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Malignant Solitary Fibrous Tumor of the Thigh Accompanied by Hypoglycemic Coma. A Case Report

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Abstract. *A case of malignant solitary fibrous tumor of the thigh with liver and lung metastasis, suspicious of non-islet cell tumor-associated hypoglycemia is reported. The patient, a 76-year-old woman, was initially aware of an increasing soft tissue tumor in the left thigh, and episodes of hypoglycemia gradually developed. These frequent episodes ceased after resection of the thigh tumor, which was diagnosed as malignant solitary fibrous tumor. Considering that (i) the primary lesion developed in a rare site, (ii) the tumor was a malignant variant (accompanied by liver and lung metastasis), and (iii) the lesion was accompanied by typical hypoglycemic episodes, this case seemed to be extremely rare.*

Case Report

A 76-year-old female was first admitted to Ayabe City Hospital, Japan, with huge soft tissue tumor of the left thigh in July 1998. She had been diagnosed as having growth hormone(GH) deficiency because of low serum GH levels and low response on GH stimulating test since 1991. The patient noted an increasing soft tissue tumor in the left thigh, forming a huge tumor and hypoglycemic episodes had gradually begun to occur. After admission, hypoglycemic coma frequently occured and was treated with glucose injections. The serum blood sugar level during the hypoglycemic coma was 10-50 mg/dl. A radiogram and CT scan of her thigh showed a large soft tissue shadow with a small area of calcification. The MR image showed a huge soft tissue tumor occupying the posterior of the thigh (18 x 12 x 10 cm) (Figure 1). A CT scan and MR image of the chest and abdomen indicated lung and liver metastases (Figure 2). An angiogram showed the hypervascularity of the tumor, and Thallium scintigram indicated abnormal accumulation in the tumor. After open biopsy, wide tumor resection was performed. Histological diagnosis of the tumor was malignant solitary fibrous tumor (Figure 3). Immunohistochemical studies showed CD34 and vimentin positivity, smooth muscle actin some positivity, S-100, myoglobin, cytokeratin and PAS negativity (Table I). Post-operatively, frequent hypoglycemia attack ceased. The blood levels of GH and somatomedin C (insulin-like growth factor I) increased and the response on the growth hormone stimulating test normalized (Table II). The liver metastasis was resected in September 2000. The histological findings of the metastatic tumor were similar to those of the thigh tumor. The patient died in August 2002 due to the multiple lung metastasis.

Discussion

Solitary fibrous tumor is a relatively rare mesenchymal neoplasm, also called fibrous mesothelioma, and occurs preferentially in the pleura and peritoneum. This tumor occurs in adults and mainly affects persons over 50 years of age. The benign variant seems to be three to four times more common than the malignant one. It was reported that hypoglycemia occurs in about 5% of cases, is more common in females than in males, and sometimes is the first manifestation of disease. Recently, this tumor type has been reported in numerous sites including the liver, orbit, nasal passages, meninges, pericardium, tunica vaginalis, testis, skin, respiratory tract, thyroid and soft tissue (1-7).

It is well known that various tumors other than insulinoma, or non-islet cell tumors, cause hypoglycemia. It is suspected that this results from the production of insulin-like growth factor II (IGF-II), which suppresses GH and IGF-I. In this case, the IGF-II blood level was

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Key Words: Malignant solitary fibrous tumor, soft tissue tumor, hypoglycemia, insulin-like growth factor II.

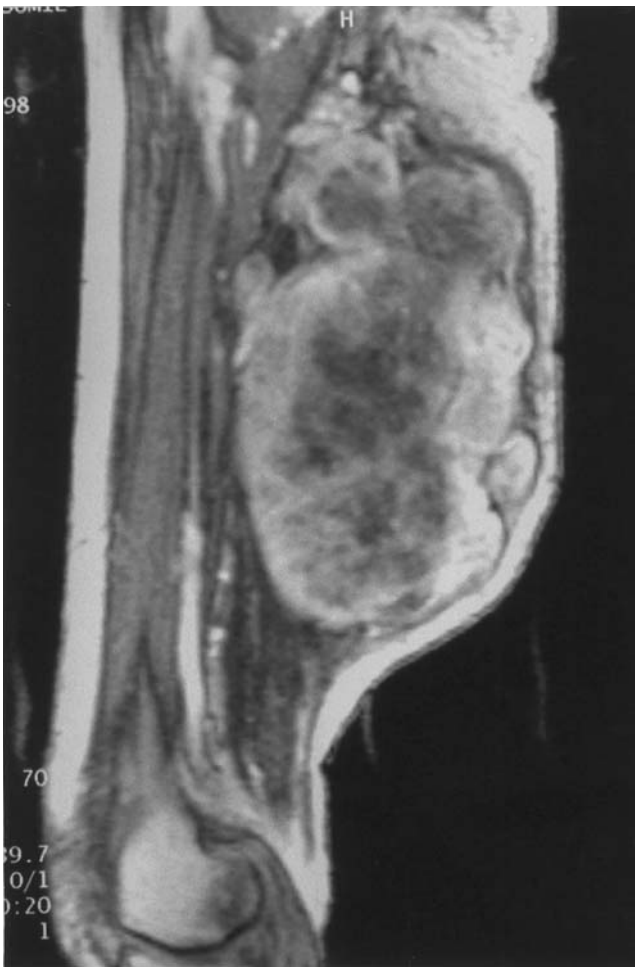

Figure 1. *MR image of the soft tissue tumor at the thigh (T1W-Gd).*

Table I. *Immunohistochemical results of the tumor.*

CD34 , vimentin	(+)
smooth muscle actin	(±)
S-100, myoglobin, cytokeratin, PAS	(–)

Table II. *(1) Endocrinological data before and after resection of the tumor.*

	before	after
C-peptide (1.0-2.5 ng/mL)	<0.1	1.2
growth hormone (0.66-3.68 μU/mL)	0.4	2.8
IGF-I (121-436 ng/mL)	37	79
IGFBP-III (2.17-4.05 μg/mL)	ND	2.22
Cortisol (2.7-15.5 μg/dL)	20.1	9.3

IGF-I: insulin-like growth factor-I, IGFBP-III: insulin-like growth factor binding protein-III, ND: not determined

(2) Serum growth hormone (GH) levels (ng/mL) on GH stimulation test.

Arginine test (0.5g/kg, *i.v.*)

	0	15	30	60	90	120 (min)
before resection	0.3	ND	1.2	0.7	0.4	0.4
after resection	3.3	ND	12.4	24.7	18.9	10.4

Insulin tolerance test (0.1U/kg, *i.v.*)

	0	15	30	60	90	120 (min)
before resection	2.6	5.0	11.8	11.5	11.7	6.9
after resection	0.3	1.7	1.6	22.7	ND	16.9

not measured. However, it seemed highly possible that secretion of IGF-II had increased, based on the patient's endocrinological data. The histology of non-islet tumors has been reported as manifold mesenchymal tumors (mesothelioma, lymphoma, hemangioperictoma, fibrosarcoma leiomyosarcoma, and so on), various types of carcinoma (hepatoma, adrenocortical carcinoma, gastric carcinoma, carcinoma of lung, and so on) and neurogenic tumors (8-14). However, some of these findings were described in older references, and these histological diagnoses were not based on current standards. To our knowledge, only one case of non-islet cell tumor arising in the extremities, described as leiomyosarcoma in the thigh, has been reported (15). In this case, we conclude that the patient had malignant solitary fibrous tumor, based on the histological findings and CD-34 positivity (7). By any standard, a tumor arising in the extremities and associated with hypoglycemia seems to be extremely rare.

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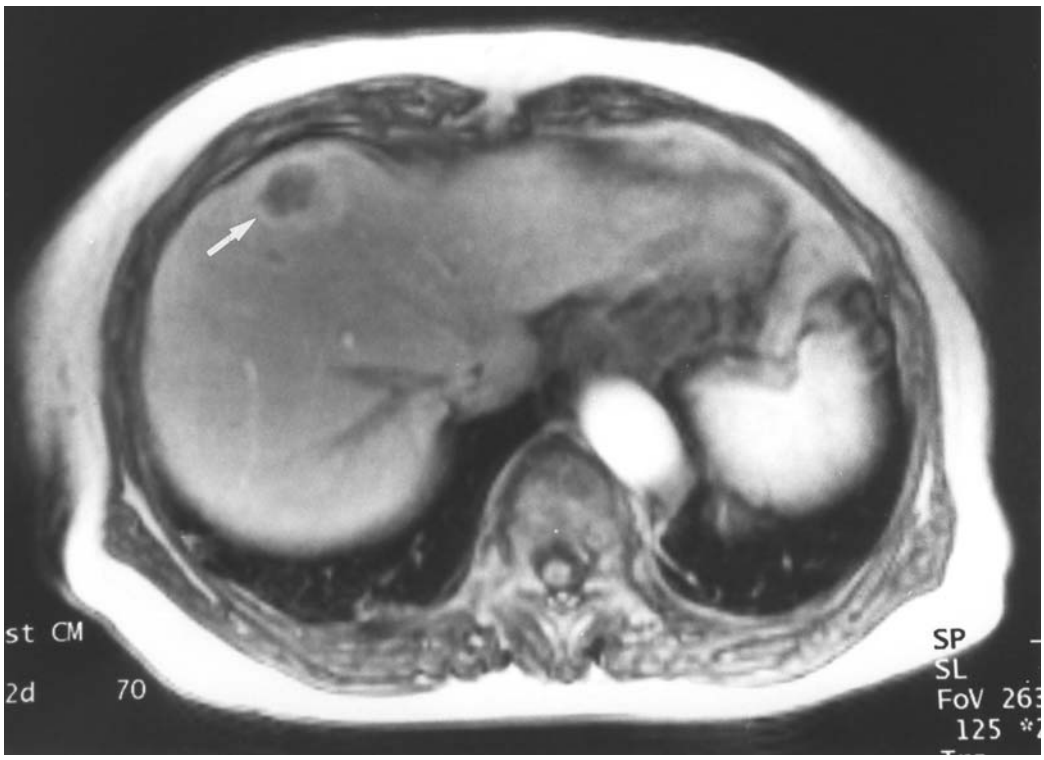

Figure 2. *MR image of the abdomen demonstrated liver metastasis.*

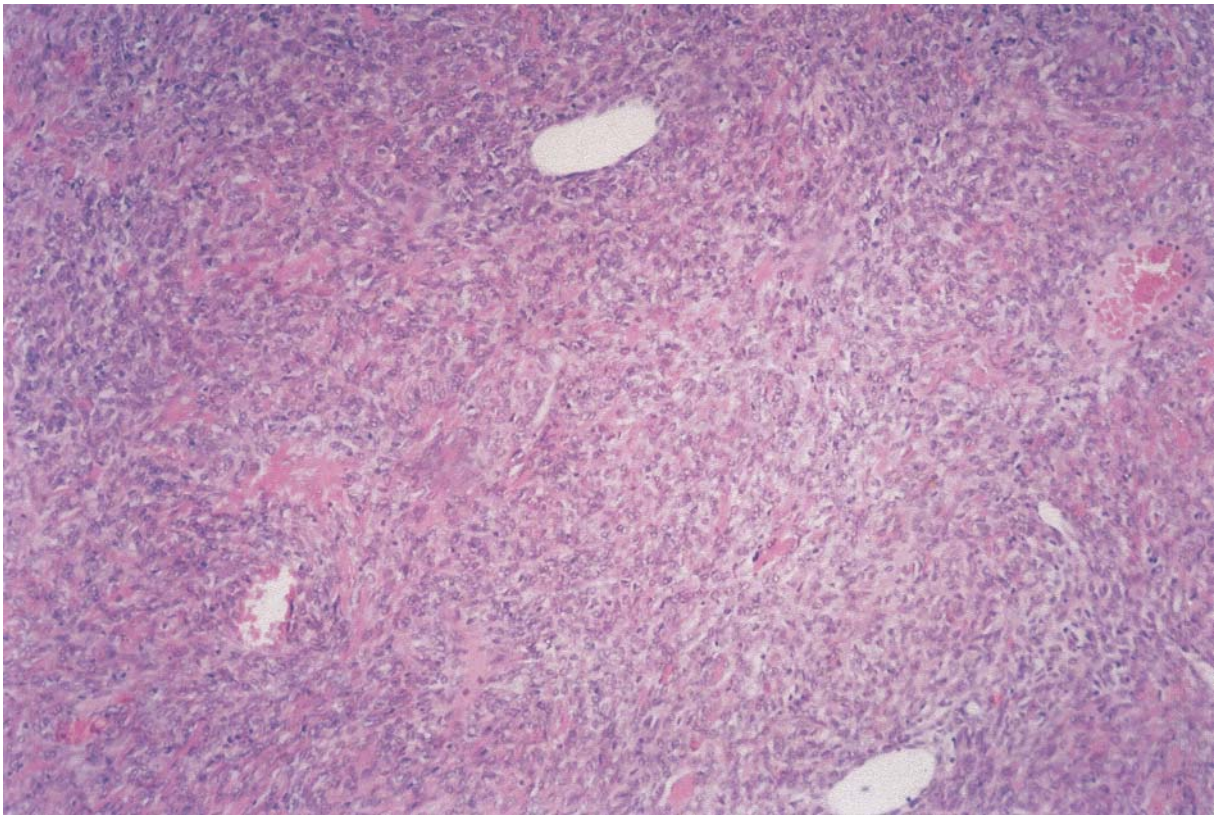

Figure 3. *The histological findings of resected specimen; the tumor was diagnosed as malignant solitary fibrous tumor.*

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