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Solitary Fibrous Tumor in the Pelvic Cavity with Hypoglycemia: Report of a Case

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Abstract
A case of solitary fibrous tumor (SFT) in the pelvic cavity with hypoglycemia is reported. The patient was a 60-year-old man who was referred to our hospital for a closer examination of hypoglycemia. Computed tomography demonstrated a mass, measuring 14 × 9 cm in size, in the pelvic cavity. Magnetic resonance imaging showed the mass to have a low signal intensity on T1-weighted images and a high intensity on T2-weighted images. Laparotomy revealed no peritoneal dissemination nor lymph node metastasis. An en bloc excision of the tumor was performed with a good recovery, and the hypoglycemia disappeared. Histologically, the tumor was composed of spindle-shaped and oval cells in sarcoma, based on a moderate mitotic rate and cellularity. Immunohistochemically, the tumor was positive for CD34 and negative for keratin, α-smooth muscle actin, desmin, S100 protein, c-kit protein, and epithelial membrane antigen. Based on these findings, the tumor was diagnosed to be malignant SFT in the pelvic cavity.

Key words Solitary fibrous tumor · Pelvic cavity · Hypoglycemia

Introduction

Solitary fibrous tumors (SFTs), which are known as either localized fibrous tumors or fibrous mesotheliomas, are rare spindle-cell neoplasms first reported as arising from the pleura.[1] Although most reported SFTs tend to arise within the thoracic cavity, extrathoracic SFTs, which occur over a wide range of anatomic sites, have also been reported.[2] Among extrathoracic SFTs, the occurrence of a primary SFT in the pelvic cavity is rare. Solitary fibrous tumors can cause non-islet cell tumor hypoglycemia (NITCH) due to the secretion of hormone-like substances. We herein present the case of a patient with an SFT in the pelvic cavity accompanied by hypoglycemia.

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Case Report

A 60-year-old man was referred to our hospital for a closer examination of a hypoglycemic crisis. On admission, the serum glucose level was 48 mg/dl, the serum insulin was below 1.0 μU/ml, and the serum C-peptide was 0.2 ng/ml. As the patient received continuous dextrose infusion, the hypoglycemic crisis disappeared. Computed tomography (CT) demonstrated a lobulated mass lesion, measuring 14 × 9 cm in size, in the pelvic cavity. On magnetic resonance imaging (MRI), the mass showed a low signal intensity on T1-weighted images and high intensity on T2-weighted images (Fig. 1). Angiography showed a hypervascular tumor fed by a branch of the inferior mesenteric artery (IMA). A laparotomy was performed through a lower median skin incision. There was no peritoneal dissemination and no lymph node metastasis was observed. The tumor was located on the left side of the pelvic cavity. An incision was made on the retroperitoneum to expose the tumor. The feeding vessels were ligated and cut then, using a blunt and sharp dissection, the tumor was completely excised. The postoperative course was good and the serum glucose level returned to normal immediately after the excision of the tumor.

Pathology

The tumor was well circumscribed and encapsulated, measuring 22 × 10.5 × 9.5 cm in size and weighing 745 g. The cut surface was grayish-white, showing components of hemorrhaging (Fig. 2). Histologically, the tumor was

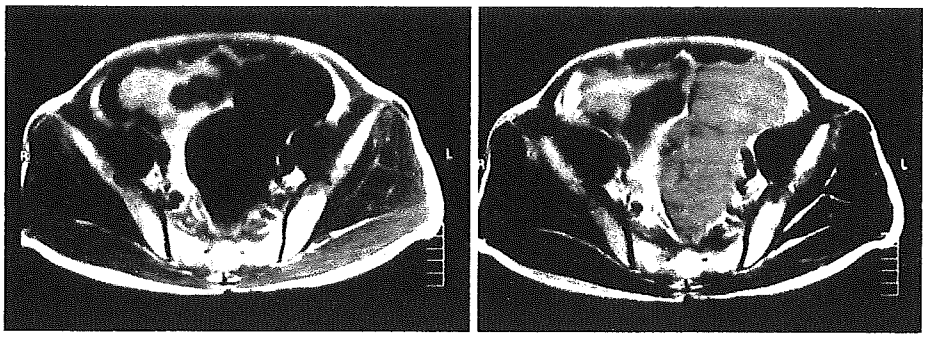

Fig. 1A,B. T1-weighted magnetic resonance imaging (MRI) showed a mass lesion of low signal intensity in the pelvic cavity (**A**). The mass showed a high signal intensity on T2-weighted MRI (**B**)

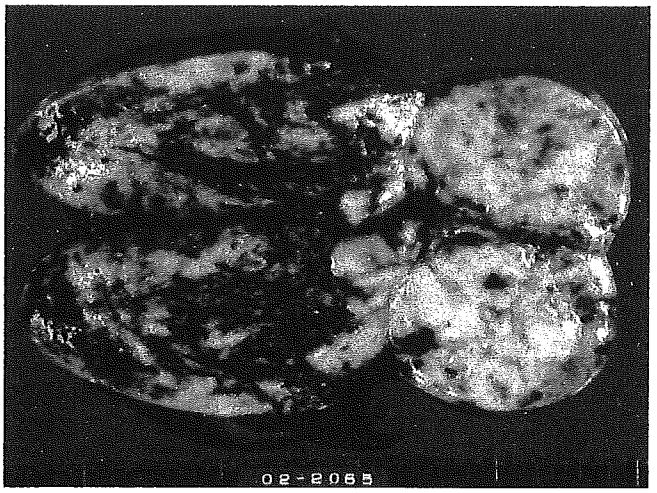

Fig. 2. The cut surface of the resected tumor was grayish-white, showing components of hemorrhaging

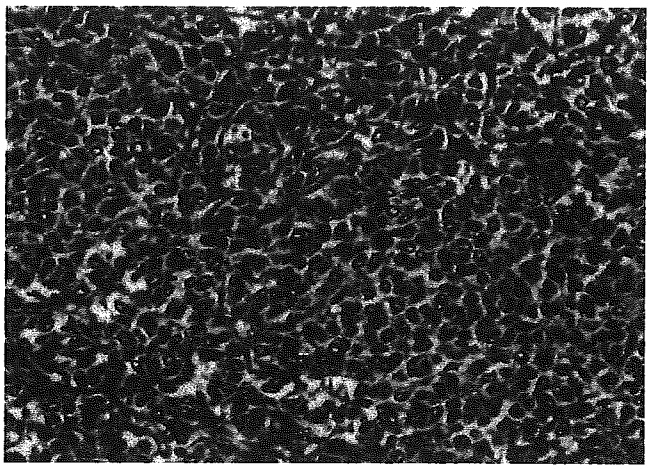

Fig. 3. Microscopic findings of the tumor. The tumor was composed of spidle-shaped and oval cells in sarcoma, based on a moderate mitotic rate and cellularity (H&E, ×200)

composed of spindle-shaped and oval cells in sarcoma, based on moderate mitotic rate and cellularity (Fig. 3). Immunohistochemically, the tumor was positive for CD34, vimentin, and bcl-2; however, it was negative for keratin, α-smooth muscle actin, desmin, S100 protein, c-kit protein, and epithelial membrane antigen (Fig. 4). Based on these findings, the tumor was diagnosed to be malignant SFT of the pelvic cavity (intermediate grade). In addition, the immunohistochemical localization of insulin-like growth factor-II (IGF-II) was analyzed on paraffin sections of the tumor using the labeled streptavidin-biotin method. The antibody used was mouse monoclonal antiserum for human IGF-II (Peninsula Laboratories, San Carlos, CA, USA). The tumor cells stained positively for IGF-II (Fig. 5).

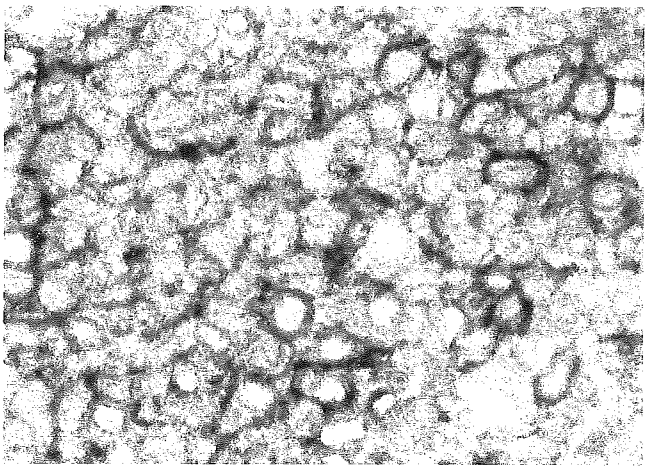

Fig. 4. Immunohistochemical findings of CD34. The tumor cells stained positively for CD34 (immunohistochemistry, ×500)

Discussion

Solitary fibrous tumors are rare spindle-cell neoplasms. Since their initial description arising from the pleura,

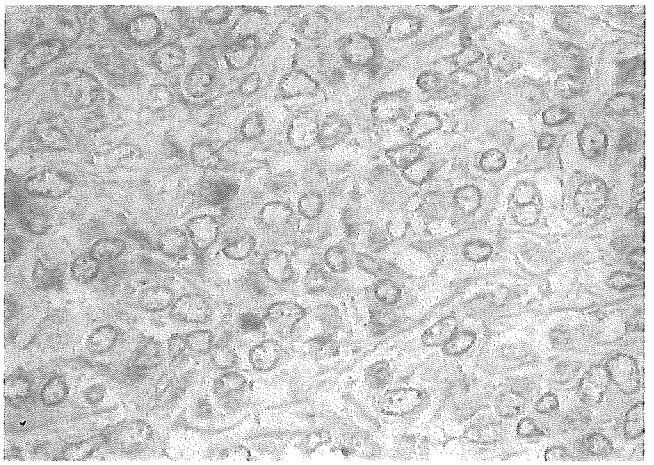

Fig. 5. Immunohistochemical findings of insulin-like growth factor-II (IGF-II). The cytoplasm of the tumor cells stained positively for IGF-II (immunohistochemistry, ×500)

SFTs have been reported to occur at a wide range of anatomic sites.[1,2] Among extrathoracic SFTs, primary SFT in the pelvic cavity is rare. Solitary fibrous tumors in the pelvic cavity occurred in 16% of 79 cases of SFTs involving various sites.[2] Thoracic SFTs are often detected when still asymptomatic on chest X-rays, while extrathoracic SFTs are often symptomatic when detected. The major symptoms of abdominal SFTs are abdominal pain, abdominal fullness, or a palpable abdominal mass.[3] Hypoglycemia rarely occurs. To our knowledge, only one case of hypoglycemia associated with abdominal SFT has been reported previously.[4] Hypoglycemia occurred in 4% of 360 cases of SFTs of the pleura.[5] As the thoracic and extrathoracic SFTs appear to involve a single biological disease entity, the incidence of hypoglycemia associated with abdominal SFTs is thus considered to occur at almost the same rate (4%).

Non-islet cell tumor hypoglycemia is a paraneoplastic syndrome occurring in patients with large slow-growing tumors such as hepatomas, sarcomas, and other tumors arising from the mesenchymal tissue.[6] The mechanism of NITCH is still not fully understood. The most common cause of NITCH is the production of IGF-II, which is structurally similar to proinsulin. Insulin-like growth factor-II, which is produced by the tumor, acts on insulin receptors thereby inducing hypoglycemia, and suppressing such hormones as insulin and growth hormone.[7–9] Recent studies have shown a large molecular form, designated big IGF-II, which is probably an incompletely processed molecule of IGF-II and may play a key role in the pathogenesis of NITCH.[9,10] The surgical removal of the tumor usually leads to a complete remission of NITCH.[8] We speculate that NITCH caused by IGF-II production was most likely associated with our case, because the clinical course of hypoglycemia and the serum biochemistry findings were compatible with those of NITCH, and the existence of IGF-II protein in tumor cells was demonstrated immunohistochemically.

A CT scan of SFT usually demonstrates a well-delineated, homogeneous, and occasionally lobulated mass of soft tissue attenuation.[11,12] On MRI, fibrous tissue of SFT has a low signal intensity on T1-weighted images. On T2-weighted images, however, mature fibrous tissue has a low intensity, while malignant fibrous tissue tends to demonstrate a high signal intensity. Although the radiological findings in our case were typical of SFT, it is often difficult to differentiate this lesion from other mesenchymal tumors. As fine-needle aspiration or a CT-guided Tru-cut biopsy are useful for the preoperative diagnosis, these examinations should be performed if the tumor is accessible by puncture needles.[13,14]

A complete en bloc surgical resection is the standard therapy for SFT. Solitary fibrous tumors have a low rate of local recurrence and metastasis after surgical resection.[2] However, tumors larger than 10 cm or those demonstrating a histologically malignant component have an increased risk of local recurrence and metastasis, and thus require a close follow-up.

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