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Case Report

Aggressive Angiomyxoma with massive ascites ☆

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Introduction

Aggressive Angiomyxoma (AAM) is a rare soft tissue neoplasm which predominantly occurs at the female pelvic peritoneum and perineum region, with female-to-male ratio is 6.6:1(Chan et al., 2000). AAM is characterized as non-encapsulated and slow-growing benign tumor. However, their locally invasive and recurrent nature often leads to difficulties in clinical treatment. On the other side, misdiagnosis is also a problem because of their no-special clinical representations. So far, 310 papers were retrieved in Pubmed using search key word "Aggressive Angiomyxoma". However, most of these papers are case report because of their rarity, which is also responsible for the deficiency of diagnosis and treatment experience of this disease. In this paper, we present a case of small-sized AAM complicating with massive ascites which has not been reported before. We hope that the clinical procedure of this case would offer more useful informations on this disease.

Case report

A 30 years-old woman was transferred to our unit from department of gastroenterology for unexplainable massive ascites. The patient's abdomen circumference and body weight have been increasing gradually in the past 2 years. She has been admitted to department of gastroenterology in our hospital followed by detailed systemic examination including CT scanning. However, the source of ascites was not found. So laparoscopic exploration surgery was recommended. The patient had no history of hepatitis or tuberculosis before. Both HBV and HCV were negative in serum testing. Sonography and CT scanning showed no distinguishable tumor lesions.

Laparoscopic surgery was performed under general anesthesia. There was approximately 10,000 ml ascitic fluid within the abdominal cavity. Multiple intestine adhesions were observed. One tumor lesion was found on the surface of the uterorectal fossa peritoneum. The diameter of this tumor was 2 cm. There were also some other small lesions scattering on the surface of the peritoneum as shown in Fig. 1. These tumor lesions were all removed. The intra-operative frozen section result was necrotic adipose tissue. Postoperation pathology showed diamond-shaped or stellate tumor cells in loose interstitial tissues. The final pathological diagnosis was Aggressive Angiomyxoma as shown in Fig. 2. Immunohistochemical staining results showed: CD34(+), SMA(+), Desmin(+), Vimentin(+), mesothelial cell(+), ER(+), and PR(+). Positive CD34 signals were shown in Fig. 3.

The patient received further GnRH-a(Goserelin) treatment (3.6 mg i.h. per 28 days, 6 cycles totally) after surgery. GnRH-a was effective in controlling the ascites. However, GnRH-a was replaced by oral contraceptive drug(Marvelon) because of its heavy medical cost 6 months later. The ascitic fluid appeared again. Eventually, the patient received another surgery to remove all pelvic floor perineum tissues. The treatment was successful with no ascites appeared after the second operation. No additional therapy was used (GnRHa) since then and she was followed up with no evidence of recurrence.

Discussion

Aggressive Angiomyxoma (AAM) was first described by Steeper and Rosai at the 72th Meeting of Internal Academy of Pathology in 1983(Steeper and Rosai, 1983). AAM mainly occurs in the female vulvar and abdominal cavity. This disease has the nature of "being aggressive" which means that it has its locally infiltrative and recurrent characters. Patients are often asymptomatic or presents as valva tumors. Compression to urinary system or intestinal system occurs only when the tumor is large enough. The size of AAM varies greatly ranging from 1 cm to 60 cm based on reports. It is hard to diagnose AAM without pathologic evidence. Misdiagnosis rate is as high as 82% before surgery (Gungor et al., 2004). Imaging examinations including CT, MRI and pelvic DSA are essential for evaluating the extent of lesions. Signals of immunohistochemical examination are useful for diagnosis in some cases. However, its value is also limited because of the overlap with other diseases such as angiomyofibroblastoma and cellular angiofibroma. Almost all AAM show positive Vimentin and Desmin signals and negative S-100 signals. The positive rates of CD34, SMA, a-SMA,

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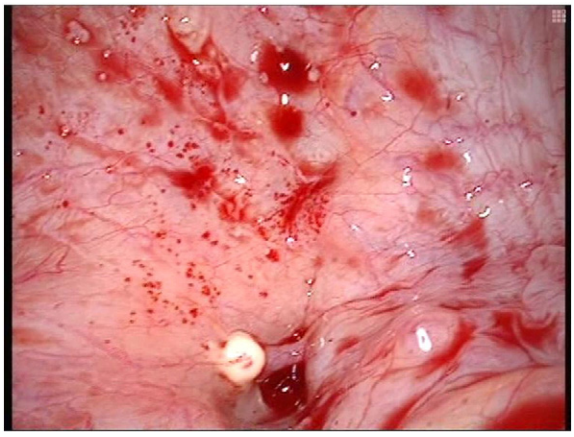

Fig. 1. Tumor lesions located on the surface of peritoneum.

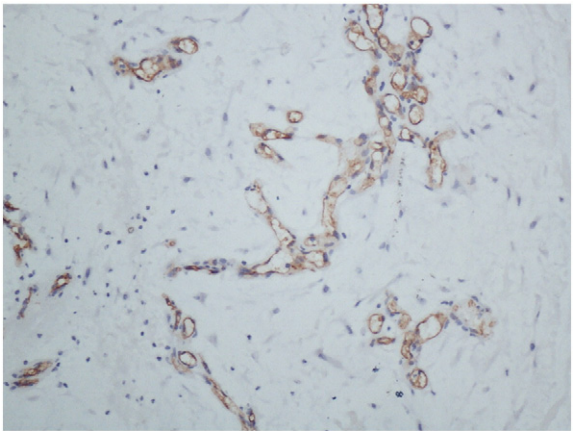

Fig. 3. Positive CD34 signals in the vascular endothelium cells.

PCNA, F8, ER or PR have been reported variously in other papers. In our case, immunohistochemical staining test results showed CD34(+), SMA (+), Desmin(+), Vimentin(+), mesothelial cell(+), ER(+) and PR(+).

AAM often occurs in the reproductive ages. Some reports revealed that the lesion growed during pregnancy, suggesting that hormones may play roles in its development and progression (Han-Geurts et al., 2006; Fishman et al., 1995). Fetsch et al (Fetsch et al., 1996) reported that the positive rates for ER and PR in AAM tissues were 93% and 90%, respectively. Havel et al reported positive ER and negative PR, while Htwe reported one case whose tumor growed rapidly during pregnancy with strong positive PR and negative ER (Havel et al., 1994; Htwe et al., 1995). So AAM is believed to be hormone-dependent tumor, which is the theory basis of hormone treatment.

The standard treatment procedure for AAM has not been established yet. In order to remove the lesions completely, wide local resection is the traditional choice. However, the recurrence rate is rather high because the tumor often infiltrates into surrounding fat and muscle tissues. For the tumors with positive ER and/or PR which could not be resected completely, hormone treatment is reasonable. Successful cases have been reported using GnRH agonist treatment against primary or recurrent tumors (McCluggage et al., 2006; Shinohara et al., 2004). In most of these cases GnRH agonist was used as postoperative adjuvant treatment. However, some authors reported the primary treatment of AAM with GnRH agonist without surgery or prior to surgery. Though the drug dose was almost the same, there was no consensus on the treatment cycles. Most of the cases received 6 cycles treatment. In our case GnRHa was also proved to be effective. For the possibility of recurrence, long-term follow-up is recommended.

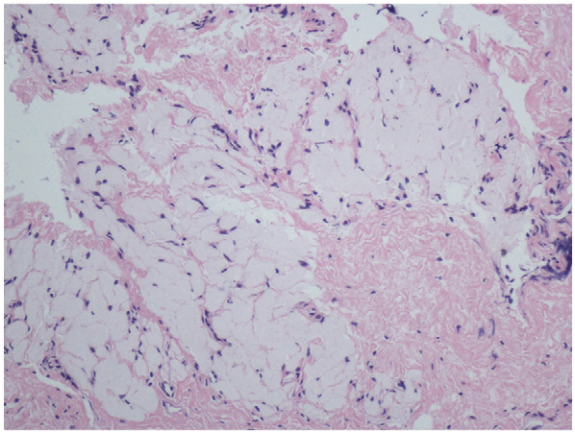

Fig. 2. Hematoxylin and Eosin staining of the tumor tissues.

This case was different from others. The patient presented unexplainable massive ascites without obvious lesions. Examinations of ascitic fluid and CT scanning found no evidence of tumor. Eventually, exploration surgery seems to be the last choice. During operation only a small tumor was found locating at the uterorectal fossa. The final diagnosis was AAM. And so, when a woman presents unexplainable ascites, the possibility of AAM should be considered despite of its rarity. Postoperative GnRH agonist treatment was successful in this case. However, it was very strange that is why such a small lesion could induce massive ascites. In this case, all other potential causes of ascites such as liver diseases, tuberculosis as well as malignant tumor were excluded. Anyway, the etiology of the ascites in this case of angiomyxoma was unknown.

Conclusion

In this case an AAM patient who presented unexplainable massive ascites was reported. The disease was treated successfully by surgery and postoperative GnRH agonist administration. Due to the rarity of this disease, more informations are still needed.

Conflict of interest statement

The author has no conflicts of interest to disclose.

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