[症例報告]Retroperitoneal Paraganghoma : A Case Report

Ryukyu Med. J, 7(1): 51~56, 1984.

Retroperitoneal Paraganglioma: A Case Report

Takashi Azuma, Yoshihiro Muto, Kotaro Kurihara
Shigeyoshi Terukina* and Yoshiyuki Sho

The First Department of Surgery, School of Medicine,
University of the Ryukyus
* The First Department of Internal Medicine, School of Medicine,
University of the Ryukyus

Key words : retroperitoneal paraganglioma, malignant paraganglioma

Abstract

A case of 47-year-old woman of a retroperitoneal paraganglioma arising from the organs of Zuckerkandl with metastatic tumors is presented herein. The tumor was apparent 7 years after pancreaticoduodenectomy for carcinoma of the duodenal papilla, and located in the retroperitoneum along the abdominal aorta just proximal to the inferior mesenteric artery. It was hypervascular, lobular with a cystic area and 4 × 3.5 × 2 cm. in size. Microscopically paraganglioma was characteristic. Although only conservative excision of the mass in the retroperitoneum was of surgical treatment, the patient has been free from any symptoms and doing well 8 months after surgery.

Introduction

Paraganglioma arising from the neural crest is rare and it's incidence has been reported to be as high as 0.012 per cent of the surgical specimens[1-5]. Especially only 8 cases have been reported in Japan[6-11]. We recently experienced a case of malignant paraganglioma arising from the organs of Zuckerkandl with metastatic foci in the liver. The case appears to be the 9th case in literatures in Japan.

Case Report

A 47-year-old woman suffering from an abdominal tumor with intermittent episode of abdominal pain for 3 months was admitted to the Ryukyu University Hospital on September 2, 1983. The patient had a past history of pancreaticoduodenectomy for adenocarcinoma of the duodenal papilla 7 years ago, in December 1976.

A non-tender, hard tumor was palpated in the abdomen just above the umbilicus. It was globular and approximately 5 cm. in diameter. Laboratory data were almost within norma limits except for 6.0 ng/ml of carcinoembryonic antigen (CEA). Alfa-

fetoprotein (AFP) was within normal level. An ultrasonography (US) revealed a hypoechogenic tumor in the retroperitoneum on the left to the superior mesenteric artery (SMA). A few foci of similar echogenic features were found in the right lobe of the liver. A computed tomograph (CT) also showed the tumors with high density in the same locations as US. At this time, recurrence of carcinoma of the duodenal papilla was highly suspected. However, serial selective arteriograph of the SMA revealed hypervascularity of the tumors with displacement of the SMA to the right (Fig. 1).

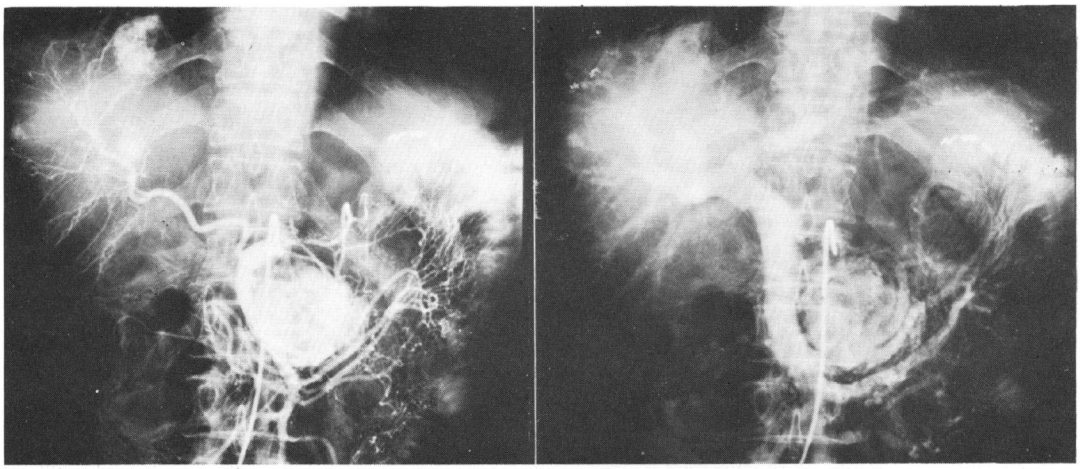

Fig. 1 Angiogram
Superior mesenteric arteriograph shows highly hypervascular tumors in the liver and in the retroperitoneum. The retroperitoneal tumor is present to the left of the SMA, displacing the SMA medially. The right hepatic artery has its origin in the SMA (left, arterial phase; right, venous phase).

Thereby, the tumor in the retroperitoneum about 3 cm. below the origin of the SMA, presumably at the level of the inferior mesenteric artery (IMA) did not seem to be a recurrent tumor, but to be a primary lesion, and a retroperitoneal sarcoma of unknown origin or multiple hemangiomas was highly suspected preoperatively.

Laparotomy was performed on October 28, 1983. Both adrenal glands appeared normal. A hard tumor located in the retroperitoneal space, lying along the aorta at the level of origin of the IMA. It was covered with the mesenterium, assuming a mesenteric tumor. The tumor was attached to the retroperitoneal space by the fibrovascular tissues. It was difficult to control bleeding during surgery because of a comparable vessels on the whole surface of the tumor. Blood loss was 1,350 ml. and no significant change in blood pressure occurred. It was impossible to take a surgical approach to the hepatic lesions because of extensive adhesions between the liver and surrounding organs.

Macroscopic Findings

The resected tumor was globular, 4 × 3.5 × 2 cm. in size and 52 gm. in weight. It was partially encapsulated and relatively well-circumscribed. Cutting the specimen, it was brownish, lobular and rather soft. There was a cystic area, 7 mm. in diameter, in the center of tumor.

Microscopic Findings

The tumor was incompletely encapsulated with a thin connective tissue. The tumor cells were oval or polygonal and arranged in nests or "zellballen" around the elaborate vasculature. They possessed abundant homogeneous or fine granular eosinophilic cytoplasm. The nuclei were round, oval or somewhat angular with small nucleoli. Large cells with hyperchromatic nuclei were scatterred (Fig. 2). Multinucleated giant cells and division figures were not found. The cystic area showed fibrosis.

Postoperative Course

The postoperative course of the patient was uneventful with normal urinary excretion of catecholamines. No change of the size and shape of the liver tumors was demonstrated by a postoperative arteriography 6 months after surgery. The patient has been well with no episode of abdominal pain and any other symptoms 8 months after surgery.

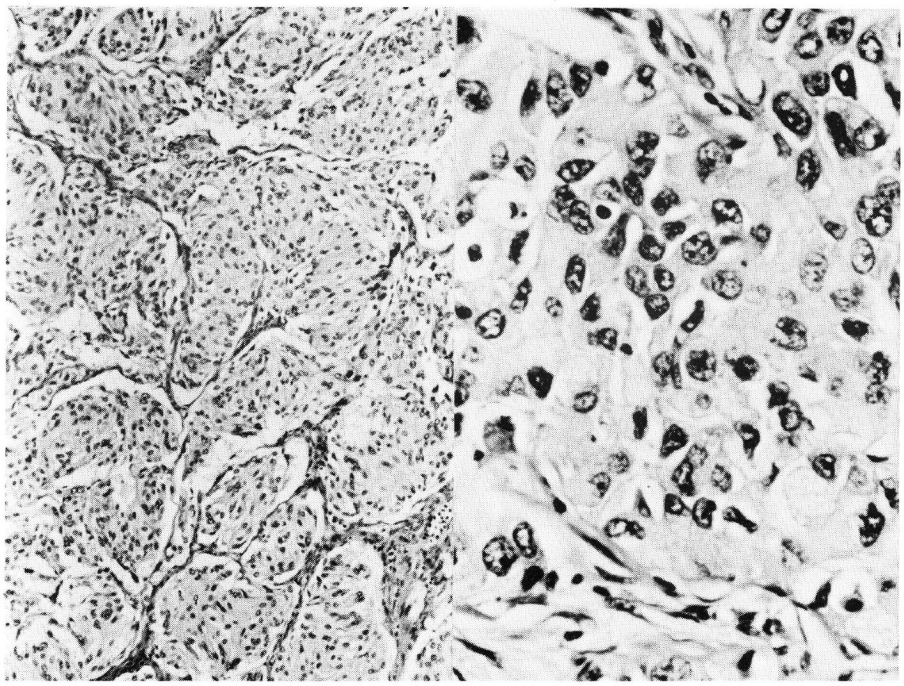

Fig. 2 Histology of the resected specimen

The tumor shows a distinctly organoid arrangement and consists of nests of cells with surrounding vascular stroma (left) (HE, × 100). The tumor cells have abundant homogeneous or finely granular eosinophilic cytoplasm with hyperchromatic oval or angular nuclei (right) (HE, × 100).

Discussion

The paraganglioma commonly arises from the carotid body. Much less frequently it occurs in the retroperitoneum and others. Retroperitoneal, extra-adrenal paraganglioma arises from paraganglia lying along the aortic axis in close association with sympathetic chain, including the organs of Zuckerkandl. Physiologically these paraganglia are prominent in early infancy but gradually regress shortly after birth within 12 to 18 months, leaving behind only small microscopic residua[1]. Paraganglioma in the present case originated in the organs of Zuckerkandl at the level of IMA.

Retroperitoneal paragangliomas occur at a relatively earlier age than those in the head and neck. Most patients are between the ages of 30 and 45. Men and women are equally affected in most series[4]. In a review of the literature in Japan[6-11], only 9 acceptable cases including the present case have been described (Table 1). There were 4 males and 5 females. The age ranged from 13 to 69 years with an average age of 48 years, and it was older than that in other countries[1-5].

Clinical manifestations were variable, but abdominal pain and / or abdominal mass were most common. Eight of nine patients were suffered from an abdominal pain and / or abdominal mass. Whereas, in 30 patients in the English literature[5], abdominal pain was apparent in only 50 per cent.

The great majority of the retroperitoneal paragangliomas were found along the aorta from the celiac axis down to the iliac artery. They were relatively well-circumscribed and encapsulated with focal necrotic or hemorrhagic areas. The biggest one was 6, 500 gm. in wieght[7]. The present case was the smallest with 52 gm. in weight.

In the majority of cases, surgical excision was performed excepting one patient who expired 9 hours after aortography prior to operation. One patient underwent postoperative radiation. The postoperative prognosis in survival estimated of 6 cases was 6 to 36 months, however it was less enough to definite postoperative prognosis. It is generally acceptable that complete removal of the tumor is of choice and that the widely extended tumors are out of surgical indication. In the present case, however, the operation was carried out to establish the diagnosis and to alleviate the abdominal pain. Although removal of the retroperitoneal tumor was conservative, the patient has been free from abdominal pain 8 months after surgery.

With regard to malignancy of paraganglioma under discussion, it is generally believed that these tumors are benign, possessing long survival even when unresectable or locally recurrent. A small percentages have been to be malignant by virtue of metastases[1)4)5)]. In a review of the English literature4), 6 of 21 cases (28 per cent) developed clinical evidence of metastases, however it was proven histologically in only 4 cases. In the reported cases in Japan, 3 of 9 cases (33 per cent) seemed to be malignant. One of them showed local invasion and the remaining two cases metastases to the liver. In some cases documented in the English literatures, there were 2 or more distinctly separate tumors which were considered to be autochthonous primary tumors. Although it was not determinable whether the liver tumors represented metastases or

Table 1. Summary of retroperitoneal paragangliomas reported in the literature in Japan

No.	Age	Sex	Symptoms and signs	Tumor size weight	Treatment blood loss	Metastses	Survival
1[6]	46	M	Abdominal mass	7 × 7 × 5 cm	Excision	None	No follow-up
2[7]	69	F	Abdominal mass	6,500 gm	None	None	Autopsy
3[8]	46	M	Abdominal mass	375 gm	Excision	Invation	Alive, 2 yrs. and 3 mos.
4[9]	13	F	Abdominal mass	17×12×10 cm 1,330 gm	Excision 15,527 ml	None	Alive
5[10]	48	F	Abdominal mass	15×10 cm 1,480 gm	Excision 4,200 ml	Liver	Death following operation
6[10]	43	M	Abdominal mass	2,800 gm	Excision 25,000 ml	None	Alive, 2 yrs. and 6 mos.
7[10]	54	F	Fever	10×15 cm 1,950 gm	Excision 3,730 ml	None	Alive 1 yrs. and 6 mos.
8[11]	66	M	Abdominal mass	10×10×13 cm 1,300 gm	Excision 2,700 ml	None	Alive, 3 yrs. and 3 mos.
9*)	47	F	Abdominal mass	4 ×3.5×2 cm 52 gm	Excision 1,350 ml	Liver	Alive, 6 mos.

*) present case

autochthonous lesions, the tumors in the liver in the present case were considered to be metastatic foci because the neural crest is not present in the liver[12)13)].

The microscopic feature of malignancy has not been well delineated and the close correlation between the histological appearance and the clinical behavior is poorly established. Therefore, it is not reliable to define th malignancy of the tumor only by histologic examination and the more important histological criteria is demonstrated of invasion to the adjacent organs or tissues or of metastases. Paraganglioma is generally slow-growing tumor, and long-term survival has been documented in the presence of local invasion and even in patients with metastases[1)]. Thus, surgical excision of the tumor should be attempted once the diagnosis has been made.

References

1) Enzinger, F.M. and Weiss, S.W. : Soft tissue tumors, The C.V. Mosby Co., St. Louis, 1983.
2) Brantigan, G.O. and Katase, R.Y. : Clinical and pathologic features of paragangliomas of the organ of Zuckerkandl. Surgery 65 : 898-905, 1969.
3) Glenn,F. and Gray, G.F. : Functional tumors of organ of Zuckerkandl. Ann. Surg. 183 : 578-581, 1976.
4) Olson, G.R. and Abell, D.M. : Nonfunctional nonchromaffin paragangliomas of the retroperitoneum. Cancer 23 : 1358-1367, 1969.
5) Hall, G.M., Morris, G.M. and Masson, G.R. : Nonfuctional retroperitoneal paragangliomas. Amer. J. Surg. 139 : 257-261, 1980.
6) Kanai, H., Abe, T., Muto, T. and Endo, S. : A case report of the retroperitoneal paraganglioma. Jap. J. Cancer Clin. 11 : 701-705, 1965 (in Japanese).
7) Koike, R. and Yokoyama, M. : : A case of the huge paraganglioma. J. Clin. Urol. 21 : 676-677, 1967 (in Japanese).
8) Takayasu, H., Yokoyama, M. and Hoshino, Y. : A case of nonfunctioning malignant paraganglioma. Nihon-Rinsho 26 : 2104-2107, 1968 (in Japanese).
9) Sakurai, H., Yamaoka, I., Arai, M., Sasaki, G., Iwafuchi, M. and Ukishima, J. : A case of successfully excised nonchromaffin retroperitoneal paraganglioma (1,330 gm). Jap. J. Clin. Surg. 32 : 63-69, 1971 (in Japanese).
10) Hara, K., Shigematsu, S., Korehisa, H., Morioka, Y. and Sugawara, K. : Three cases of retroperitoneal paraganglioma, preoperatively diagnosed as hepatoma. Jap. J. Gastroenterol. Surg. 10 : 293-299, 1977 (in Japanese).
11) Kawarada, Y., Yishimura, A., Nishida,S., Sekoguchi, T. and Mizumoto, R. : Nonfunctioning retroperitoneal paraganglioma : A case report, diagnosed as hepatoma preoperatively. Jap. J. Clin. Surg. 41 : 1074-1080, 1980 (in Japanese).
12) Pearse, A.G.F. : The APUD cell concept and its implications in pathology. Pathol. Ann. 9 : 27-41, 1974.
13) Sidhu, G.S. : The endodermal origin of digestive and respiratory tract APUD cells - Histologic evidence and review of the literature. Amer. J. Pathol. 95 : 5-20, 1978.