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Case Report

A Case of Primary Malignant Fibrous Histiocytoma of the Duodenum

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ABSTRACT

We report a case of primary malignant fibrous histiocytoma of the duodenum. A 53-year-old man was admitted to our hospital with chief complaints of epigastralgia and fever of several days' duration. Radiologic and endoscopic examinations revealed a duodenal neoplasm metastatic to the lung. Histopathologic examination of biopsy specimens revealed storiform pleomorphic malignant fibrous histiocytoma. As surgical resection was impossible, multiagent chemotherapy was administered (cisplatin, 100 mg/m² ; doxorubicin, 60 mg/m² × 2 cycles) ; however, treatment was unsuccessful, and the patient died 5 months after admission. To our knowledge, the present case of primary malignant fibrous histiocytoma is the first to be reported in Japan and the third ever reported. (Jikeikai Med J 2005 ; 52 : 55-8)

Key words : malignant fibrous histiocytoma, duodenal neoplasm

INTRODUCTION

Malignant fibrous histiocytoma (MFH) is the most common soft-tissue sarcoma in men 60 years or older[1,2] ; however, primary MFH rarely occurs in the alimentary tract. Approximately 20 cases of MFH of the stomach[3,4] or other parts of the alimentary tract[5-9] have been reported. However, a search of the literature yielded only 2 cases of primary MFH of the duodenum[8,9]. We describe what we believe to be the first case of MFH of the duodenum to be reported in Japan, including results of immunopathologic analysis of an endoscopic biopsy specimen. MFH of the digestive tract is generally treated with excision, chemotherapy, and radiotherapy ; however, because early detection of this tumor is difficult, excision tends to be incomplete, and metastasis and local recurrence result in a poor prognosis.

CASE REPORT

A 53-year-old man was admitted to our hospital with chief complaints of epigastralgia and high fever of several days duration. He had been well until 1 month previously, when he noted dark stools, unaccompanied by other symptoms. No significant past history was elicited. On abdominal examination, muscular rigidity was noted. Laboratory studies revealed severe anemia (hemoglobin, 4.8 g/dl) and were suggestive of inflammation (C-reactive protein, 8.41 mg/dl). Serum levels of carcinoembryonic antigen were elevated at 12.8 ng/ml, but CA19-9 was normal. Free air was noted under the diaphragm on abdominal X-ray films. These findings suggested perforative peritonitis.

Computed tomography (CT, Fig. 1) revealed a 4-cm-diameter duodenal mass displacing the head of the pancreas, with no evidence of metastasis to the liver. No dilation of the biliary or pancreatic ductal

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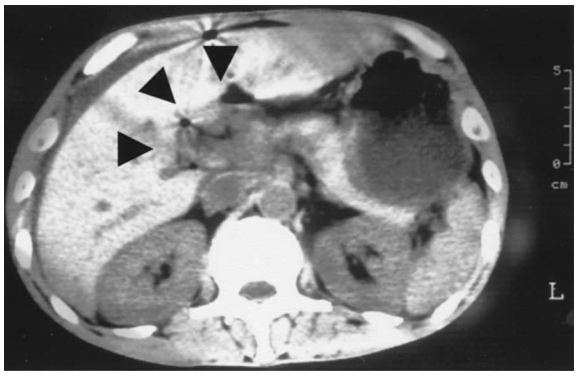

Fig. 1. Abdominal CT : A low-density mass 4 cm in diameter is visualized in the lower region of the uncinate process of the pancreas.

system was seen. Chest X-ray films revealed a 3-cm-diameter area of opacity in the upper lobe of the left lung. Endoscopy demonstrated a firm, elastic, hemorrhagic tumor measuring 4 cm in diameter arising from the duodenum, at the anal side of the papilla of Vater (Fig. 2). The perforation could not be visualized but was suspected to be near the tumor. Because CT confirmed that the pancreas and pancreatic ductal system were normal and because the tumor was seen to arise from the duodenal wall rather than being extramural, a diagnosis of duodenal neoplasm with pulmonary metastasis was made.

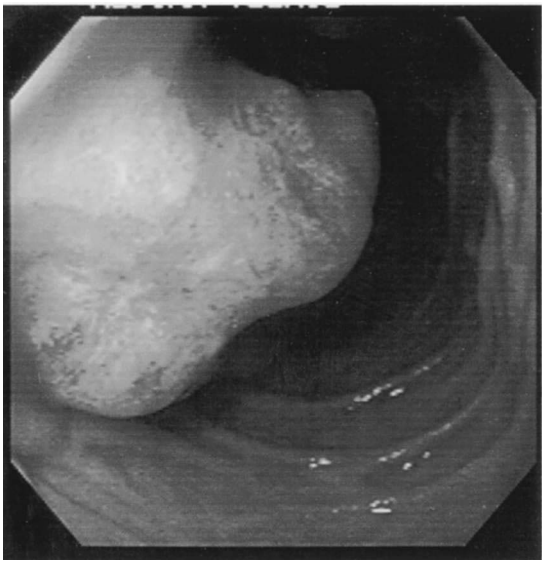

Fig. 2. Endoscopy : A firm, elastic, hemorrhagic 4-cm-diameter tumor occupies the anal side of the duodenum beyond the papilla of Vater.

Histopathologic examination of the biopsy specimen revealed a nonepithelial sarcomatous lesion with pleomorphic tumor cells tightly packed within sparse mesenchymal tissue. Tumor cells had clear cytoplasm, and irregular nuclei with multinuclear and giant nuclear formation were evident. Nucleoli were distinct, and nuclear divisions were observed in many tumor cells (Fig. 3-A). Immunostaining was performed with an automated immunostainer (NX System, Ventana, Tucson, AZ, USA) and a Basic DAB Detection Kit (# 760-001, Ventana). Tumor cells showed intense and extensive immunoreactivity for vimentin (Fig. 3-B), α1- antitrypsin (α1-AT) (Fig. 3-C), and α1-antichymotrypsin (α1-ACT) (Fig. 3-D), whereas reactions were negative for periodic acid-Schiff, leucocyte common antigen, UCHL-1, L-26, keratin, Grimelius stain, chromogranin A, S-100 protein, synaptophysin, insulin, somatostatin, glucagons, and Dupan-II. On the basis of these results, a histopathologic diagnosis of storiform pleomorphic MFH was made.

Treatment for perforative peritonitis was started, with antibiotics and total parenteral nutrition being administered and aspiration performed via an indwelling nasogastric tube. This treatment improved the patient's general condition. As complete excision of the tumor was impossible, chemotherapy was started with cisplatin, 100 mg/m^2, and doxorubicin, 60 mg/m^2×2 cycles ; however, this treatment was not effective, and the patient died 5 months after admission. An autopsy was not performed.

DISCUSSION

MFH was defined by Weiss and Enzinger in 1978 as a neoplasm composed of spindle-shaped (fibroblast-like) and rounded (histiocyte-like) cells accompanied by pleomorphic giant cells and inflammatory cells[2,10]. MFH can be classified into storiform, pleomorphic, fibrous, inflammatory, giant cell, mixoid, and angiomatoid types, all of which are positive for α1-AT and α1-ACT[11]. To our knowledge, specific serum tumor markers for MFH have not been reported, despite the elevation of carcinoembryonic antigen in the present case.

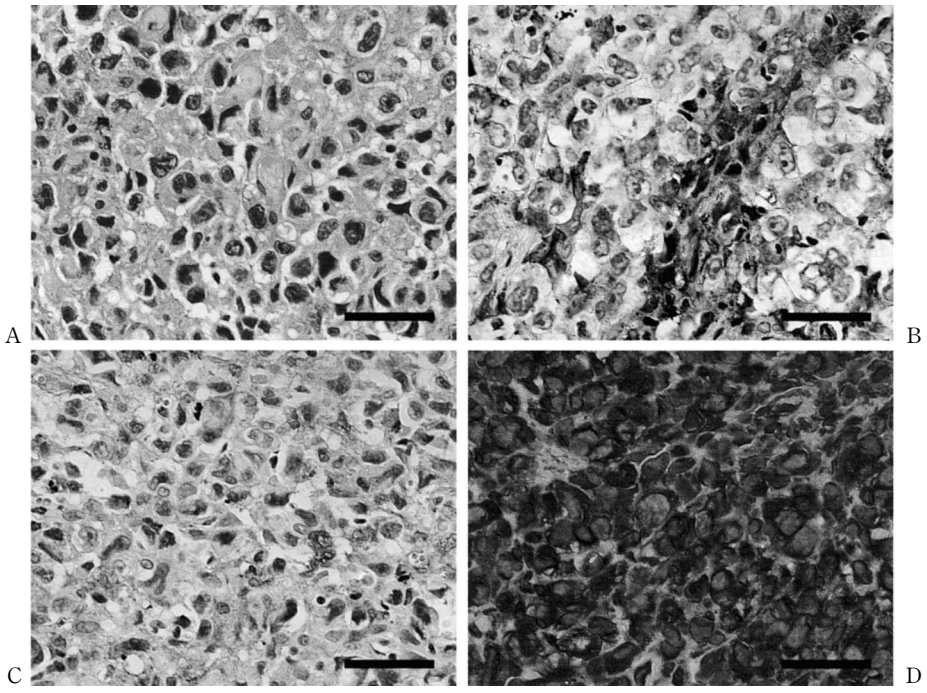

Fig. 3. A. Hematoxylin-eosin staining : Nonepithelial sarcomatous lesion. Tightly packed pleomorphic giant cells are observed, within sparse mesenchymal tissue. Tumor cells exhibit clear cytoplasm and irregular nuclear formations, with multinuclei and giant nuclei. Nucleoli are distinct, with nuclear fission evident in many tumor cells. B. Vimentin : Immunoreactivity is strong in neoplastic cells and mesenchymal tissue and slight in blood vessels. C. α1-AT : Neoplastic cells exhibit staining, particularly of the cytoplasm. D. α1-ACT : Neoplastic cells show slight immunoreactivity. Bar=50 μm.

Surgical excision is the treatment of choice in MFH, but local recurrence and distant metastases frequently occur. Weiss and Enzinger have reported that in cases with incomplete excision, the rate of local tumor recurrence is 44% and the rate of metastasis is 42%[2]. Metastasis occurs most frequently to the lung (82%) and lymph nodes (32%). Moreover, only complete excision appears to improve prognosis[12]. Chemotherapeutic agents, such as doxorubicin, cisplatin, cyclophosphamide, vincristine, ifosmide, and actinomycin D, are used in combination[13-16]. Leite et al have reported that 33% of patients in their series responded to combination chemotherapy, a rate similar to that seen in other types of sarcoma, but all patients died within 2 years[13]. However, Kawamoto et al. and Yoshitani et al. have reported that STI 571, a tyrosine kinase inhibitor, inhibits cell growth and proliferation of MFH cell lines[17,18]. The prognosis of MFH may improve with new types of chemotherapy.

To our knowledge, including the present case, only 3 cases of primary MFH of the duodenum have been reported ; 2 of these cases presented with abdominal pain from perforative peritonitis and 1 case presented with melena. Whipple's procedure was performed in 2 cases ; however, both patients died soon after surgery (Table 1).

The location of the tumor is likely an important prognostic factor in retroperitoneal or abdominal

Table 1. Reported cases of primary MFH of the duodenum

Patient	Age (year)	Sex	Perforation	Therapy	Histologic Type	Follow-up	Ref.
1	29	F	+	Whipple procedure	storiform pleomorphic	died postoperatively	8
2	61	F	−	Whipple procedure	storiform pleomorphic	died of tumor at 2 months	9
3	53	M	+	Chemotherapy	storiform pleomorphic	died of tumor at 5 months	present case

MFH. Late detection of these tumors owing to their location is responsible for a poorer prognosis than for soft-tissue tumors of the extremities, as illustrated by detection of pulmonary metastasis in the present patient upon admission. Examination of a larger number of patients with MFH would allow a more precise evaluation of the biological and clinical behavior of this tumor and lead to more specific and more effective treatment.

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