

Case Report

Intrapelvic aggressive angiomyxoma with inferior vena cava involved

Guohua Yu^{1*}, Lingling Kong^{2*}, Guimei Qu¹, Xubo Pan¹, Jianglin Cong³, Jian Chen⁴

¹Department of Pathology, Affiliated Yantai Yuhuangding Hospital, Medical College of Qingdao University, Yantai, China; ²Department of Psychology, Binzhou Medical College Yantai, China; ³Department of Gynecology, Affiliated Yantai Yuhuangding Hospital, Medical College of Qingdao University, Yantai, China; ⁴Department of Central Laboratory, Affiliated Yantai Yuhuangding Hospital, Medical College of Qingdao University, Yantai, China. *Equal contributors.

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Abstract: Aggressive angiomyxoma is defined as a rare and benign mesenchymal tumor. Local infiltration, high risk of local recurrence and predominance in the female genital tract are typical features. Here we reported the second case of aggressive angiomyxoma with the inferior vena cava involved illustrating the unusual biological behavior of this tumor. A 37-year-old female presented initially with abdominal distention of six months' evolution. Imaging showed large mass in the pelvic cavity and hypo-dense shadow in the inferior vena cava. The tumors both in the inferior vena cava and in the pelvic cavity were excised by surgery and diagnosed pathologically as aggressive angiomyxoma.

Keywords: Aggressive angiomyxoma, pelvic cavity, neoplasm, inferior vena cava

Background

Aggressive angiomyxoma is a mesenchymal tumor found mainly in female. The clinical characteristics include locally aggressive, slow growth, recurrence and benign appearance in morphology without evidence of nuclear atypia or mitosis. However, this tumor involving the vena cava is unusual. According to our PubMed research, only one case of aggressive angiomyxoma of the pelvis with inferior vena cava involved has been reported [1]. In our present study, we report another case of intrapelvic aggressive angiomyxoma with inferior vena cava involved and clinicopathologic features are discussed.

Case presentation

A 37-year-old female presented with a six-month history of abdominal distention. During physical examination, some abnormal signs were found including abdominal protuberance and superficial vein dilation. Abdominal palpation was positive for hard neoplasm with obscure boundary and no pressing pain involving

the whole hypogastrium. Enhanced computed tomography scan demonstrated hypo-dense shadow in the inferior vena cava (**Figure 1A**) and pelvic cavity (**Figure 1B**). There were no other imaging findings in portal vein, hepatic vein, bilateral renal vein and deep veins of lower limbs. The chest X-Ray did not reveal any other abnormality. Her blood biochemistry was normal except lactic dehydrogenase (LDH) 447 IU/L, total protein 58.16 g/L and albumin 34.22 g/L. Complete blood counts, erythrocyte sedimentation rate, C-reactive protein, blood biochemistry, electrolytes, serum creatinine, urine analysis and the endocrine profile were within normal range.

The patient received surgical therapy under general anesthesia. Firstly, the tumor in the inferior vena cava was removed. During open thoracotomy, the inferior vena cava was opened to reveal the mass. By blunt and sharp dissection, the tumor was excised completely. Subsequently, exploratory laparotomy was made. Enormous and lobulated neoplasm was found in the whole pelvic cavity. The basal part of tumor was wide and originated from the

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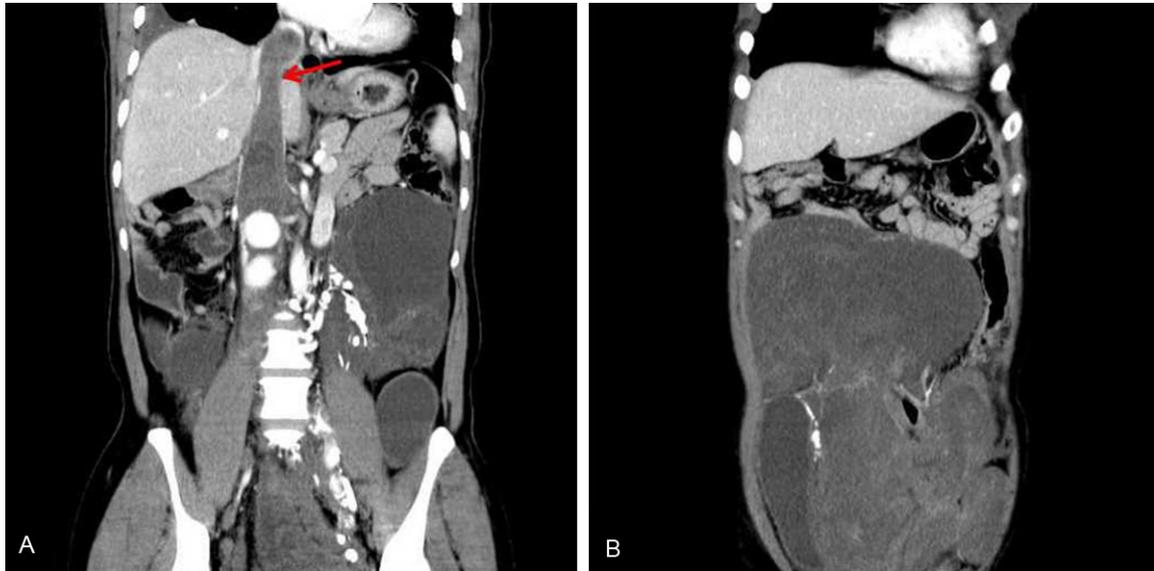


Figure 1. Imaging shows (A) hypo-dense shadow in inferior vena cava (arrow) and (B) in pelvis by enhanced computed tomography scan.

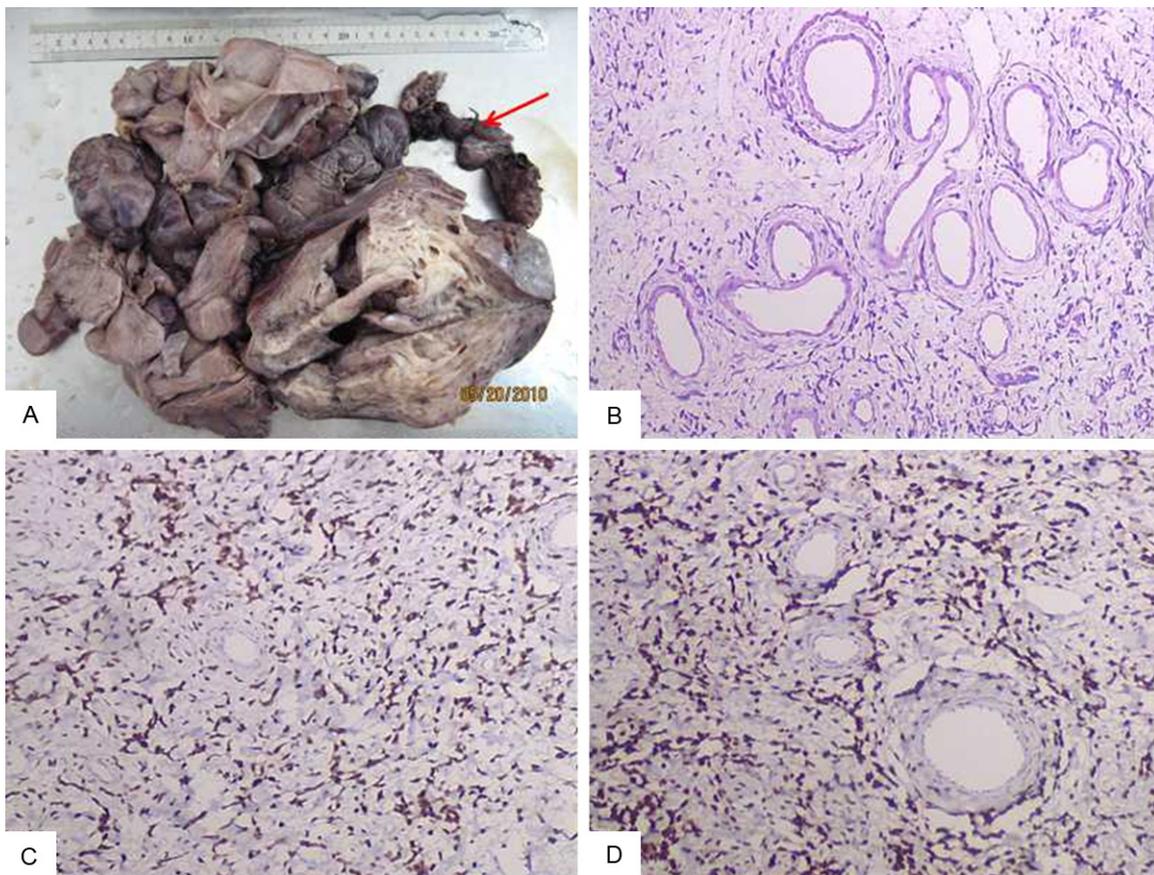


Figure 2. (A) Photograph shows the neoplasm is lobulated and cut surface gives a cystic or solid appearance. The arrow shows the tumor in the inferior vena cava. Photomicrograph shows (B) the tumor cells and numerous blood vessels are in a background of loose mucous matrix (Haematoxylin & eosin, $\times 40$); (C) ER positive cells on immunohistochemistry (Nuclear staining, $\times 40$); and (D) cells expressing PR on immunohistochemistry (Nuclear staining, $\times 40$).

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Table 1. Results of the immunohistochemical study

Antibody	Clone	Source	Dilution	Result
Anti smooth muscle actin	1A4	Dako	1:100	+
Anti ER	SP1	Lab Vision	1:100	++
Anti PR	SP2	Lab Vision	1:100	++
Anti Desmin	CD33	Lab Vision	1:100	+
Anti CD34	QBEnd/10	Lab Vision	1:100	Part of neoplastic cells are positive
Anti S100	Polyclonal	Lab Vision	1:100	-
Anti Ki67	MIB-1	Lab Vision	1:100	Labeling index is about 3%

Dako, Glostrup, Denmark; Lab Vision Corporation, Fremont, USA.

sacrococcygeal region. Little intestine was pushed aside towards the right upper quadrant. Descending colon, part of sigmoid colon, rectum, bladder and the right adnexa of the uterus were adhered. The tumor was dissected along the margin.

The intrapelvic neoplasm was lobulated, measured 32 cm×25 cm×15 cm and weight 1450 g totally. The tumor separated from inferior vena cava was about 15 cm in length and 1 cm to 3 cm in diameter. Cut surface gave a cystic or solid appearance, grey in color and rubbery in quality (**Figure 2A**). Under low power, the tumor cells were in a background of loose myxoid matrix. Numerous variable-sized, thin-walled and thick-walled blood vessels were distributed among the tumor cells (**Figure 2B**). Under high power, the tumor cells were spindle or stellate-shaped and had slender, eosinophilic cytoplasm and bland nuclei. No nuclear atypia and mitoses were found.

By immunohistochemical staining, the tumor cells were positive for estrogen receptor (ER), progesterone receptor (PR), desmin, smooth muscle actin (SMA) and were negative for S100 (**Figure 2C, 2D**). The Ki67 labeling index was approximate 3%. All the primary antibodies are listed in **Table 1**. The features of histology and immunohistochemistry of tumor cells from the inferior vena cava were similar with those from pelvic cavity. On the basis of histological features and immunohistochemical characters, the diagnosis of intrapelvic aggressive angiomyxoma with inferior vena cava involved was made. The patient did not receive further treatment and discharged from hospital after surgery. Three months after operation, imaging revealed no sign of recurrence and the patient was subsequently lost to follow-up.

Discussion

Aggressive angiomyxoma has been classified by the World Health Organization under “tumors of uncertain differentiation” and occur much more commonly in the reproductive women [2-4]. The reported ratio of female to male is approximately 6-7:1. Aggressive angiomyxoma generally involves female reproductive tract, such as pelvic cavity, vulvar region and perineum while extra-genital tract can also be found [5, 6]. The patient in our study was a young female who presented with not only a huge mass in the pelvic cavity but also the tumor in the inferior vena cava. There was no positive finding in other blood vessels. Associating with the discovery of Irene et al, our report illustrate the existence of this unusual behavior that aggressive angiomyxoma involves the inferior vena cava(1). Interestingly, two cases of metastatic aggressive angiomyxoma have been retrieved [7, 8]. One case described a 63-year-old woman suffered from aggressive angiomyxoma of the pelvis with massive bilateral pulmonary, mediastinal, aortic lymph node and peritoneal metastases, which led to the death in the end. Another case showed a 27-year-old woman with aggressive angiomyxoma of vulvar relapsed repeatedly. Nine years later, multiple lung metastases were detected and the patient died next year. Whether this so-called metastasis is true or organic invasion is caused by intravenous growth and extension? We would like to swing the pendulum towards the latter on the basis of theoretical hypothesis that the lung could be invaded by aggressive angiomyxoma via right atrium, right ventricle and pulmonary artery since the tumor could grow in the inferior vena cava and extend toward the right atrium. However, the evidence of intravenous growth is not mentioned in two above cases of metastat-

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ic aggressive angiomyxoma and the tumor even occurs in the lymph node. So it is too early to draw a conclusion. Much more cases and further research are needed in the future.

The diagnosis of aggressive angiomyxoma is regarded as presumptive when the typical morphological characteristics of spindle cells and numerous variable-sized, thin-walled or thick-walled blood vessels in a background of loose myxoid matrix are seen with light microscopy, as designated when additional stains such as estrogen receptor (ER), progesterone receptor (PR), Desmin and SMA are positive and S100 is negative. Angiomyofibroblastoma, which shares many histological features with aggressive angiomyxoma, is the most critical disease in distinguishing diagnoses [9, 10]. Angiomyofibroblastoma shows the presence of a sharply circumscribed border. This tumor involves thin blood vessels and plump epithelioid tumor cells are arranged in an observably perivascular distribution. Other histological differential diagnoses, including leiomyosarcoma, myxoid lipoma, myxomatous neurofibroma and myxoid leiomyoma should be cautiously considered and properly performed immunohistochemical staining could be helpful for a correct diagnosis.

The best treatment of aggressive angiomyxoma is surgical excision with wide free margins. The use of chemotherapy and radiotherapy is not particularly effective due to the low mitotic activity. However, hormone therapy was applied and suggested to be useful in the patients with recurrence or in whom surgical excision is not possible because of the expression of ER and PR [11]. Imaging examination is the preferred choice for detecting recurrences.

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Disclosure of conflict of interest

None.

Address correspondence to: Dr. Jian Chen, Department of Central Laboratory, Affiliated Yantai Yuhuangding Hospital, Medical College of Qingdao University, 20 Yuhuangding East Road, Yantai 264000, China. E-mail: ygh0536@163.com; Dr. Jianglin Cong, Department of Gynecology, Affiliated Yantai Yuhuangding Hospital, Medical College of Qingdao University, 20 Yuhuangding East Road, Yantai 264000, China. E-mail: yn702@163.com

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