An Extrarenal Malignant Rhabdoid Tumor Suspected to Originate from the Mesentery in an Adult: Report of a Case

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Abstract
A malignant rhabdoid tumor is very rare and its prognosis is extremely poor. It was first described as a distinctive and highly malignant neoplasm of the infant kidney. Tumors with a similar appearance have been reported in various extrarenal sites. We herein report a case of a 41-year-old man who was admitted to our hospital complaining of a lower abdominal mass. After one series of examinations, the patient underwent a laparotomy. Most such tumors are situated in the mesentery and involve the small intestine, and thus we diagnosed it to originate from mesentery. This tumor could not be resected and only an excisional biopsy was done. It was histologically composed of a solid sheet arrangement with ovoid, round, and lateralized nuclei and mild acidophilic cytoplasm. Inclusion body-like structures were found in the cytoplasm. Immunohistochemically, the tumor cells were positive for cytokeratin, epithelial membrane antigen, vimentin, and CAM5.2. The patient died 2 weeks after operation due to multiple organ failure.

Key words Rhabdoid tumor · Mesentery neoplasm · Small intestine · Immunohistochemistry

Introduction
A malignant rhabdoid tumor was first described in 1978 by members of the Wilms' tumor study group. It was originally described as a rare renal sarcoma of the infant kidney. However, tumors with a similar histological and clinical appearance have been reported at various extrarenal sites. Malignant extrarenal rhabdoid tumors are both morphologically and biologically homologous to malignant renal rhabdoid tumors. The clinical behavior of malignant rhabdoid tumors showed an extremely poor prognosis with a high potential for extensive metastases to various organs. In the same way, just like malignant rhabdoid tumors, the course of malignant extrarenal rhabdoid tumors is short and its prognosis is very poor.

Histologically, the tumor cells are highly cellular, consisting of monomorphic round or polygonal cells arranged in solid sheets or in alveolar or trabecular patterns with abundant eosinophilic cytoplasm containing globular hyaline-like inclusions. Immunohistochemical staining for vimentin, cytokeratin, and cytoplasmic epithelial membrane antigen (EMA) has generally revealed positive results. The histogenesis of malignant rhabdoid tumors still remains uncertain.

Case Report
A 41-year-old man who presented with 10-day history of lower abdominal distension and mass with no pain was admitted our hospital. The patient complained of slight nausea and a loss of appetite. The physical examination revealed a child-head-sized tumor with elastic hardness and mild tenderness in the lower abdomen. The body temperature of the patient was 37.2°C. He demonstrated mild anemia but did not notice any tarry stool. A complete blood cell count showed a red cell count of $380 \times 10^4$ mm$^{-3}$, a hemoglobin of 11.0 g/dl, a leukocyte count of 11 100/mm$^3$, and a platelet count of 34.0 $\times 10^4$ mm$^3$. Lactate dehydrogenase was elevated at 716 IU/l. C-Reactive protein was elevated at 9.6 mg/dl. Hypoalbuminemia with 3.0 g/dl was noted. Soluble interleukin-2 receptor was slightly elevated at 620 U/ml. All tumor markers examined, namely, carcino-
embryonic antigen, carbohydrate antigen 19-9, squamous cell carcinoma antigen, and α-fetoprotein were all within the normal limits. Interleukin-2 receptor was slightly elevated at 620 U/ml. The remaining laboratory findings were all within the normal limits.

Ultrasonography revealed an isoechoic mass measuring 7.0 × 6.0 cm containing linear high-echoic parts in the lower abdomen. The tumor was unclear concerning continuity with small intestines. Abdominal enhanced computed tomography revealed a large mass with a high-density wall and necrotic center in the lower abdomen (Fig. 1a). Magnetic resonance imaging (MRI) revealed a solid mass with a central hollow in the whole lower abdomen (Fig. 1b). Contrast enema with barium showed extrapolmonary oppression of the ascending colon and sigmoid colon due to the tumor. Gallium-67 scintigraphy revealed an uptake image in the lower abdomen consistent with a tumor.

After one series of examinations, we established a preoperative diagnosis of malignant lymphoma of the small intestine. The patient underwent a laparotomy. The tumor was found in a lump of the mesentery and also invaded the small intestine from the jejunum to ileum. A greater part of the tumor occupied the mesentery than the small intestine. The lumen of the small intestine remained open. In these operative observations, we determined that the tumor originated from the mesentery rather than from the small intestine. A resection of the tumor en bloc was judged to be impossible and only an excisional biopsy was performed. The cut surface bled easily. The tumor was histologically composed of a solid sheet arrangement, medullary proliferation, and trabecular formation. The tumor cells revealed such findings as ovoid or round nuclei, prominent nuclei, lateralized nuclei, and mild acidophilic cytoplasm. Inclusion body-like structures were found in the cytoplasm of some tumor cells (Fig. 2). Tumor cells were immunohistochemically positive for cytokeratin, EMA, vimentin, and cytokeratin marker (CAM5.2) (Fig. 3). They were negative for leukocytic common antigen, B-lineage antibody (L26), T-lineage ligand (UCHL-1), CD79α, neuron-specific enolase, keratin, myoglobin, chromogranin, and biclonal anticytokeratin antibody (AE1/AE3).

The patient’s condition worsened due to ileus and peritonitis probably causing a minor perforation of the small intestine due to tumor necrosis, and died 2 weeks after the operation due to multiple organ failure. No autopsy was performed.