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Case report

Giant retroperitoneal myxoma: A case report and literature review

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ABSTRACT

Introduction and importance: Myxoma is a benign tumor and is mesenchymal in origin. Myxomas of the retroperitoneum are extremely rare entities.

Case presentation: We here report a case of a 67-year-old male who presented with progressive abdominal distention for 3 years. Laboratory investigations revealed a reduction in erythrocytes, lymphocytes, hemoglobin, and an elevation in carbohydrate antigen 19-9. Imaging findings showed a multilocular cystic mass in the right abdomen with thin septa and internal calcifications. Laparotomy revealed that the mass had arisen from the retroperitoneum and the histological study suggested the diagnosis of myxoma.

Discussion: Myxoma features as a "cystic mass" in imaging studies. Therefore, the possibility of a cystic lymphangioma, cystic mesothelioma and myxoma should be considered when a multicystic lesion in the retroperitoneal space is observed. Due to the rarity of retroperitoneal myxomas and lack of specific manifestations and diagnostic methods, preoperative diagnosis is often delayed or incorrect. And until now, only a few cases of retroperitoneal myxoma have been reported.

Conclusion: The report will increase the understanding of the diagnosis and treatment of retroperitoneal myxomas. A brief review of the related literature was also carried out.

1. Introduction

Myxoma is a benign tumor and is mesenchymal in origin. It has been described in several locations, mainly in the heart, followed by bone, muscle, skin and subcutaneous soft tissues. However, it is rare to find myxoma in the retroperitoneal space. No detailed imaging report on myxoma of the retroperitoneal space. Here, we report a rare case of retroperitoneal myxoma in a 67-year-old male. Myxoma is most frequently diagnosed in patients in the fourth to seventh decades of life and has been reported to range in size from 1 to 17 cm [1]. However, in our patient, the longest diameter of the tumor was 21 cm, which was greater than the maximum size of myxomas reported in the literature. Due to a lack of clinical and imaging features, histological analysis is required for a definitive diagnosis. The initial criteria for the diagnosis of myxoma were established by Stout in 1948, and myxoma is defined as a true mesenchymal neoplasm composed of undifferentiated stellate cells in a myxoid stroma [2]. As myxomas are benign, surgical resection is recommended when present with significant clinical signs [3]. A review of the current literature was also provided to contextualize the findings of the present study.

2. Presentation of case

A 67-year-old man attended our hospital with progressive abdominal distention for 3 years. The patient had an abdominal mass found during examination, he received no treatment, and the size of the mass increased during the following three years, resulting in serious abdominal distention. He had a history of laparoscopic appendectomy and underwent surgery for a retroperitoneal tumor ten years earlier. The patient had no significant family history, no regular medications, no allergies, and no relevant genetic information, additionally, he had a smoking history of more than 40 years. His blood collection data were as follow: white blood cell was 9.4×10^9/L, neutrophil was 8.35×10^9/L (reference value:1.8–6.3), lymphocyte was 0.49×10^9/L (reference value:1.1–3.2), hemoglobin was 136 g/L, and there was an increase in carbohydrate antigen 19–9 (594.5 U/mL) and carcinoembryonic antigen (361.9 U/mL), other tumor markers are within normal limits.

CT scanning showed a large multilocular cystic mass (35 × 24 × 13 cm), which was well-circumscribed in its entirety. The mass extended from the epigastrium to the pelvis and almost filled the right abdominal and pelvic cavity. The majority of the mass was hypo-attenuation, with

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iso-density internal septations and calcifications visible inside the mass (Fig. 1). The lesion caused an extrinsic mass effect on adjacent structures. On magnetic resonance imaging (MRI), a cystic mass was found, which were hypointense and markedly hyperintense on T1- and T2-weighted imaging, respectively (Fig. 2A–B), with low signal septations showing intense enhancement on post-contrast images (Fig. 2C). The boundary between the lesion and lumbar spine was distinct, and no bony destruction was detected (Fig. 2D). Imaging findings were suggestive of a multi-cystic lesion in the retroperitoneal space and the possibility of a cystic lymphangioma and cystic mesothelioma or a rare retroperitoneal myxoma was considered.

Based on the assessment of a large mass with thin septa and multiple calcifications in the retroperitoneal space in an elderly male, a preoperative diagnosis of retroperitoneal lymphangioma was considered. However, histopathological examination was consistent with a low-grade myxoma, elements of atypia or mitotic activity were infrequent, and part of the lesion was of high grade (Fig. 3).

The surgery was performed by an associate chief physician whose professional career was more than 15 years. Intra-operatively, the mass was confirmed to have arisen from the retroperitoneum and complete excision was performed (Fig. 4). No obvious abnormalities in adjacent organs (liver, spleen, pancreas, stomach) were found during surgical exploration, and there was a clear thrombus in the inferior vena cava.

The postoperative follow-up was poor with lots of complications, unfortunately, the patient's general condition gradually worsened, and died due to septic shock.

2020 SCARE guidelines was used for the construction of this paper [4].

3. Clinical discussion

Our patient, a 67-year-old male, presented with abdominal distention. A benign lesion was suspected on clinical and imaging examinations due to the long disease duration and no obvious invasion of adjacent structures. CT and MRI scans were suggestive of a multi-cystic lesion and the possibility of a benign or low-grade malignant cystic lesion was considered. Macroscopically, the tumor was composed of stellate cells in the myxoid stroma, consistent with low-grade myxoma. A myxoma of the retroperitoneum in a male is a very rare clinical entity.

Myxomas are mucus-rich neoplasms that mirror the structure of the umbilical cord and do not show any other type of differentiation according to Virchow [5]. It is worth mentioning that myxoma can occur over a wide age range, and the majority occurs in the fifth decade of life, with a slight female predilection [6]. The most common location is the heart, followed by somatic soft tissues, and myxoma in the retroperitoneum is extremely rare with less than 10 cases reported in the literature [7–9].

The clinical manifestations range from no obvious abnormalities to symptoms caused by the mass effect, such as abdominal pain or distention. A slow-growing painless mass is the most common clinical sign. The patient reported herein did not present with obvious clinical symptoms at the early stage, which may have been due to no significant

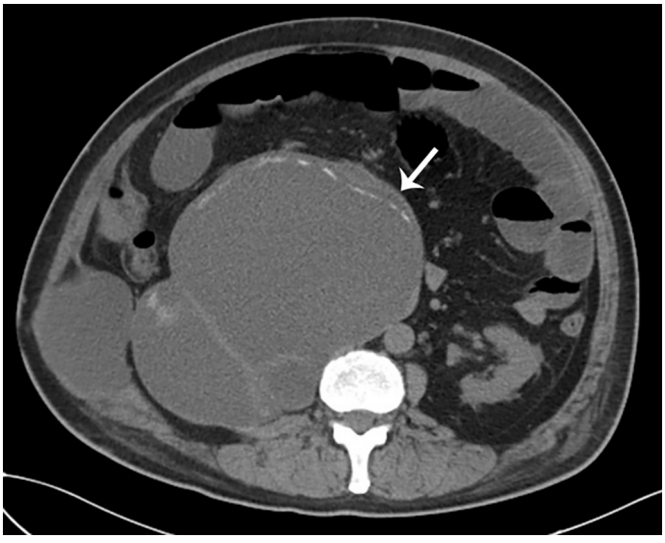

Fig. 1. CT findings. The isodensity internal septations and calcifications (white arrow) were visible inside the mass.

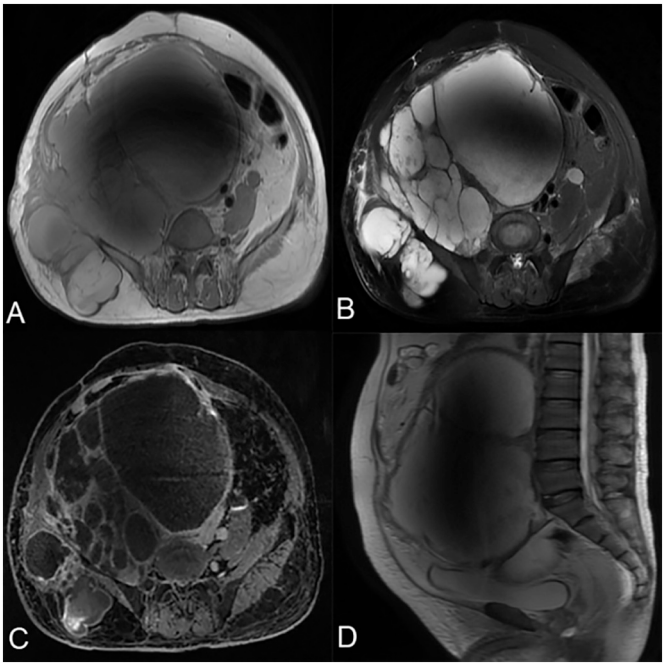

Fig. 2. MRI findings. (A–B): The lesion was predominantly hypointense and hyperintense on T1-and T2-weighted images, respectively. Axial T2-weighted imaging (B) also revealed the characteristic multi-loculated mass. (C): Axial post-contrast T1-weighted fat-suppressed images showed avid enhancement of the internal septations. (D): Sagittal T2-weighted images demonstrated that the boundary between the lesion and lumbar spine was distinct, and no bony destruction was found.

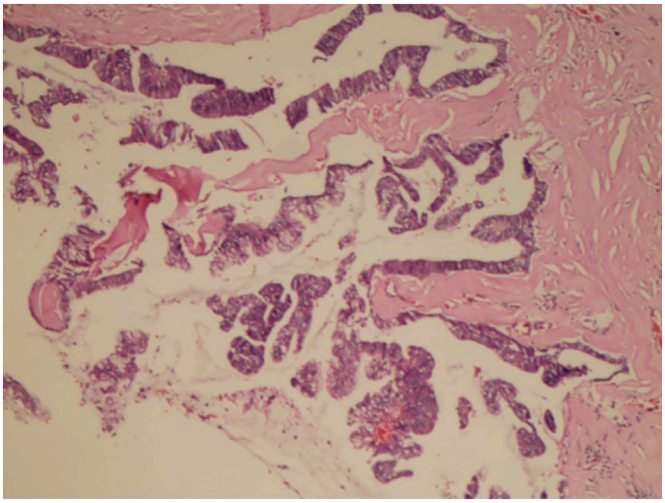

Fig. 3. Histopathological features. Histopathology of the resected specimen: The cells in the lesions were stellate-shaped with myxoid stroma, and elements of nuclear atypia or mitotic activity were infrequent.

Fig. 4. Surgical specimen: Gross appearance of a giant retroperitoneal myxoma.

mass effect on adjacent abdominal structures.

Additionally, this patient presented with multiple complications, such as pulmonary embolism and cerebral infarction. Patients with cardiac myxoma have been reported to present with thromboembolic symptoms [10]. Typically, embolization is a premier symptom of myxomas. Our patient developed a thrombus in the inferior vena cava, which may have been due to severe compression caused by the large tumor, resulting in reduced blood flow in the vessels. Under these conditions, emboli and thrombi form more easily and cause infarctions in many systems in the body. There is another possible explanation for this condition, it was reported that embolic events can be caused by a thrombus from the tumor surface or a myxomatous fragment of the tumor [11].

Carbohydrate antigen 19-9 (CA19-9) is a well-known tumor marker with little specificity, the elevation of which has been reported to be associated with several malignancies, mainly of hepatopancreatic biliary origin, and CA19-9 can be overexpressed in several benign gastrointestinal disorders [12]. In a reported case of an intramuscular myxoma, the level of serum CA19-9 increased preoperatively and returned to a normal level six months following surgery, Theodorou D et al. assumed that normal values were restored after resection may be the cause of an association between CA 19.9 and IM [13]. In a previous study, Jing JX et al. indicated that CEA and CA19-9 levels in patients with vascular embolism were significantly higher than those without vascular embolism [14]. Taking these into considerations, an embolus may be included as a possible reason for CA19-9 elevation when our patient presented with multiple embolisms.

Surgical resection-based comprehensive treatment is the optimal choice for a retroperitoneal tumor. Outcomes of the retroperitoneal tumor depend on its degree of malignancy and if radical resection can be adopted. In a case of myxoma with cerebral and pulmonary metastasis, after irradiation and five courses of chemotherapy with doxorubicin and ifosfamide, the metastases disappeared completely [15]. In addition, Vescini F et al. reported the use of bisphosphonate therapy providing a clear reduction in the diameter of myxoma after the use of zoledronic acid for 4 years [16].

Familiarity with radiologic features, in combination with clinical information, allows for more accurate diagnosis [17]. On CT, myxoma is often seen as a well-circumscribed hypo-attenuating lesion, or a lesion with uniformly low and high signal intensity on T1- and T2-weighted images, with variable heterogeneous enhancement. Sometimes, cystic spaces and multiple internal septa are also observed [5]. Imaging findings of the lesion in our case are consistent with the features mentioned above.

The differential diagnosis of retroperitoneal myxoma includes cystic lymphangioma, myxoid sarcoma, cystic mesothelioma, synovial cyst, bursa, and a ganglion. Generally, the size of the last three lesions is not as large as that in our patient; moreover, the most frequently affected sites are the musculoskeletal system, rather than the retroperitoneum. Cystic lymphangioma most frequently occurs in the head and neck. MRI reveals a well-circumscribed multi-cystic mass, with an intensity similar to that of “water density”, without internal enhancement, and wall calcifications are rarely seen. Cystic mesothelioma (CM) is a rare lesion that has a predilection for middle-aged women, when it occurs in the abdomen, it is more commonly found in the pelvic space rather than the retroperitoneum [18]. On MRI, CM typically appears as a hypointense and hyperintense cystic mass on T1- and T2-weighted images, respectively, with minimal to a mild peripheral wall and septal enhancement [19]. Myxoid liposarcoma is seen in younger populations, with the incidence peaking in the fourth to fifth decades of life. They most commonly occur in the lower extremities. The findings of high signal intensity (fat or hemorrhage) on T1-weighted images are helpful in the diagnosis.

4. Conclusion

The retroperitoneal space is an uncommon location for a myxoma. Women are aged 40–70 years are the most commonly affected population [2]. Since myxomas are benign, tend to infiltrate surrounding tissues, and do not give metastases, treatment should include wide resection of the involved structures with close follow-up control of the patient [20]. Chemoradiotherapy may be the possible option for the treatment of myxoma. Recurrence is the most common complication of myxomatous surgical excision which occurs in approximately 30% of cases at a median of 8.5 years [21]. Radiologists should be aware of differential diagnosis when identifying retroperitoneal cystic lesions, including cystic lymphangioma, cystic mesothelioma, and myxoma.

“Highlights”: It is the very first detailed imaging report on myxoma of the retroperitoneal space, and can also provide information on differential diagnosis and treatment of retroperitoneal myxoma.

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Ethical approval

No ethical approval necessary.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Research registration

Not applicable.

Guarantor

Lingling Song.

Provenance and peer review

Not commissioned, externally peer-reviewed.

Patient perspective

The procedure of intervention was explained to the patient with all findings and possible complications. He accepted the procedure and informed consent was given.

CRediT authorship contribution statement

Zhang ZZ designed the report, reviewed the literature, edited the images and wrote the paper.

Zhang ZZ and Song LL analyzed the data and revised the paper.

All the authors have read and approved the final manuscript.

Declaration of competing interest

The authors report no declaration of interest.

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