An Effective Chemotherapy Regimen for Liver Metastasis from Retroperitoneal Fibrosarcoma: Report of a Case

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Abstract: Retroperitoneal fibrosarcoma is a rare disease that has proven difficult to treat due to its high incidence of postoperative local recurrence. We recently experienced a patient in whom retroperitoneal fibrosarcoma was followed by liver metastasis without local recurrence. A 34-year-old woman who initially presented with right upper quadrant pain was found to have a retroperitoneal tumor by diagnostic imaging techniques. Extirpation of the tumor was performed and the histopathological diagnosis was fibrosarcoma. A solitary metastasis was detected in the lateral segment 1 year after this operation and a lateral segmentectomy was carried out; however, a short time later, multiple liver metastases were found. Initially, ethanol injections were given with little effect, following which CYVADIC chemotherapy, consisting of cyclophosphamide, vincristine, farmorubicin, and dacarbazine was administered. An excellent responsiveness without severe toxicity was achieved after five cycles, with a significant reduction in tumor size, being estimated as a complete response. Thus, we consider that this chemotherapy regimen could be a promising mode of treatment for liver metastasis from retroperitoneal fibrosarcoma without local recurrence.

Key Words: retroperitoneal tumor, fibrosarcoma, liver metastasis, chemotherapy

Introduction

Retroperitoneal soft-tissue sarcomas account for 15% of all total soft-tissue sarcomas, among which fibrosarcoma is rare, accounting for just 5%, compared with liposarcoma or leiomyosarcoma, which account for 50% and 25%, respectively. A high local recurrence rate ranging from 40% to 82% has been reported in all series of retroperitoneal sarcoma, while the most common sites of distant metastases are the lungs (6%) and liver (9%).

CYVADIC is an effective combination chemotherapy in the treatment of advanced sarcomas, and consists of cyclophosphamide, vincristine, farmorubicin, and dacarbazine. Pooled data have shown that the response rate for metastatic sarcoma ranges from 15% to 60%; however, liver metastasis is considered less responsive than pulmonary metastasis.

We report herein an unusual case of metachronous liver metastasis without local recurrence from retroperitoneal fibrosarcoma. We considered that treatment of the liver metastasis was critical to the patient's prognosis and administered CYVADIC chemotherapy, achieving excellent results after five cycles.

Case Report

A 34-year-old woman presented to our hospital with right upper quadrant pain. Her past medical history and family history were not significant. Physical examination revealed a hard mass in the right upper quadrant, but hematological examinations and blood chemistry showed no abnormal values, including those of the tumor markers. Computed tomography (CT) scan of the abdomen demonstrated a large mass, 4 x 3 cm, in the right upper quadrant, the internal density of which was not homogeneous, and whose margin was clearly distinguishable from the surrounding tissue. A T1-weighted magnetic resonance imaging (MRI) scan of the sagittal section showed a mass located close to the lower surface of the liver and the anterior surface of the right kidney (Fig. 1). No metastatic mass was found in the liver. Abdominal angiography showed a hypovascular round area with no tumor vessels.

A laparotomy was performed under the diagnosis of a retroperitoneal tumor. The tumor had a fibrous capsule and was easily separated from the surrounding tissue.
except for an area of adhesion to the lateral wall of the inferior vena cava (IVC), which was resected using a side-clamping technique and repaired by direct suturing with 5-0 prolene. The surgical specimen was an elastic-hard, well-circumscribed tumor, the cut surface of which was white and solid with necrosis and hemorrhage (Fig. 2). Microscopic examination revealed that it was mainly composed of pleomorphic fibroblast-like spindle cells focally arranged in a storiform pattern and composed of oval to round histiocyte-like cells, admixed with abundant cytoplasm and bizarre nuclei containing prominent nucleoli; confirming a diagnosis of fibrosarcoma (Fig. 3). The surgical margin proved to be tumor-free, including the resected IVC wall. However, 1 year after the operation, a solitary liver mass was detected in the lateral segment and a lateral segmentectomy was performed. Histologic examination confirmed this tumor to be a metastasis. A MRI scan 3 months after hepatectomy revealed further liver metastases (Fig. 4a). At first we attempted treatment with ethanol injections for these lesions in place of arterial embolization because of the poor vascularity, but this proved ineffective. Next we administered systemic chemotherapy, in the form of CYVADIC, each course of which consisted of 400 mg cyclophosphamide (CPM), 1 mg vincristine (VCR), 40 mg farmorubicin (FAM), and 200 mg dacarbazine (DTIC). This was repeated five times at monthly intervals. After five cycles, the liver metastases were found to have reduced in size remarkably, resulting in a complete response (Fig. 4b). During therapy, the patient complained only of slight nausea, and the laboratory data, including leukocytes, platelets, and liver function tests, showed normal values. Diagnostic imaging tests revealed no evidence of local recurrence throughout the course.

**Discussion**

The overall 5-year survival rate for patients with retroperitoneal sarcoma ranges from 15% to 35%, which