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Recurrent superior mediastinal primary hemangiopericytoma 23 years after the complete initial excision: a case report.

Akio Hiraki* Tomoyuki Murakami† Keisuke Aoe‡

Eisuke Matsuda** Tadashi Maeda††

Yoshiki Umemori‡‡ Hiroshi Ueoka§

*NHO Sanyo National Hospital,

†NHO Sanyo National Hospital,

‡NHO Sanyo National Hospital,

**NHO Sanyo National Hospital,

††NHO Sanyo National Hospital,

‡‡NHO Sanyo National Hospital,

§NHO Sanyo National Hospital,

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Abstract

We describe here a patient with a recurrent hemangiopericytoma of the superior mediastinum 23 years after an initial complete resection. In the current biopsy specimen, the tumor cells were much more anaplastic than those seen 23 years ago. Although the patient was treated with chemotherapy, which consisted of ifosfamide and epirubicin, the tumor was unresponsive and he died 6 months later from disease progression. Careful long-term follow-up is mandatory for patients with hemangiopericytomas because recurrence with greater malignancy can develop following an extended disease-free interval.

KEYWORDS: primary hemangiopericytoma, recurrence, mediastinal tumor

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Case Report

Recurrent Superior Mediastinal Primary Hemangiopericytoma 23 Years after the Complete Initial Excision: A Case Report

Akio Hiraki[a], Tomoyuki Murakami[b,c], Keisuke Aoe[a,b]*, Eisuke Matsuda[d], Tadashi Maeda[a,b], Yoshiki Umemori[d], and Hiroshi Ueoka[a]

Departments of [a]Respiratory Medicine, [b]Clinical Research, [c]Pathology, and [d]Surgery, NHO Sanyo National Hospital, Respiratory Disease Center, Ube, Yamaguchi, 755–0241, Japan

We describe here a patient with a recurrent hemangiopericytoma of the superior mediastinum 23 years after an initial complete resection. In the current biopsy specimen, the tumor cells were much more anaplastic than those seen 23 years ago. Although the patient was treated with chemotherapy, which consisted of ifosfamide and epirubicin, the tumor was unresponsive and he died 6 months later from disease progression. Careful long-term follow-up is mandatory for patients with hemangiopericytomas because recurrence with greater malignancy can develop following an extended disease-free interval.

Key words: primary hemangiopericytoma, recurrence, mediastinal tumor

Hemangiopericytoma is an uncommon soft-tissue sarcoma first described by Stout and Murray in 1942 [1]. The origin of the tumor is pericytes, cells first defined by Zimmermann [1, 2], that are normally arranged around capillaries and postcapillary venules and modulate blood flow and permeability. Consequently, they may occur anywhere capillaries are found. Hemangiopericytomas occur most frequently in the extremities, pelvis and retroperitoneum, head and neck, and meninges, and are rarely encountered in the mediastinum [2]. Here, we describe a patient with a recurrent hemangiopericytoma of the superior mediastinum 23 years after an initial complete resection.

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*Corresponding author. Phone: +81-836-58-2300; Fax: +81-836-58-5219
E-mail: aoe@sanyou-dr.jp (K. Aoe)

Case Report

An asymptomatic 69-year-old Japanese man was referred to us for further examination of a mediastinal mass identified on January 1, 2003. He had already undergone a complete excision of a mediastinal tumor in our hospital 23 years previously. The pathologic diagnosis at that time was reported as a probable thymoma with a pericytomatous pattern.

Physical examination on admission revealed no abnormalities. Laboratory findings including tumor markers were within normal limits, apart from mild anemia. Serum tumor markers were not elevated, including cytokeratin 19 fragment, carcinoembryonic antigen, α-fetoprotein, and human chorionic gonadotropin. Plain radiography and computed tomography (CT) of the chest revealed a 10×8 cm mass in the anterior mediastinum (Fig. 1). CT scans of the abdomen and brain and bone scintigraphy demonstrated no evidence of distant metastases. The patient under-

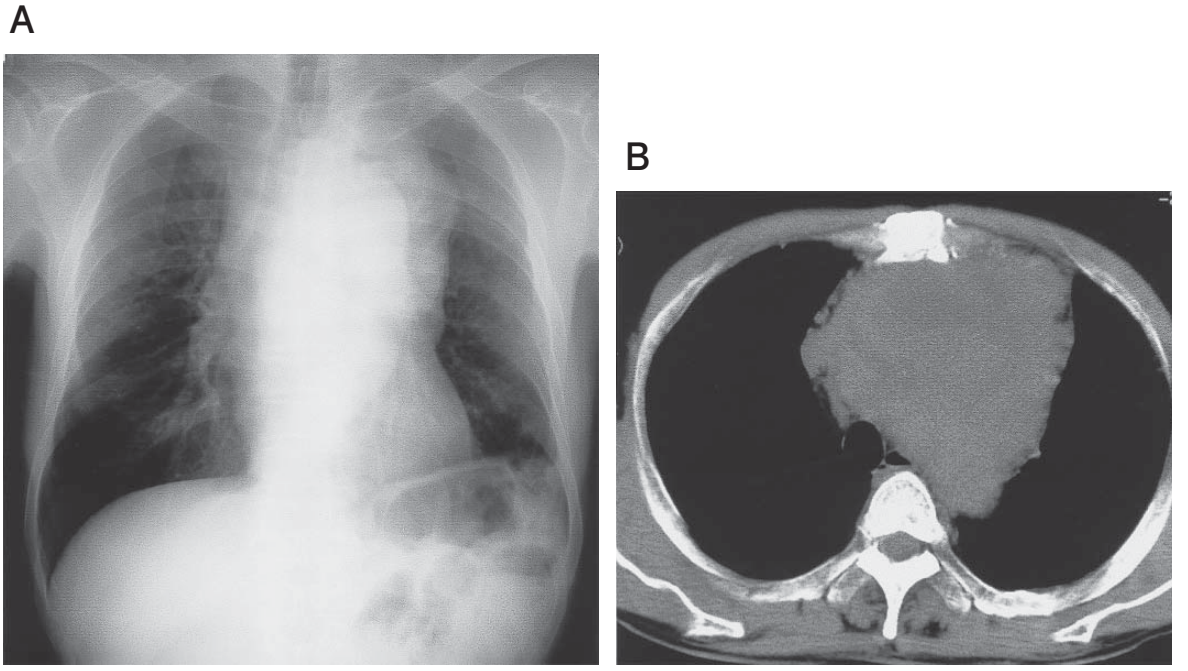

Fig. 1 A chest plain radiography and computed tomography (CT) of the chest revealed a 10 × 8 cm anterior mediastinal mass.

went percutaneous needle biopsy, which revealed an irregular proliferation of anaplastic spindle cells with foci of pericytomatous patterns (Fig. 2A). Most of the tumor cells were immunohistochemistrically reactive against vimentin and CD34, and none of the tumor cells were reactive against cytokeratin AE1/AE3, epithelial membrane antigen, factor Ⅷ, S-100, desmin, α-smooth muscle actin, myoglobin, HBME-1, synaptophysin, or chromogranin A-3. Two pathologists re-evaluated the hemtoxylin-eosin (HE)-stained sections of the tumor that was resected 23 years ago (Fig. 2B). The histologic appearance was compatible with typical hemangiopericytoma, and the tumor cells were much less anaplastic than those seen in the present biopsy specimen. From these findings, the present tumor was diagnosed as a recurrent primary mediastinal hemangiopericytoma with anaplastic changes.

Treatment was initiated February 24, with chemotherapy consisting of ifosfamide (1.5 g/m^2) on days 1 to 3 and epirubicin (60 mg/m^2) on day 1. The patient showed no response after completion of 2 cycles of chemotherapy. A systemic evaluation, including CT scans of the abdomen and brain and bone scintigraphy, demonstrated new distant metastases to the bone, liver, and brain. The patient was discharged with supportive care. He died 6 months later of disease progression.

Discussion

Hemangiopericytoma is a rare soft-tissue sarcoma derived from mesenchymal cells with pericytic differentiation [1]. There are only a few published reports on this type of soft-tissue sarcoma; information on the clinical features and management of this tumor is seriously lacking.

Surgery remains the mainstay of treatment, and the role of chemotherapy and irradiation has not yet been established, because experience in the management of this tumor is limited [3]. The overall prognosis of this tumor is relatively favorable, with 5- and 10-year actuarial survival rates of 86% to 47%, respectively [3–5]. The interval to disease recurrence in a previous report was longer than has been observed for other sarcomas. One report stated that the median time to local recurrence after curative intent was 29 months (range, 2 to 225 months) [5].

Microscopic diagnosis is based primarily on the recognition of an architecture characterized by a pericytomatous pattern with tightly packed cells around ramifying thin-walled, endothelium-lined vas-

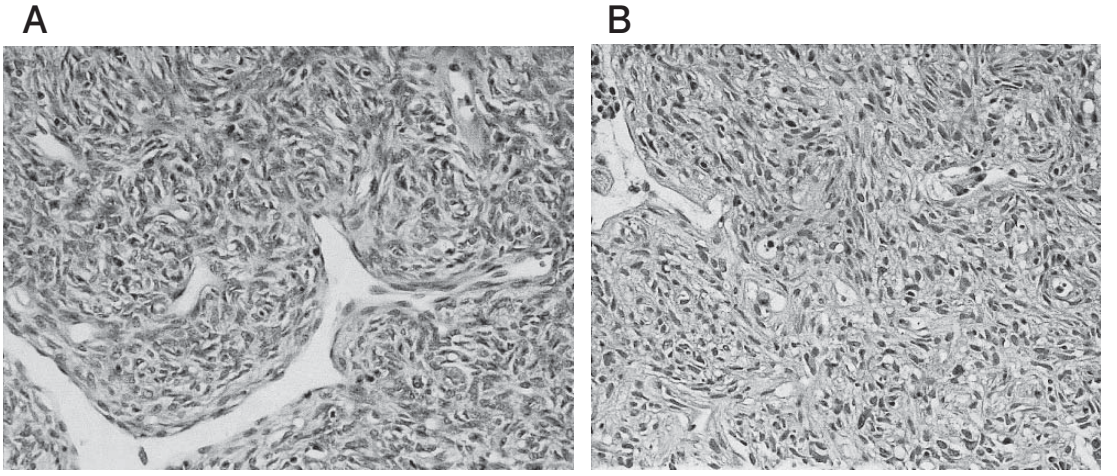

Fig. 2 Histologic findings of the hemangiopericytoma relapse (**A**) and on the first presentation 23 years ago (**B**). (Hematoxylin and eosin stain; original magnification, ×200).

cular channels ranging from small capillary-sized vessels to large gaping sinusoidal spaces [2, 6]. The pericytomatous pattern, however, occurs in a variety of benign and malignant neoplasms, such as fibrous histiocytoma, synovial sarcoma, mesenchymal chondrosarcoma, angiosarcoma, mesothelioma, extrapleural solitary fibrous tumor and others including thymoma [6]. Therefore, distinguishing of hemangiopericytoma from other tumors may be difficult, especially when the characteristic features of other neoplasms are inconspicuous.

Recently, immunostaining has become a powerful technique for definitive differentiation [7]. Our immunohistochemical examination demonstrated that the tumor was non-epithelial (vimentin-positive and cytokeratin-negative) and had CD34 antigens, a marker of pericytes and hematopoietic progenitors. More than 20 years ago in Japan, immunostaining was not available in routine examinations, and histologic examination was performed only with HE staining combined with other histochemical analyses. Therefore, pathologists in the past faced greater difficulties in the distinction of hemangiopericytoma from other tumors.

The difficulties of predicting the clinical behavior of hemangiopericytomas have been repeatedly stressed in the literature. However, Enzinger and Smith [2] in a review of 106 cases, reported that a malignant clinical course is associated with a large tumor (greater than 5 cm), an increased mitotic rate (greater than 3 mitotic figures per HPF), high cellularity, immature and pleomorphic tumor cells, and foci of hemorrhage and necrosis. In the present case, we evaluated HE sections of the primary tumor according to that criteria and found that none of the factors indicated a poorer prognosis, though the cellularity was somewhat high. These findings might explain the very long interval to recurrence in this case.

On the other hand, the recurrent tumor showed immature and pleomorphic (anaplastic) cells, and the specimen was highly cellular. The small biopsy specimen, however, did not allow us to check mitotic index, hemorrhage or necrosis. The anaplastic feature and the large size of the recurrent tumor may explain the resistance to chemotherapy and the development of multiple metastases. Even for recurrent cases, the histologic features of hemangiopericytomas should be taken into consideration for treatment strategy and prognosis.

The residual tumor cells after the initial excision likely grew and accumulated genetic and phenotypic changes to become more highly malignant at the time of clinical recurrence. Few accounts have been found of such a highly anaplastic and malignant change in a recurrent hemangiopericytoma; therefore, our case serve as an important warning of that risk.

In summary, we describe a patient with recurrent hemangiopericytoma of the superior mediastinum 23 years after initial curative resection. Careful long-

term follow-up is mandatory for patients with hemangiopericytomas because recurrence with greater malignancy can develop following an extended disease-free interval.

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