroma, etc. Aggressive fibromatosis of abdominal wall was first described by MacFarlane in 1832, and it was named for the first time by Muller according to its general appearance and texture in 1838. This disease has been mistaken for a benign lesion for a long time because when the cells were examined by pathology often show normal mitosis, and distant metastases are not found clinically, but actually the disease is locally invasive and shows a local invasive growth. So it is a rare low-grade malignant soft tissue tumor. At present, the main treatment for the disease is operation, and radiotherapy and hormone therapy have a certain effect, but these therapies are not ideal. The author describes a case of AF of the abdominal wall that was misdiagnosed as abdominal wall endometriosis before surgery. The clinical characteristics, treatment strategies, and prognosis are discussed, together with a literature review. The study procedure was approved by Ethics Committee of the First Hospital of Jilin University. All the subjects had given the written informed consent.

1. Case report

A 32-year-old woman presented to the hospital with the complaint of a mass on the abdominal wall for several weeks. This gravida 2 para 1 patient usually had regular menses, with a cycle length of approximately 22 to 23 days, and moderate flow lasting for about 5 days. She had mild and tolerable dysmenorrhea. The patient had a cesarean section 3 years earlier in a local hospital, and was found to have a palpable lower abdominal mass with mild tenderness and symptom exacerbation during her menstrual period. The patient was hospitalized for a mass on the abdominal wall. She had no abdominal pain, distension, fever, dizziness, fatigue, palpitations, or shortness of breath, and had a good diet and sleep quality, with normal stools and urine, and no obvious change in weight. She had a previous history of left adnexectomy for an “ovarian cyst” 10 years prior in another hospital, and was told that the pathological findings were benign.
and is also seen in the mesocolon, gastrocolic ligament, greater omentum, and retroperitoneum.\[9\]

The etiology of AF is unknown, but it may have a genetic origin, or may be associated with endocrine abnormalities or physical trauma.\[10\] The disease often occurs in those aged 10 to 40 years. The incidence in women is 2 to 3 times higher than that in men.\[11\]

AF of the abdominal wall shows no specificity on imaging, and is difficult to distinguish from other soft tissue tumors, especially abdominal wall endometriosis. Abdominal wall ultrasonic imaging shows a hypoechoic mass, while color Doppler ultrasound shows an enhanced blood signal. Imaging is helpful to determine the tumor location and extent of infiltration, but lacks diagnostic specificity, and cannot distinguish AS from other soft tissue tumors. Pathologic examination is the primary means of diagnosis. Gross specimens range from 3 to 20cm in size, according to the location. The mass often appears irregular, with unclear boundaries, a coarse cross section, and pale color, with braiding and scar-like tissue during invasive growth. Under a microscope, the tumor is composed of abundant collagen fibers and small fiber cells arranged in parallel, with cells demonstrating abnormal division. Striated muscle islands can often be seen at the tumor margin, surrounded by tumor tissue.

2.2. Treatment and prognosis

There is no standard treatment for AS because the biological characteristics include local invasion and a high recurrence rate. Treatment includes surgery, radiotherapy, chemotherapy, hormonal therapy, and conservative management. Individualized comprehensive treatment based on surgery is recommended. The appropriate extent of tumor excision is controversial, because of difficulty in determining tumor aggressiveness before surgery. The operative principle is removal of the tumor to the greatest possible extent, with maximum protection of surrounding vital organs. It is generally acknowledged that resection of the tumor with a margin of at least 2 to 3cm is necessary.\[13\] A frozen pathological examination should be performed if complete excision is uncertain. Some studies have reported that a positive margin is unrelated to postoperative recurrence. However, most researchers believe that a positive surgical margin is associated with tumor recurrence and that patients with a positive margin have a much higher postoperative recurrence risk. A negative margin does not rule out postoperative recurrence. Moreover, local structure and function may be adversely affected by an attempt to achieve a negative margin. The goal of complete tumor excision should be maintained while trying to reduce the extent of surgical resection.

Some researchers believe that radiotherapy does not reduce the postoperative local recurrence rate, and may lead to complications such as edema, cellulitis, fibrosis, ulcers, and pathological fractures. Radiotherapy even has the potential to induce other local malignant tumors, and should not be performed in patients with an initial negative margin but may be considered during postoperative follow-up.\[14\] Patients with a positive margin should receive postoperative radiotherapy because of its curative effect on AF. Tumor regression after radiotherapy may ensure complete resection under the premise of protection of vital organs. Postoperative supplemental radiotherapy can result in a curative rate in patients with a positive margin similar to that of patients with a negative margin.\[15\] Postoperative supplemental radiotherapy is standard for patients with a positive margin, and can achieve an 80% local control rate.\[12\] Postoperative
radiotherapy is recommended for patients undergoing reoperation after recurrence, while radiotherapy alone can be effective for patients who cannot tolerate surgery. Postoperative supplemental radiotherapy should be performed as early as possible since the local recurrence rate is as high as 50% in cases with a negative margin after secondary surgery.

Surgery and radiotherapy have good therapeutic effects in AF. Chemotherapy and endocrine and targeted therapy are generally reserved for patients who cannot undergo surgery or radiotherapy because the AF has invaded vital organs or the patient has had multiple postoperative recurrences. For such patients, these treatments can reduce pain and reduce the size of the mass, creating opportunities for further surgery. Studies have confirmed the effectiveness of adriamycin combined with nitroimidine-amine in the treatment of AF and that of antiestrogen therapy for patients whose are estrogen receptor-positive. The targeted therapeutic effect of imatinib has gained attention for its significant effect on AF, and is undergoing further clinical research.

AF rarely causes death from distant metastasis. Patients can survive with the tumor for a long time, and the prognosis is good. However, long-term follow-up is needed, and recurrences require prompt treatment.

This case report and literature review may be helpful in the diagnosis and treatment of AF, and may ensure timely, correct, and appropriate treatment.

References

[1] Ibrahim M, Sandogji H, Allam A. Huge intrathoracic desmoid tumor. Ann Thorac Med 2009;4:146–8.
[2] MacFarlane J. Clinical Reports on the Surgical Practice of the Glasgow Royal Infirmary [M]. 5th ed.1832;Glasgow Royal Infirmary, Glasgow:63–66.
[3] Muller J, Ueber den Feineren Bau und Die Formen der Krankhaften Geschwulste [M]. 1838;G Reimer, Berlin:60.
[4] Fujishina T, Yoshida H, Obi S, et al. Analysis of factors influencing hepatocellular carcinoma detection: efficient use of computed tomography during arterial portography and during hepatic arteriography. J Gastroenterology 2005;40:266–73.
[5] Hani S, Zadeh T. Desmoin tumors of the pediatric mandible. Annplast Surg 2009;62:213–9.
[6] De Bree E, Keus R, Melissas J, et al. Desmoid tumors: Need for an individualized approach. Expert Rev Anticancer Ther 2009;9:525–35.
[7] Kreuzberg B, Koudelova J, Ferda J, et al. Diagnostic problems of abdominal desmoid tumors in various locations. Eur J Radiol 2007;62:180–5.
[8] Kriz J, Eich HT, Haverkamp U, et al. Radiotherapy is effective for desmoid tumors (aggressive fibromatosus) - long-term results of a German multicenter study. Oncol Res Treat 2014;37:255–60.
[9] Ren YY, Fu YQ. The progress of diagnosis and treatment of neurofibromatosis. J Zhongming Chin Med Uni 2010;34:127–8.
[10] Xu HM, Ma SZ, Wang ZJ. The treatment of abdominal desmoid tumor. Chin J Gen Surg 2010;25:254–6.
[11] Hosalkar HS, Fox EJ, Delaney T, et al. Desmoid tumours and current status of management. Orthop Clin North Am 2006;37:53–63.
[12] Micke O, Seegenschmiedt MH. Radiation therapy for aggressive fibromatosism (desmoid tumors) — results of a national patterns of care study. Int J Radiat Oncol Biol Phys 2003;57(Suppl):2–5.
[13] Dong RZ, Shi QY, Wang CM. Clinical analysis for 84 cases of abdominal wall ligament tumor. China Cancer 2008;17:1079–81.
[14] Nadler RB. Bladder training biofeedback and pelvic floormy-algia. Urology 2002;60(6 suppl):42–3.
[15] Micke O, Seegenschmiedt MH. Radiation therapy for aggressive fibromatosism (desmoid tumors): results of a national patterns of care study. Int J Radiat Oncol Biol Phys 2005;61:882–91.
[16] Shreyaskumar R, Harry L, Robert S. Combination chemotherapy in adult desmoid tumors. Cancer 1983;72:1244–7.
[17] Piciariello L, Sala SC, Martineti V, et al. A comparisios of methods for the analysis of low abundance proteins in desmoid tumor cells. Anal Biochem 2006;354:203–12.
[18] Baker LH, Wathen K, Gough R, et al. Activity of imatinibmesylate in desmoid tumors: interim analysis of a Sarcoma Alliance for Research through Collaboration (SARC) phase 2 trial (abstract). Proc Am Soc Clin Onc 2004;22:821.