Primary pulmonary hemangiopericytoma

Hemangiopericytoma is a potentially malignant tumor of vascular origin. It usually is a solitary lesion and may arise in the chest or in extrathoracic areas. It is usually encapsulated. Microscopic features vary with the degree of differentiation of the individual tumor. The more differentiated variety exhibits abundant capillaries with open lumina surrounded by ovoid tumor cells; in the less differentiated tumors, the capillaries lack lumina and the tumor cells are spindle shaped. The prognostic significance of the microscopic pattern is controversial. However, prominent mitotic activity, necrosis, hemorrhage, and increased cellularity are ominous signs and are usually noticed in tumors that later exhibit malignant behavior. The larger the lesion, the more likely that it will be symptomatic. Treatment of choice is ample surgical resection.

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Hemangiopericytoma is an uncommon, potentially malignant tumor of vascular origin. Although it may arise in the lung and mediastinum, in most reported cases the lesion was located outside the thorax, with definite musculoskeletal predilection. In the lung, the tumor may present as a small asymptomatic nodule, usually discovered on a routine chest x-ray film, or as a large symptomatic lesion. For an intraoperative diagnosis to be made, the possibility of this tumor existing must be appreciated during frozen-section examination. The following case is reported to call attention to this tumor and to emphasize the value of accurate recognition, because the malignant potentiality of the lesion has a bearing on appropriate therapy.

Case report

A 54-year-old man was admitted to the hospital because of a well-circumscribed density in the lower lobe of the left lung discovered on routine chest roentgenograms. He had no symptoms related to the mass. Results of physical examination and laboratory tests were essentially within normal limits. Findings from bronchoscopic and mediastinoscopic examinations were normal. At thoracotomy, the mass was well rounded and measured 2.5 by 2 cm. in diameter. It was located in the periphery of the left lower lobe, and the overlying pleura was not puckered. Wedge resection was performed. Grossly, the mass was somewhat firm. On cut section, it appeared to be an encapsulated, pale-tan tumor with focal areas of hemorrhage. The report from frozen section examination was "benign spindle cell tumor," and wedge resection was thought adequate for such a lesion. However, permanent sections indicated hemangiopericytoma.

Microscopically, there were numerous capillaries lined with flattened endothelium and surrounded by sheets of tumor cells resembling pericytes (Fig. 1). The tumor cells were round or oval, possessed pale cytoplasm and vesicular nuclei, and showed occasional mitoses. Reticulin stain showed a fine network radiating from the vascular channels and encircling either individual tumor cells or small clusters of cells (Fig. 2).

The electron microscope showed that the endothelial cells and the tumor cells were surrounded by continuous basal lamina giving the appearance of the capillaries being surrounded by a double basement membrane (Fig. 2).

Two years after the operation, the patient is alive, well, and free of evidence of clinical disease.

Discussion

Tumors of vascular origin are subdivided into two groups: those composed of pericytes, the hemangiopericytoma and glomus tumor, and those composed of endothelial cells, the hemangiendothelioma. Analogous counterparts are lymphangioma and lymphangiosarcoma that arise from lymph vessels.

Hemangiopericytoma has been described as a mesenchymal neoplasm of vascular origin characterized by numerous capillaries surrounded by tumor cells and a rich reticulin network. Macroscopically, the tumor varies from light gray to tan and frequently

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**Fig 1.** *Left.* Photomicrograph showing numerous well-developed capillaries surrounded by sheets of tumor cells (pericytes). *Right.* Reticulin network capillaries and tumor cells. (Hematoxylin and eosin. Original magnifications: *left,* ×70; *right* ×250.)

**Fig. 2.** Electron micrograph showing endothelial cells (*1*) and tumor cells (*2*) surrounded by a continuous basal lamina giving the appearance of the capillaries being surrounded by a double basement membrane (*arrows*). (Original magnification ×14,950.)
exhibits areas of hemorrhage and necrosis. A capsule is present but may be incomplete. Owing to its variegated histologic appearance, hemangiopericytoma should be differentiated from bronchial adenoma, reticulin cell sarcoma, vascular fibrosarcoma, and various vascular, histiocytic, neural, and smooth muscle tumors. The lack of bronchial communication, the presence of more extensive hemorrhage, necrosis, and the presence of a rich reticulin network in hemangiopericytomas help in differentiating it from these other tumors.

Clinically, the larger the lesion is, the more likely that it will be symptomatic and the worse the prognosis will be. Surgery is the treatment of choice because of the tumor’s malignant potential, unpredictable behavior, and tendency to local recurrence. If the extent of the lesion allows, ample resection should be performed. A prolonged period of follow-up is essential, because the tumor may recur up to 20 years following excision, as observed in extrathoracic lesions.

Experience with radiotherapy for primary pulmonary hemangiopericytoma has been minimal. It has been useful in palliation of pulmonary metastases and relief of superior vena cava obstruction. Its use as an adjunct to surgery has been recommended in cases of recurrence, if symptoms in inoperable lesions demand palliation, or preoperatively in larger lesions involving the chest wall where diagnosis of hemangiopericytoma has been established. Experience with chemotherapy has been limited and disappointing.

References