Clinical Case Report

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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 lesions 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.[1] Aggressive fibromatosis of abdominal wall was first described by MacFarlane in 1832,[2] and it was named for the first time by Muller according to its general appearance and texture in 1838.[3] This disease has been mistaken for a benign lesions 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

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(no report was available). She was told she had "hepatitis B" 3 years earlier, without formal diagnosis or therapy. She had no history of hypertension, coronary heart disease, diabetes mellitus, or tuberculosis. She did not drink or smoke.

On admission, the abdomen was flat and soft, without varicosities or visible masses. A mass with diameter of about 3 cm was palpable near the cesarean section scar in the periumbilical region. The mass was hard, with a smooth surface, little tenderness, a clear boundary, and good mobility; there was no rebound tenderness or guarding. Abdominal ultrasonography showed a hypoechoic mass in the abdominal wall musculature in the left hypogastric region, measuring 29 mm × 29 mm × 18 mm. The mass had a clear boundary and an irregular appearance, with surrounding punctate blood flow signals. Endometriosis could not be ruled out.

At surgery, the periumbilical caesarean scar was found to be about 1 cm wide. The scar tissue in the original incision was excised. The skin and subcutaneous fat were incised and the mass could be palpated anterior to the peritoneum. The mass had a diameter of about 5 cm, and was palpable in the extraperitoneal musculature below and to the left of the periumbilical incision after the peritoneum was opened. There was a clear boundary between the mass and the peritoneum and there were no adhesions. The abdominal wall musculature was adjacent to the mass, with about 2 to 3 cm between the muscle and the border of the mass. Two curved forceps were used to clamp the muscles, and the mass was cut, sutured, and divided. The wound surface was sutured to stop bleeding and the abdominal wall mass was completely excised.

Pathological findings revealed AF of the abdominal wall (ligament tumor of the abdominal wall), measuring 2.5 cm × 2.5 cm × 2 cm. The tumor was present at the excision margin. Immunohistochemical findings were positive for SMA and nuclear beta-catenin in sporadic cells, whereas CD34, desmin, and S-100 were negative. Less than 5% of cells were Ki67-positive. The diagnosis was an intermediate tumor with low-grade malignant biological behavior and tendency for recurrence.

2. Discussion

2.1. Clinical characteristics

AF is a rare tumor derived from tendon membrane structures, and is formed by excessive proliferation of fibroblasts. This disease was classified as a fibroblast/myofibroblast intermediate (locally invasive) tumor by the World Health Organization in the report on soft tissue tumor pathology and genetics in 2002, and was characterized by deep soft tissue cloning with fibroblast proliferation.[4] AF accounts for about 0.03% of all soft tissue tumors.[5] AF has no envelope and is always closely adherent to nearby muscles and nerves; therefore, local recurrence is likely, even after extensive resection. About 8% of patients die because of rapid recurrence, and 20% to 30% achieve stability or spontaneous remission.[6] This characteristic distinguishes AF from other benign tumors. AF can be divided into extra-abdominal, abdominal wall, and intra-abdominal types. The extra-abdominal type is most common (50%–60%), followed by the abdominal wall (25%) and intra-abdominal (15%) types.[7] The tumor can occur in any location, but usually involves the back, head and neck, chest and abdominal wall, abdominal cavity, or extremities. AF in the abdomen is always found in the small bowel mesentery, as in familial adenomatous polyposis,[8] 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 wall 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 20 cm 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 3 cm 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 nitreneamidine-amine in the treatment of AF,[16] and that of antiestrogen therapy for patients whose are estrogen receptor-positive.[17] The targeted therapeutic effect of imatinib has gained attention for its significant effect on AF, and is undergoing further clinical research.[18]

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.

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