Primary Retroperitoneal Myxofibrosarcoma: a case report and review of the literature

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ABSTRACT

Myxofibrosarcoma is a neoplasm of fibroblastic origin. It commonly occurs in the extremities in the elderly, but rarely arises in the trunk, especially in the retroperitoneum. Herein, we report a rare case and the imaging findings of a primary retroperitoneal myxofibrosarcoma in a 69-year-old male patient without any significant past history who had complained of intermittent abdominal pain and constipation for two months. The imaging studies, including ultrasonography, X-ray of the kidneys, ureter, and bladder (KUB), abdominal computed tomography (CT), and 3.0-Tesla magnetic resonance imaging (MRI), revealed a large well-circumscribed mass lesion arising from the left side of the retroperitoneum. The lesion was filled with great amount of homogeneous fluid-like component without prominent calcification or a fat component. After surgical excision, a diagnosis of retroperitoneal myxofibrosarcoma was made. On the follow-up CT obtained ten months after surgery, a local recurrent tumor was observed. Although myxofibrosarcomas rarely arise from the retroperitoneum, the possibility of these tumors should be kept in mind in those patients who have a retroperitoneal lesion with imaging findings similar to that of the tumor.

CASE REPORT

A 69-year-old male patient, who was healthy previously without any history of major systemic disease, visited a physician for intermittent abdominal pain and constipation for two months. Physical examination revealed a protuberance in the left side of the abdomen without tenderness or rebound tenderness. Routine blood investigation was unremarkable. On abdominal ultrasonography, the normal left kidney was not seen. Instead, there was a huge mass lesion with a heterogeneous, relatively high echogenicity (Fig. 1). On an abdominal CT (Fig. 3), a well-defined homogeneous mass, measuring 22 × 22 × 15 cm³, was noted in the left side of the retroperitoneum. It blurred the left psoas muscle line and displaced the bowel loop to the right-hand side. After surgical excision, a diagnosis of retroperitoneal myxofibrosarcoma was made. On the follow-up CT obtained ten months after surgery, a local recurrent tumor was observed. Although myxofibrosarcomas rarely arise from the retroperitoneum, the possibility of these tumors should be kept in mind in those patients who have a retroperitoneal lesion with imaging findings similar to that of the tumor.
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The mass. The stomach, colon, small bowel, and left kidney were displaced by the lesion. On 3.0-Tesla MR imaging (Fig. 4), the mass presented T1-hypointense and T2-hyperintense signals with internal septation. Correlating to fat-suppressed T1-weighted MR imaging (Fig. 4b), there was no prominent macroscopic fat signal within the tumor. Although the lesion showed hyperintensity on T2-weighted MR imaging, there was a marked signal loss in the center of the image, probably due to a “standing wave artifact”, related to dielectric resonances at an ultrahigh field strength (3.0 Tesla and above) MRI. After contrast administration, the tumor showed a delayed and heterogeneous enhancement pattern (Fig. 4d-4f). The imaging findings were suggestive of a mass with suspicious myxoid stroma. The differential diagnoses included myxoid liposarcoma, myxofibrosarcoma, and neurogenic tumor.

Under the impression of a large retroperitoneal tumor, the patient received surgical treatment and the operation proceeded smoothly without immediate complications. Macroscopically, the tumor was tan-whitish and elastic with a multilobulated and gelatinous cut surface. Focal hemorrhage was observed. Microscopically, it was characterized by variable cellularity with myxoid stroma and numerous elongated curvilinear vessels (Fig. 5a). Focal increased cellularity of atypical fibroblastic cells with ill-defined, slightly eosinophilic cytoplasm and hyperchromatic nuclei were noted (Fig. 5b). Immunohistochemically, the tumor cells showed CD34 (-), S100 protein (-), and smooth muscle actin (-). The final diagnosis was myxofibrosarcoma, intermediate grade.

After the operation, the patient recovered well without any major complaints. On a follow-up CT obtained ten months after surgery, a local recurrent tumor was observed in the left iliacus muscle, confirmed by pathologic examination.

DISCUSSION

Primary retroperitoneal neoplasm

Primary retroperitoneal neoplasms are a relatively rare and diverse group of tumors including benign and malignant spectrums. CT and MRI can assist in recognizing the characteristics of a mass, but it is still a challenge for radiologists to diagnose these kinds of tumors. Nishino et al. described some important clues in the diagnosis of primary retroperitoneal neoplasms [6]. The first step is to decide whether the tumor is indeed located in the retroperitoneal space. Anterior displacement of retroperitoneal organs or major vessels could strongly suggest that tumors arise from the retroperitoneal space. Occasionally, the peritoneal organs are also displaced when the tumors are large, as in our patient. The second step is to exclude that the tumor has arisen from the organs of the retroperitoneal space. Some indirect signs could help to identify the origin of a tumor,
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Figure 3

Unenhanced computed tomography of abdomen, axial view a. and coronal view b. showed a well-defined homogenous mass lesion (asterisk), measuring 22x22x15 cm³, in the left-sided retroperitoneum. There was no obvious calcification, hemorrhage, or fat component within the mass lesion. The left kidney (white arrow) showed superior displacement.

Figure 4

3.0-Tesla abdominal MR imaging. a. Axial T1-weighted MR imaging demonstrated hypointensity of this mass lesion (asterisk in a to f). b. Axial T1-weighted fat-saturated MR imaging showed no prominent signal loss in this mass lesion. Thus, there was no macroscopic fat component within the mass lesion. c. On axial T2-weighted MR imaging, the lesion showed hyperintensity. There was marked signal loss in the center of this image probably due to standing wave artifact. On dynamic contrast-enhanced axial T1-weighted fat-saturated MR imagings obtained in the c. corticomedullary, e. nephrographic, and f. excretory phases, the lesion showed heterogeneous and delayed enhancement.
such as the “beak sign”, the “embedded organ sign”, etc. [7].
If a tumor arises from the retroperitoneal organs, it could
deform the edge of an adjacent organ into a beak shape
(beak sign). When part of an organ appears to be embedded
in the tumor (positive embedded organ sign), this also
suggests that the tumor has arisen from this organ. In our
case, no prominent beak sign nor positive embedded organ
sign could be identified, so the origin of the tumor may not
be a retroperitoneal organ. Besides, anterior and superior
displacement of the left kidney may hint that the mass arose
from the left side of the retroperitonium.

Some characteristic components of tumors could
provide clues and make differential diagnosis more specific,
especially if they are easily recognized on CT or MRI.
Fat is one of the characteristic components. It is easily
recognized due to a low attenuation on CT and hyperin-
tensity on T1-weighted MR imaging with signal loss in a
fat-suppressed image. Lipoma is a tumor consisting almost
entirely of fat and has a regular, well-defined border. In
contrast, if the tumor is irregular and ill-defined, liposar-
coma may be considered. Liposarcoma is the most common
sarcoma of the retroperitoneum and is classified into

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well-differentiated, pleomorphic, myxoid, and de-differentiated types [8, 9]. A well-differentiated liposarcoma often contains much fat, but de-differentiated types of liposarcoma may not. In our case, no obvious fat component was seen on CT and MRI, and therefore the diagnosis of lipoma was excluded. But, myxoid or de-differentiated types of liposarcoma could not be completely excluded.

Myxoid stroma appear markedly hyperintense in T2-weighted MR imaging and delayed contrast medium enhancement. A few tumors contain myxoid stroma, including neurogenic tumors (schwannoma, neurofibroma, ganglioneuroma, ganglioneuroblastoma, etc.), myxoid liposarcoma, and myxoid-fibrosarcoma. Relatively uncommon tumors containing myxoid stroma are desmoid tumors, leiomyoma, and leiomyosarcoma. Back to our case, the mass presented hyperintensity in T2-weighted images, and it was filled with myxoid stroma, which may be included in the list of differential diagnoses. As a result, neurogenic tumor, myxoidfibrosarcoma, desmoids tumor, and leiomyoma were considered.

Calcification is more easily demonstrated on CT than MRI. Retroperitoneal masses containing calcification include ganglioneuroma, hemangioma, neuroblastoma, osteosarcoma, teratoma, etc. Schwannoma, paraganglioma, myelolipoma, leiomyoma, leiomyosarcoma and hematoma uncommonly contain calcification. However, no prominent calcified component was recognized in the tumor in our patient.

Vascularity is an important feature to distinguish the nature of retroperitoneal masses. Hemangioma and paraganglioma are extremely hypervascular tumors. Myxofibrosarcoma, leiomyosarcoma and other sarcomas are moderately hypervascular. Low-grade liposarcoma, lymphoma, and many other benign tumors are of a hypovascular distribution [10]. In the dynamic T1-weighted MR imaging of our patient, the tumor showed heterogeneous and mildly delayed enhancement. Its vascularity was moderate or low.

In conclusion, the mass has no fat or calcification component, and it presented marked hyperintensity in T2-weighted MR imaging, with moderate or low vascularity. The list of differential diagnoses may include myxoid liposarcoma, myxofibrosarcoma, and some neurogenic tumor with atypical appearance.

**Myxofibrosarcoma**

Myxofibrosarcoma is one of the most common sarcomas in the elderly. It was formally termed as a myxoid variant of malignant fibrous histiocytoma by Enzinger and Weiss [1]. Angervall and Kinndblom classified it as a distinct entity and used the term “myxofibrosarcoma” to focus on its myxoid and fibroblastic elements [2]. Recently, myxofibrosarcoma was generally recognized as a distinct entity and included in the 2002 World Health Organization classification of tumors.

According to the review by Mansoor et al. [11], myxofibrosarcoma present in the extremities (82%), trunk (14.7%), head and neck (2.7%), and retroperitoneum. They are usually palpable, painless, and slow-growing. The clinical symptoms are not acute and usually progress slowly. In our case, the chief complaints included intra-abdominal dull pain and constipation. Tenderness or rebound tenderness
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were not found.

According to Kaya et al. [12], myxofibrosarcomas in the limbs have the tendency to show diffuse infiltration along the fascial planes (17 of 21 cases); all cases present a myxoid component, which is shown by hypointense-T1 and extra-hyperintense-T2 weighted signals. An increased perifascial T2 signal is characteristic in myxofibrosarcoma and multi-directional signal spreading is observed in cases of diffuse infiltration. Most of these myxofibrosarcomas have a heterogenous enhancement pattern (12 of 16 cases). Post-contrast studies and diffusion-weighted MR imaging may help to demonstrate the infiltration pattern.

In our case, the MRI findings were compatible with the findings described by Kaya et al. [12]. The tumor was well-circumscribed, arising from the left side of the retroperitoneum, and displaced the visceral organs such as the stomach, left transverse colon, and left kidney. It was hypointense-T1 and hyperintense-T2 weighted, and showed homogeneous signals. After contrast medium infusion, the tumor also showed heterogeneous enhancement. However, it was a local well-circumscribed tumor with no evidence of diffuse infiltration. Whether it is the main difference between myxofibrosarcoma in the limbs and that in the body or not, it needs further study in the future.

Surgical excision is recommended for myxofibrosarcoma. Due to the tendency towards infiltration, MRI study is important for surgical planning. Although primary myxofibrosarcoma of the retroperitoneum is not as invasive as that in the limbs, MRI still has advantages in terms of evaluation of tumors with soft tissue components.

Primary myxofibrosarcoma of the retroperitoneum is rare. Only 4 cases have been mentioned in a review article and 2 case reports [3-5]. Recurrence was reported in one case [4]. In our case, the patient received abdominal CT for follow-up after ten months. Intra-abdominal recurrence was confirmed. Considering the high recurrence rate of myxofibrosarcoma in the limbs, it should be assumed that there is a high possibility of recurrence, and close follow-up should be arranged for patients with this disease.

REFERENCES