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Spindle Cell Hemangioendothelioma

—A case report—

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Spindle cell hemangioendothelioma is a rare vascular tumor which is presented with subcutaneous nodules and follows a benign indolent course but has a recurrent tendency, and is histologically resembling a cavernous hemangioma and Kaposi's sarcoma. We present a case of spindle cell hemangioendothelioma possessing clinical aggressiveness with painful bony erosion, histologic pleomorphism and mitoses.

A 20-year-old man presented with a recurrent painful mass on the left ankle. The mass was dark brown and firm with irregular margins and measured 1.5 cm in diameter, which affected and eroded the underlying medial malleolus of the left tibia. Microscopically, the tumor was composed of cavernous endothelial-lined blood spaces and spindle cellular areas mimicking Kaposi's sarcoma. The spindle cells intermingled with plump epithelioid cells and showed a moderate degree of pleomorphism with occasional mitoses. Immunohistochemically, the spindle cells were focally positive for factor VIII-associated antigen and vimentin, and negative for S-100 protein, desmin, and epithelial membrane antigen.

Key Words : *Hemangioendothelioma, Spindle cell hemangioendothelioma, Deep soft tissue, Bony erosion.*

INTRODUCTION

Spindle cell hemangioendothelioma is a rare vascular neoplasm described by Weiss and Enzinger as a low-grade angiosarcoma resembling a cavernous hemangioma and Kaposi's sarcoma (Weiss and Enzinger, 1986). Clinically, spindle cell hemangioendothelioma appears as a solitary or multiple cutaneous and subcutaneous nodules, usually involves the upper extremity of young adults, and follows a benign indolent course, but has a recurrent tendency (Weiss and Enzinger, 1986; Scott and Rosai, 1988). Most cases of spindle cell hemangioendothelioma reported are located in the superficial soft tissue and therefore are diagnosed clinically as various benign dermatologic lesions such as epidermal inclusion cyst, sebaceous cyst, hemangioma or dermatofibroma. Furthermore, there has been few cases of spindle cell hemangioendothelioma located in deep soft tissue, which also produce no specific symptoms.

Even though spindle cell hemangioendothelioma has been known as low-grade malignancy, there has been few reported cases of spindle cell hemangioendothelioma with local aggressive and invasive be-

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haviors (Silva et al., 1986; Lai et al., 1991).

We report a case of recurrent spindle cell hemangioendothelioma with unusual clinical behavior in that it affected the underlying bone and caused painful bony erosion, which has not been reported so far as we know.

CASE REPORT

A 20-year-old man visited a local clinic in 1990 for treatment of a nodule on the left ankle. The nodule first appeared on the anterior surface of the left ankle when he was 15 years old. The lesion measured up to 2 cm in diameter and was relatively firm. We could not ascertain the pathologic diagnosis of that lesion since the pathologic examination has not been performed.

The patient was admitted to Capital Armed Forces General Hospital 4 years later with a similar nodule which was incidentally found 1 cm away from the previous one. On palpation, it was non-tender, firm and measured 3 cm in diameter. The excised lesion was diagnosed as a hemangiopericytoma.

The wound healed, but the patient returned 6 months later with pain on the medial malleolus 2 cm away from the second operation site. The pre-operative X-ray revealed a small-sized bony erosion with sclerotic margins on the medial malleolus, but there was no palpable mass (Fig. 1). The bone scan revealed that there was a hot uptake on delayed image in the medial malleolus of the left distal tibia, which was suggestive of osteomyelitis or soft tissue tumor. On operation, there was a 1.5 cm-sized, red soft tissue mass within the undermined bone lesion. However, the medial malleolus appeared normal except for dimpling of the cortical bone. All the laboratory tests were within normal limits.

PATHOLOGIC FINDINGS

Grossly, the second excised specimen was a well circumscribed, oval, grayish white and firm mass, measuring 3.5×2.5×1cm. The cut surface was gray tan and solid. There were no areas of necrosis. The third excised mass within the undermined malleolus was dark brown and firm with irregular margins, measuring 1.5×0.5×0.5cm.

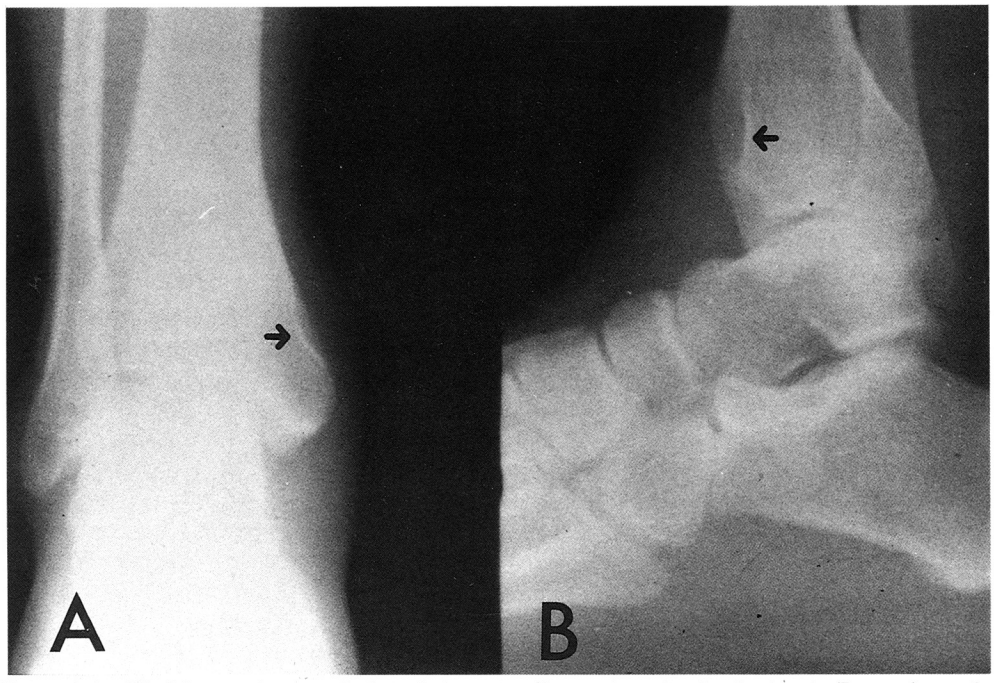

Fig. 1. X-ray of the left tibia reveals a scalloping of the cortex (A, arrow) and sclerotic margin (B, arrow) on the medial malleolus.

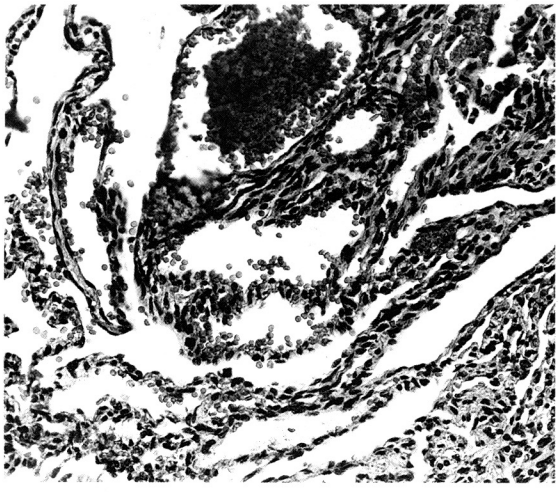

Fig. 2. A portion of tumor illustrating features of typical carvernous hemangioma (×100).

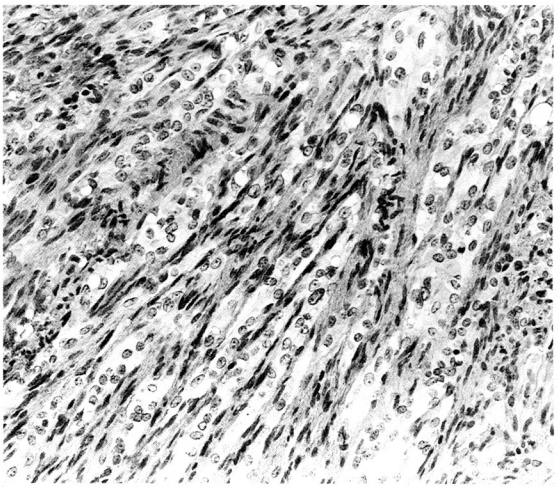

Fig. 4. The spindle cells are intermingled with plump epithelioid cells (×400).

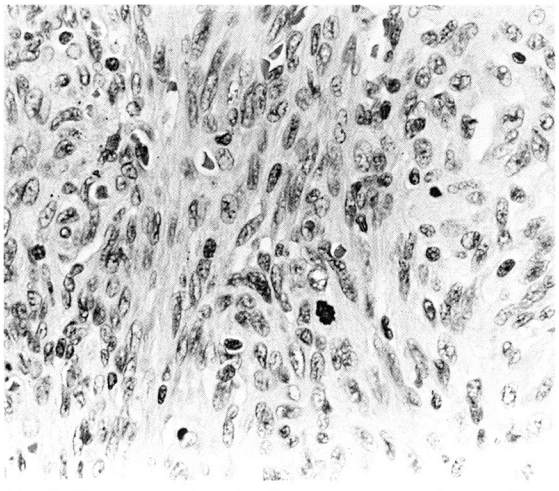

Fig. 3. High-power view of cellular components shows spindle cells with moderate degrees of pleomorphism and mitosis (×400).

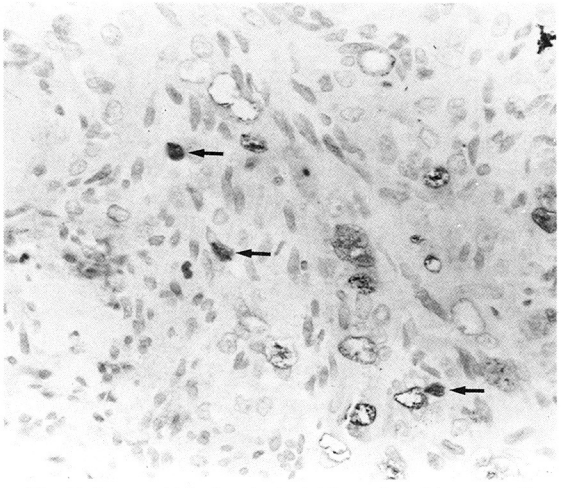

Fig. 5. Immunohistochemical staining of cellular area illustrates the spindle cells and vacuolar cells occasionally stained with anti-Factor VIII-associated antigen antibody (arrows).

Microscopically, the two tumors were basically identical and composed of cavernous endothelial-lined blood spaces and cellular zone with less differentiated spindle cells (Fig. 2, 3). The cellular zones mostly consisted of spindle cells that formed a kaposiform mixture of cleft and round vascular spaces intermingled with a few vacuolated cells and plump epithelioid cells (Fig. 4). Mitoses were occasionally found, and there was a moderate degree of pleomorphism of the spindle cells (Fig. 3). There was neither organized intravascular thrombi nor calcification.

Immunohistochemically, antibodies against factor VIII-associated antigen and vimentin stained most of the endothelial cells lining the cavernous blood spaces and also occasional spindle cells (Fig. 5), which were stained blue on Masson's trichrome staining. The epithelioid cells with plump cytoplasm were negative for factor VIII-associated antigen as well as

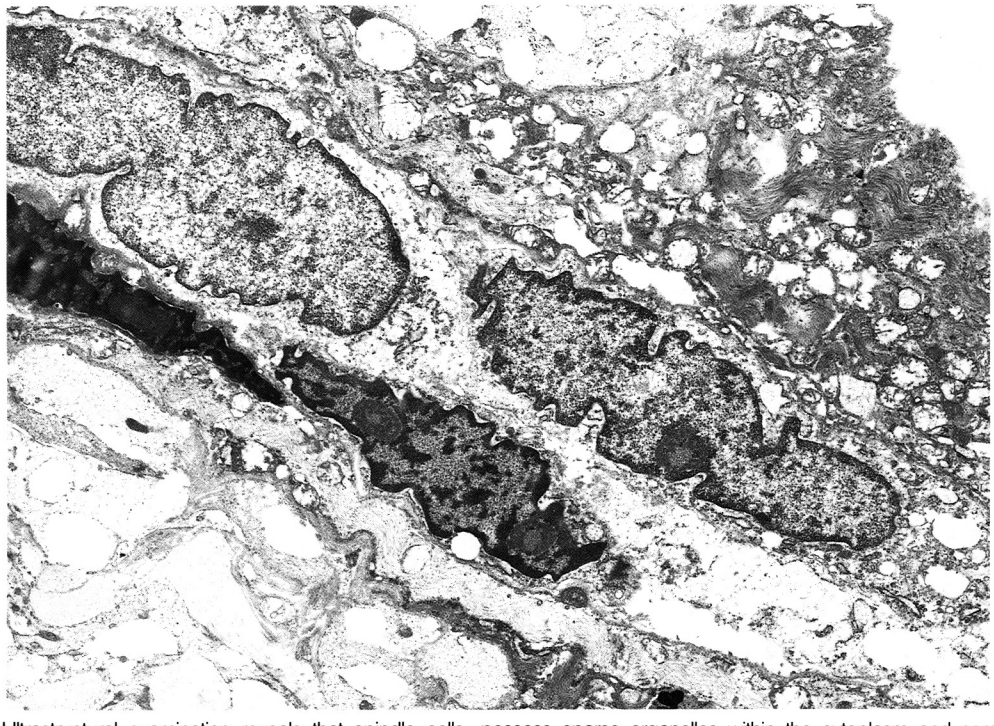

Fig. 6. Ultrastructural examination reveals that spindle cells possess sparse organelles within the cytoplasm and some intermediate filaments (×8.600)

epithelial membrane antigen (EMA). Desmin and S-100 protein were not recognized within all kinds of tumor cells.

Ultrastructurally, the cells lining vascular channels possessed well formed basal lamina and collections of intermediate filaments, suggesting endothelial origin (Fig. 6). However, Weibel-Palade bodies and hemosiderin granules were not noted. The spindle cells possessed sparse organelles within the cytoplasm and did not have basal lamina, which appeared to be the primitive mesenchymal cells.

DISCUSSION

Spindle cell hemangioendothelioma is a rare vascular tumor and characterized by the combined histologic features of Kaposi's sarcoma and cavernous hemangioma. The amounts of these components vary from lesion to lesion and from case to case. A typical lesion is composed of small nodules in the dermis or subcutis which seem to be well demarcated but give rise to tiny capillary sprouts at their periphery (Weiss and Enzinger, 1986). In this case the spindle cell components are more abundant and the tumor is located in the deeper portion compared with other more typical cases.

Although the spindle areas, out of context, resembled Kaposi's sarcoma, they differed principally by the focal presence of plump or round epithelioid cells as well as cavernous hemangiomatus areas (Weiss and Enzinger, 1986). The epithelioid cells were forming small nests or lining vascular channels with sometimes vacuolar changes, generally regarded as intracytoplasmic lumen formation by single cell. However, the epithelioid cells were negative for Factor VIII-associated antigen and EMA like epithelioid cells of the epithelioid hemangioendothelioma, whereas all carcinomas and epithelioid sarcoma are usually positive for EMA.

There are different views on the nature of the spindle cells, which have been suggested to represent primitive endothelial cells (Weiss and Enzinger, 1986) or pericytes (Scott and Rosai, 1988) or fibroblasts (Imayata, 1992). On ultrastructural and clinical

study, Imayata (1992) described that spindle cell hemangioendothelioma may develop from a cycle of recanalization after thrombosis and most of the cells appear simply to be fibroblasts with features of pericytes. Whereas the ultrastructural features of the spindle cells in the present case appeared to be those of primitive mesenchymal cells, the spindle cells were reactive for factor VIII-associated antigen on immunohistochemical study, which is suggestive of vascular origin. The immunohistochemical analysis of the spindle cells with factor VIII-associated antigen and Ulex europaeus agglutin 1 (UEA-1) vary from study to study (Weiss and Enzinger, 1986; Povysil and Janousek, 1993; Murakami et al., 1993).

Even though the clinical behaviors of spindle cell hemangioendothelioma are marked by numerous local recurrences and slow but steady progression of the disease, the lack of cellular atypism and no distant metastasis have raised questions regarding the biological nature of spindle cell hemangioendothelioma. Eltorky et al. (1994) suggested that spindle cell hemangioendothelioma would be a non-neoplastic lesion and not a neoplasm of borderline malignancy on the basis of the facts that there are the repeated presence of organized intravascular thrombi, early clinical presentation in life and presence of some degree of vascular malformation at the periphery of the lesion. However, considering its clinical behaviors and histologic findings, it is unlikely that this case would be attributed to reactive vascular changes. The tumor developed skipped recurrent masses around the left ankle, eroded the underlying medial malleolus of the left tibia, and possessed the histologic features of moderate cellular pleomorphism and occasional mitoses in the spindle cellular areas but no organized intravascular thrombi. Whereas it has been well known through extensive pathologic studies (Weiss and Enzinger, 1986; Scott and Rosai, 1988) that spindle cell hemangioendothelioma is usually lack of cellular atypism and mitoses, the cellular components of the present case showed several foci of moderate degrees of cellular pleomorphism and mitotic activity, which would be related to its aggressiveness in biological behavior. There have also been few cases of spindle cell hemangioendothelioma with aggressive clinical behavior such as local metastasis(Silva et al., 1986; Lai et al., 1991). Despite the occurrence of local metastases, this tumor has not progressed to widespread dissemination, which clinically sets it apart from epithelioid hemangioendothelioma (Weiss and Enzinger, 1982; Lai et al., 1991). It is likely that requiring metastases in the diagnostic criteria for the malignancy may result in failure to recognize this entity as low-grade malignant or borderline lesions.

In summary, we have presented a case of spindle cell hemangioendothelioma that developed recurrent painful nodules around the left ankle and showed aggressiveness in biological behavior and some pleomorphism and mitoses in histologic aspects.

REFERENCES

Eltorky M, McC-Chesney T, Sebes J, Hall JC. *Spindle cell hemangioendothelioma : Report of three cases and review of the literature. J Dermatol Surg Oncol 1994, 20; 196-202.*

Imayata S, Murakamai Y, Hashimoto H, Hori Y. *Spindle cell hemangioendothelioma exhibits the ultrastructural features of reactive vascular proliferation rather than of angiosarcoma. Am J Clin Pathol 1992, 97; 279-87.*

Lai FM-M, Allen PW, Yeun PM, Leung PC. *Locally metastasizing vascular tumor spindle cell, epithelioid, or unclassified hemangioendothelioma? Am J Clin Pathol 1991, 96; 660-3.*

Murakami I, Sarker AB, Teramoto N, Horie Y, Taguchi K, Akagi T. *Spindle cell hemangioendothelioma. a report of two cases. Acta Pathol Jpn 1993, 43; 529-34.*

Povysil C, Janousek M. *Spindle cell hemangioendothelioma. Cesk Pathol 1993, 29; 140-3.*

Scott GA, Rosai J. *Spindle cell hemangioendothelioma report of seven additional cases of a recently described vascular neoplasm. Am J Dermatopathol 1988, 10; 281-8.*

Silva EG, Phillips MJ, Langer B, Ordonez NG. *Spindle and histiocytoid(epithelioid) hemangioendothelioma primary in lymph node. Am J Clin Pathol 1986, 85; 731-5.*

Weiss SW, Enzinger FM. *Epithelioid hemangioendothelioma. A distinctive vascular tumor often mistaken for carcinoma. Cancer 1982, 50; 970-81.*

Weiss SW, Enzinger FM. *Spindle cell hemangioendothelioma a low-grade angiosarcoma resembling a cavernous hemangioma and Kaposi's sarcoma. Am J Surg Pathol 1986, 10; 521-30.*