Case Reports

RETROPERITONEAL FIBROSARCOMA WITH DISTINCTIVE CHARACTERISTICS ON GROWTH PATTERN AND FINDINGS IN DIAGNOSTIC IMAGING

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Abstract: A 47-year-old man with lumbago had been found to have a perirenal hematoma at the lower pole of the left kidney on CT. MR imaging also demonstrated a hematoma, 8×8×9cm in size. After 1-month observation, the mass had very rapidly increased in size, suggesting it to be a neoplasm. Consequent surgery resulted in non-curative tumor extirpation, because of invasion to the descending colon, left kidney and surrounding tissues. Histologically, the tumor consisted of uniform spindle-shaped cells with interlacing solid and bundle forms, showing a herring-bone pattern. Immunohistochemical studies finally led to the diagnosis of fibrosarcoma with markedly high malignant potential. Additional therapy was not performed. Rapid local spread of the tumor was demonstrated when bowel obstruction developed 2 months later. The patient died of the disease 3 months after surgery. No distant metastasis was observed throughout the clinical course.

Key words : retroperitoneal fibrosarcoma, herring-bone pattern, immunohistochemical studies

INTRODUCTION

Fibrosarcoma in the retroperitoneal space is a very rare malignant tumor. The treatment of this tumor requires prompt surgical extirpation. We encountered a case of retroperitoneal fibrosarcoma, which showed a distinct growth pattern and very rapid local spread without distant metastasis. Similar findings to hematoma in diagnostic imaging modalities led to a complicated preoperative diagnosis.

CASE REPORT

A 47-year-old man presented with a 2-week history of gradually increasing lumbago in the middle of March 2007. He had been found to have a perirenal hematoma at the lower pole of the left kidney on CT. At admission, he had tenderness in the left lower abdomen without a palpable tumor. Laboratory data showed anemia, hemoglobin of 10.4 g/dL, and C reactive

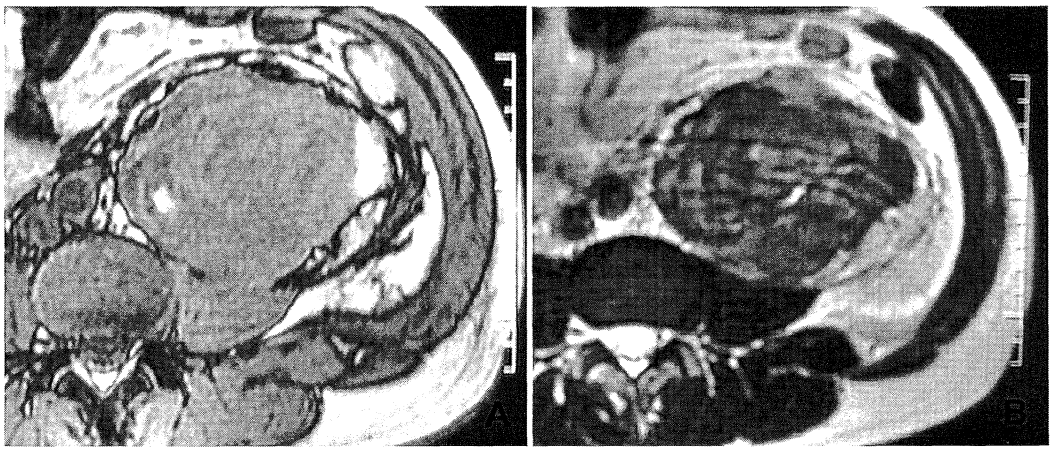

Fig. 1. MR imaging at the first admission.
A : Plain T1-weighted image, showing a perirenal mass with very high intensity sports within the mass is demonstrated. B : Plain T2-weighted image, showing scattered low intensity spots. There was no contrast enhancement inside the mass. These findings suggested that the mass was a hematoma with various points of bleeding in intermingled chronic and acute phases.

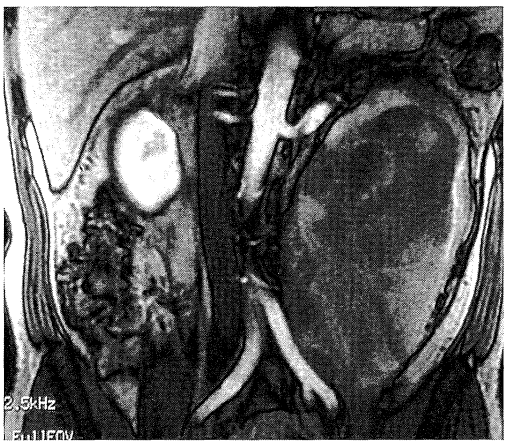

Fig. 2. MR imaging at the second admission.
The tumor had spread over the upper pole of the kidney, and recurrent intratumoral hemorrhage was suspected..

protein of 6.3 mg/dL. Hematuria was not observed. Plain MR imaging also demonstrated a perirenal mass with very high intensity spots inside in T1-weighted images, and scattered low intensity spots in T2-weighted images. No contrast enhancement was observed inside the mass. These findings suggested that the mass was a hematoma with various kinds of spots of bleeding in intermingled chronic and acute phases (Fig. 1). A neoplasm had to be ruled out. We explained the necessity of aingiography and subsequent surgical treatment. However, the patient left the hospital because of disappearance of the symptom and the patient's personal reasons. During the hospitalization, anemia did not progress, and growth

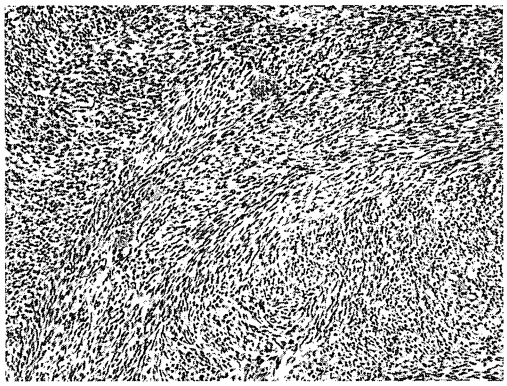

Fig.3. Microphotograph of the resected specimen.(H-E stain)
A herring-bone pattern : the tumor consisted of uniform spindle-shaped cells with interlacing solid and bundle forms.

of the tumor was not observed on CT. At re-admission one month later, he had a painful, palpable mass in the left lower abdomen. In MR imaging, the tumor had unexpectedly spread over the upper pole of the kidney, and a recurred intra-tumor hemorrhage was suspected (Fig. 2). Distant metastasis was not detected. Subsequently performed left renal arteriography did not demonstrate active bleeding, vascular malformation, or tumor feeding vessels. Considering a possible neoplasm, tumor extirpation was attempted with a transperitoneal approach.

The red-brown tumor accompanied with partial necrosis and hemorrhage was non-capsulated and fragile, involving the descending colon, left kidney and surrounding tissues. Although extended resection including these organs was attempted, curative surgery was not achieved. The external and communicating iliac lymph nodes were enlarged.

Histologically, the tumor consisted of uniform spindle-shaped cells with interlacing solid and bundle forms, showing a herring-bone pattern (Fig. 3). The results of immunohistochemical studies finally led to the diagnosis as fibrosarcoma: positive only for vimentin and negative for CK7, wide keratin, S-100, CD34, CD68, CD99, EMA, cytokeratin and calretinin. The additional findings indicated that the tumor had extremely high malignant potential: an increase in cell density and mitotic figures, scarcity of collagen, accompanying necrosis, and 70-80% of MIB-1 index. Metastasis was not observed in the lymph node specimens.

Postoperatively, we informed the patient and his family that: 1) curative surgery was not achieved, 2) the tumor seemed to have extremely high malignant potential, 3) the previously reported response rates of chemotherapy for the disease were not high, and 4) no effective therapy for the residual lesion had been established. As a result, no additional treatment was performed according to their decision. Two months postoperatively, the patient returned to the hospital due to bowel obstructive symptoms of emesis and abdominal pain. MR imaging demonstrated the extremely rapid spread of local tumor lesion. The patient died of the disease 3 months postoperatively. Metastasis of distant organs and lymph nodes were not observed until death.

Table 1. Results of immunnhisto chemical staining for retroperitoneal fibrosarooma : The Jaoanese literature

No	Reported year	Results		Source
		Positive	Negative	
1	1996	Vimentin	S100	Arata, et al. : Nishinihon J Urol 58: 1111
2	1997	Vimentin	S100, NSE, Actin, Myosin, Desmin	Takei, et al.: Acta Urol Jpn 43: 487
3	1998	Vimentin	none	Nemoto, et al.: Jpn J Clin Urol 52: 323
4	2001	none	S100, CD34, Actin, Desmin	Asano, et al.: Obstet Gyne Practice: 50: 1265
5	2001	Vimentin	S100, Cytokeratin	Ogata, et al.: Jpn J Urol Surg 14: 1199
6	2004	Vimentin	S100, Actin, Desmin, HHF-35	Takeuchi, et al.: Acta Urol Jpn 50: 525
7	2006	Vimentin	S100, Desmin, α-SMA	Kakuta, et al.: Acta Urol Jpn 52: 271
8	2009	Vimentin	S100, CK7, Wide keratin, EMA, Cytokeratin, CD34, CD68, CD99, EMA, Calretinin	Our case

DISCUSSION

Malignant tumors account for 70 to 80% of retroperitoneal tumors.[1] Fibrosarcoma is rare among malignant soft tissue tumors. Only 4.6% of fibrosarcomas developed in the retroperitoneal space.[2] The disease-related symptoms are an abdominal mass, weight loss, abdominal pain and fullness, which are caused by enlargement of the tumor.[3] Our case presented with lumbago. Thus, the delayed development of subjective symptoms is one of the reasons for the poor prognosis of this disease.

Among diagnostic imaging modalities, MR imaging would be useful for the diagnosis of fibrosarcoma.[4] However, there were no reports in the Japanese literature in which MR imaging led to the accurate preoperative diagnosis of fibrosarcoma, because the findings vary according to the tumor differentiation, and the degree of degeneration, necrosis and hemorrhage.[3] In our case, in fact, the findings of MR imaging suggested a hematoma at the initial presentation and even at re-admission when marked spread of the tumor was observed.

The final diagnosis should be reached by histological examinations. In hematoxy-eosin staining, fibrosarcoma often shows a herring-bone pattern: Uniform spindle-shaped cells are arranged in interlacing solid and bundle forms. However, it is well known that it is very difficult to diagnose soft tissue tumors only by a routine histological examination. In the past two decades, immunohistochemical studies have been playing a role to assist the differential diagnosis. Takeuchi et al.[5] accumulated 37 cases of retroperitoneal fibrosarcoma in the Japanese literature. We collected 8 reports including our case, which were examined immunohistochemically for the final diagnosis (Table 1). The only positive finding was obtained with vimentin as well as that in our case. For the differential diagnosis, solitary fibrous tumor, malignant fibrous histiocytoma and synovical sarcoma were on the list. In our case, CD34 was negative, suggesting solitary fibrous tumor was not the diagnosis because 90% of solitary fibrous tumor is positive for CD34.[6] Being negative for both CK7 and wide keratin withdrew synovical sarcoma from the list.[7] Although it was very difficult to differentiate malignant fibrous histiocytoma from fibrosarcoma, the herring-bone pattern

observed in histology supported our diagnosis. The following findings indicated this case to have extremely high malignant potential: the increased cell density, frequent mitotic figures, scarcity of collagen, existence of necrosis, and high MIB-index of 70 to 80%.[8]

The first line treatment of fibrosarcoma would be surgical extirpation. In many cases as well as our case, however, the tumors have often become huge and have invaded to the surrounding tissues at the time of presentation. In addition, the tumor often shows a resinoid growth pattern without a capsule.[3] Thus, it has been reported that approximately 50% of fibrosarcoma resulted in non-curative surgery,[4,9] and that the recurrence rate was 70% even after curative surgery.[9]

The present case showed extremely rapid growth and high invasion potential. However, no sign of metastasis was detected until death. Although metastasis has been recognized to be rare in fibrosarcomas, the prognosis of fibrosarcoma is poor: the 5-year survival rate is around 20%.[4] This fact indicates that therapies other than surgery are not effective. Surgery, radiation therapy, and chemotherapy are treatment options, but the most important factor in the treatment of primary tumors is complete surgical resection.[10]

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