Sclerosing Pneumocytoma with Lymph Node Metastasis

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A 33-year-old woman with a family history of multiple cancers had been undergoing surveillance scans. Magnetic resonance imaging revealed cysts in the kidney and pancreas, multiple hemangiomas in the liver, and a lesion in the left lower lobe (LLL) of the lung. Repeat positron emission tomogram/computed tomography demonstrated a 1.8 × 1.6-cm nodule (Fig. 1) within the LLL.

The patient underwent endobronchial ultrasound-guided fine needle aspiration biopsy of the LLL nodule, which showed hypercellular smears containing uniform, cuboidal, round, and oval medium-sized cells. They were diffusely positive for thyroid transcription factor-1, and negative for neuroendocrine cell markers. The diagnosis was well-differentiated lung adenocarcinoma. The patient underwent video-assisted thoracoscopic LLL lobectomy, which showed a well-circumscribed 2.5-cm yellow-tan nodule composed of mostly monomorphic, cuboidal surface cells and round cells with pink to clear cytoplasm and well-defined cell borders in papillary, solid, and sclerotic growth patterns. Occasional hemorrhagic foci were also present. A focal area of mild cytologic atypia was noted. A focus of metastatic tumor deposit containing solid sheets of cells was identified in one of the peribronchial lymph nodes. The results of staining for thyroid transcription factor-1 and epithelial membrane antigen were positive in both surface and round cells, but cytokeratin 7 and cytokeratin AE1/AE3 stained only surface cells (Fig. 2). The cytomorphologic examination findings and immunoprofile were consistent with sclerosing pneumocytoma (SP) with a lymph node metastasis.

SP (previously called sclerosing hemangioma) is a rare tumor of the lung that is thought to arise from primitive respiratory epithelium. This tumor usually occurs in middle-aged adults with a male-to-female ratio of 1:5.1 Surgical resection is curative without a need for additional treatment. Although SP is considered a benign tumor, we found a lymph node (LN)
metastasis in the present case, indicating that SP might have a low malignant potential. Although this finding is not entirely unique, only rare cases of metastasis to regional LNs have been reported. In the largest series of published cases, which included 100 patients, only one patient was found to have LN metastasis.¹ A comprehensive literature review identified a total of 19 cases of SP with LN metastasis.¹⁻³ The male-to-female ratio was higher (1:1.3 versus 1:5) and the lower lobe was the more common tumor location (68% versus 23.5%) in metastasizing SP. Similarly, most of the SPs with LN metastasis were 3 cm or larger as opposed to the ones without LN metastasis (72% versus 26%). However, survival data are available for a very limited number of patients. In reports involving four patients² and one patient,³ LN metastasis did not affect the long-term survival (follow-up time range 2–10 years). Long-term surveillance of these patients would be beneficial to learn the behavior of this rare entity, especially the SPs with LN metastasis. Our patient is doing well 1 month after surgery and is enrolled in an annual surveillance program.

Figure 2. (A) Tumor nodule showing papillary structures lined by cuboidal surface cells and round stromal cells (hematoxylin and eosin [HE] staining; original magnification, ×200). (B) Tumor nodule showing sheets of mostly round cells representing solid areas (HE staining; original magnification, ×200). (C) Tumor nodule showing sclerotic areas infiltrated by strands of neoplastic cells (HE staining; original magnification, ×200). (D) Lymph node involvement by the tumor showing solid growth pattern (HE staining; original magnification, ×100). (E) Immunohistochemical staining for thyroid transcription factor-1 shows diffuse staining involving both surface cells and round cells (original magnification, ×100). (F) Immunohistochemical staining for cytokeratin 7 shows selective staining involving only surface cells (original magnification, ×100).
References

