Abstract.—A 40-year-old female was examined for complaints of left arm pain and restriction of movement in the left upper extremity for 3 months. Chest X-ray showed a mass in the left upper lung and the patient was evaluated with computed tomography that confirmed the significant mass in the left paratracheal region and also showed small nodules in both of the lungs. A whole-body FDG-PET scan was performed for the suspicion of malignancy. FDG-PET indicated high FDG accumulation in the lung lesions mainly in the left paratracheal region. FDG-PET findings were highly suspicious of malignancy so the patient had thoracoscopic biopsy of the lesion. The histological findings and immunohistochemistry tests were consistent with pulmonary epitheloid hemangioendothelioma (PEH). Epitheloid hemangioendothelioma (EH) is a systemic name that represents a rare type of malignant tumor of vascular endothelial origin, which can arise in bone, liver, soft-tissue, or lung. PEH is currently known as the lung form of EH. Consequently, our patient had resection of the left paratracheal mass.

This report presents a rare case of histologically confirmed PEH, which showed increased FDG accumulation on FDG-PET study. PEH should be added to the other causes of increased FDG uptake.

Key words: pulmonary epitheloid hemangioendothelioma, FDG, PET.

Increased FDG uptake in pulmonary epitheloid hemangioendothelioma

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INTRODUCTION

Epitheloid hemangioendothelioma (EH) is a systemic name that represents a rare type of malignant tumor of vascular endothelial origin, which can arise in bone, liver, soft-tissue, or lung. In most cases only a single organ is involved at presentation. Multigorgan involvement occurs occasionally (especially liver and lungs) and may represent metastatic disease or multicentric origin of the disease. Pulmonary epitheloid hemangioendothelioma (PEH) was initially termed as “intravascular bronchoalveolar tumor” (IVBAT) and considered to be an unusual form of bronchoalveolar malignancy as described by Dail and Liebow. By immunohistochemical and ultrastructural studies, IVBAT was subsequently established to be pathologically identical to the EH of soft-tissue or liver. It is now known as the lung form of EH and named as PEH. PEH typically occurs in women and manifests as bilateral parenchymal nodules.
Since growing malignant cells with higher glucose metabolism accumulate $^{18}$F-fluorodeoxyglucose (FDG) to a greater extend, FDG-Positron Emission Tomography (PET) imaging has been successfully used in the evaluation of patients with known or suspected malignancies\(^{10,11}\).

This report presents a rare case of histologically confirmed PEH, which showed increased FDG accumulation on FDG-PET study. The etiology, diagnosis, therapy, and prognosis of this rare condition are also discussed.

**CLINICAL CASE**

A 40 year-old female had complaints of left arm pain and restriction of movement in the left upper extremity for 3 months. Her physical examination revealed pain while moving the left arm. Laboratory tests were in normal limits.

Chest radiograph showed a mass in the left upper lung. The patient had no symptoms of chest pain, dyspnea or hemoptysis. Then, the patient was evaluated with CT that confirmed the significant mass in the left paratracheal region (fig. 1). Small nodules in both of the lungs were also noted (fig. 2).

She was referred to PET Center to be evaluated with FDG-PET for the suspicion of malignancy. A whole-body FDG-PET scan was performed, and approximately 60 min after intravenous injection of 370 MBq of FDG, whole-body PET scanning of the neck through pelvis (10 min/bed position) was obtained using an EXACT/HR scanner (CTI PET Systems, Knoxville, TN). Images were reconstructed with attenuation correction. FDG-PET revealed high FDG accumulation in the lung lesions mainly in the left paratracheal region (figs. 3, 4). FDG uptake in the center of this mass was lower when compared to the periphery of the lesion (ring like appearance). Since the FDG-PET findings were highly suspicious of malignancy, the patient had thoracoscopic biopsy from the lesion. The histological findings and immunohistochemistry tests were consistent with PEH.

Thereafter, the patient had resection of the left paratracheal mass. After the surgery, she had neither any symptoms nor progression of the other lung nodules for a follow-up period of 6 months.

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Fig. 1.—Transaxial CT images demonstrate the large mass in the left paratracheal region (black arrows).

Fig. 2.—(A-B) Transaxial CT images illustrate the small nodules in the lungs bilaterally (white arrows).

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DISCUSSION

PEH is an unusual neoplasm that presents as multiple small nodules in lung parenchyma less than 2 cm in diameter. Occasionally, this tumor might be seen as a solitary pulmonary mass measuring up to 5 cm. The majority of patients with PEH are females and the age range for diagnosis is 20-60 years. Patients may be asymptomatic and detected by abnormal chest radiography or there might be non-specific symptoms as chest pain, cough and sputum. There is no characteristic clinical or biological marker for PEH. PEH usually manifests radiologically multiple pulmonary nodules. Calcification is histologically common but it is rarely observed on chest radiographs or CT scans. Pleural effusions and hilar adenopathy can be seen in 9% of patients. Definitive diagnosis requires biopsy. Macroscopically PEH nodules (several millimeters to 5 cm) are usually rubbery or cartilage-like and the cut surface is grey-white to yellow-brown. Rarely, calcifications are observed. Histologic spectrum ranges from low-grade to higher-grade lesions according to the number of mitotic activities and necrosis. The periphery of the neoplasm is hypercellular and the center is hypocellular with coagulative necrosis, hyalinization, calcification. The tumor cells are round with abundant cytoplasm, while the nucleus is round or oval. The cell growth may form lumens of various sizes. Immunohistochemistry tests show positive immunohistochemistry for endothelial cell markers, such as factor VIII-related antigen, Ulex and CD4. Electronmicroscopy demonstrates features of epithelial vascular endothelium. It has been proposed that PEH can grow in a peculiar fashion by extending from alveolus to alveolus through pores of Kohn. Dis-
semination might be via vascular spread or lymphatic route. In contrast, it has also been suggested that involvement of multiple organs might be due to multicentric origin of the disease. There are currently no effective therapies for PEH. Radiation and multiple chemotherapeutic agents have shown to be non-effective. Surgical resection has been recommended if the lesion is solitary or the number of lesions is limited. In addition to surgery for localized lesions of EH, therapy using α-2A interferon, interleukin-2, and retinoids have been suggested. Following the asymptomatic patients with no therapy has been recommended also. Partial spontaneous regression in a few cases was reported. Prognosis of this tumor is unpredictable. Survival time of the patients reported in the literature is between 2 and 24 years. Patients with clinical symptoms, peripheral adenopathy, hepatic tumor, extensive intravascular, pulmonary or pleural involvement have poor prognosis. Death is usually from respiratory failure or complications of extrathoracic tumor.

Rapidly growing malignant cells with higher glucose metabolism accumulate FDG to a greater extent. FDG is an analogue of glucose and it is a tracer of energy substrate metabolism. Increased glucose transporters have been proposed as one of the reasons of the pathologic preparation. The ring like increased FDG uptake in the left paratracheal lymph node, the increased FDG uptake in the left paratracheal lymph nodes observed on CT images. Since the increased FDG uptake in the periphery of the lesion might be due to hypercellularity and the hypoactive area in the center of the lesion and could be related with hypocellularity and coagulative necrosis as mentioned above in the histological definition of PEH. FDG-PET also showed the multiple bilateral lung nodules of PEH in the patient (fig. 4).

In conclusion, this article describes a case of bilateral pulmonary epithelioid hemangioendothelioma along with a description of this extremely rare pulmonary tumor. We add to the literature a PEH case, which showed increased FDG accumulation. PEH should be considered among other causes of increased FDG uptake in the lungs which are listed in table 1.

Table 1

<table>
<thead>
<tr>
<th>Common causes</th>
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<td>Smoking</td>
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REFERENCES


