

Understanding the Uncommon Thoracic Tumors

Thoracic tumors continue to account for the highest number of cancer deaths in the United States. Non-small cell lung cancer (NSCLC) is the most common, followed by small-cell lung cancer (SCLC). This year, the American Cancer Society estimates that about 175,000 new cases of thoracic tumors will be diagnosed and 160,000 people will die of tumors of lung and bronchus.¹ In addition to these common thoracic tumors, several less common tumors arise from other thoracic cavity structures, including pleura, airway, mediastinum, and chest wall. The lung tissue itself can also harbor less common forms of tumors such as carcinoid and large-cell neuroendocrine carcinoma. A rough estimate indicates that this heterogenous cluster of uncommon thoracic tumors may constitute up to 5% of all thoracic tumors (Table). This in itself makes the word “uncommon” indeed debatable.

In this issue of *Cancer Control*, we explore the road less traveled: five selected uncommon thoracic tumors are reviewed by multidisciplinary experts in their particular field. The first article, by Dr Ismail-Khan and colleagues, provides an up-to-date summary of malignant pleural mesothelioma. Current state-of-the-art treatment is comprehensively reviewed, including selection criteria for extrapleural pneumonectomy and promising agents still under investigation. In the second article,

Dr Robinson focuses on a solitary fibrous tumor of the pleura, the less aggressive form of pleural tumors. Its unique radiologic characteristic and clinical course are concisely presented. The third article is dedicated to large-cell neuroendocrine carcinoma, the less common form of NSCLC. Drs Fernandez and Battafarano review the classification of pulmonary neuroendocrine tumors, the prognosis of large-cell neuroendocrine carcinoma, and the implication for its management. In the fourth article, Dr Taveira-Dasilva and colleagues provide a comprehensive review of lymphangioliomyomatosis (LAM), a tumor of smooth, muscle-like cell originating within the lung tissue. Its perplexing pulmonary cyst formation and extrapulmonary manifestation are vividly illustrated along with promising ongoing research. The last article in this series of uncommon thoracic tumors covers tracheobronchial tumors, including mucinous cystadenoma, pleomorphic adenoma, mucoepidermoid carcinoma, and adenoid cystic carcinoma. Drs Gaissert and Mark describe the unique features of these tumors and the success with segmental tracheal or bronchial resection.

While these articles cover most of the less common thoracic tumors, other infrequent but important thoracic tumors include carcinoid tumor, thymic tumor and diffuse idiopathic pulmonary neuroendocrine cell hyperplasia (DIPNECH). The pulmonary carcinoids, both typical and atypical, are part of the spectrum of pulmonary neuroendocrine tumors; both have a better prognosis than SCLC and large-cell neuroendocrine tumor. Surgery is the cornerstone of treatment, and somatostatin receptor scintigraphy, though generally useful in gastroenteropancreatic carcinoids, can also be helpful for staging. For thymic tumors, many studies have now shown that unresectable disease may be cured by treatment with induction chemotherapy followed by radiation, while

Estimated Incidence of Selected Uncommon Primary Thoracic Tumors in the United States

Thoracic Tumors	Estimated New Cases Per Year (Estimated % of All Thoracic Tumors)
Pleura:	
Malignant pleural mesothelioma	2,500 (1.4%)
Solitary fibrous tumor of the pleura	Unknown, <800 cases reported in the literature
Lung:	
Large cell neuroendocrine carcinoma	5,000 (2.8%) ²
Lymphangioliomyomatosis	Unknown, prevalence 1,000–1,500 cases
Bronchopulmonary carcinoid tumors and DIPNECH	300 (0.2%)
Lymphoepithelioma-like carcinoma of the lung	Unknown, <150 cases reported in the literature ⁶
Trachea and bronchus:	
Tracheobronchial gland tumors	300 (0.2%)
Mediastinum:	
Thymic tumors	450 (0.3%) ³
Neurogenic tumors	450 (0.3%) ⁴
Chest wall:	
Sarcomas	800 (0.4%) ⁵

recurrent disease may respond to octreotide with or without prednisone.^{7,8} DIPNECH is another interesting entity. A premalignant precursor of carcinoid tumors, DIPNECH may present with a radiographic finding mimicking metastatic cancer. Additional articles on these fascinating tumors are provided in the Ten Best Readings department in this issue.

Also included in this issue is an article on Community Clinical Oncology Programs (CCOPs) that target how best to recruit healthy individuals for participation in cancer prevention research. Dr McKinney and colleagues outline how to develop cancer prevention research in our community. CCOPs represent a new model of academic-community cooperation that, with adequate financial and technical support, can broaden community participation in preventive research. Lastly, we continue our "Cancer, Culture and Literacy" department, this time with an article that summarizes the important issues discussed at the May 2006 Cancer, Culture and Literacy Conference — "Solutions for Addressing Health Disparities Through Community Partnerships." The sessions focused on conceptual and methodological issues in cancer health disparity research within the context of culture and literacy.

In summary, the uncommon intrathoracic tumors presented in this issue may pose a challenge to both diagnosis and treatment if we are unfamiliar with them. Certainly they pose a problem with regard to clinical trial enrollment and drug development, leading to a slow progress in the understanding and treatment. One solution to this is an establishment of a central database such as the LAM registry. The details of this exciting project, which is supported by the National Institutes of Health, can be accessed at <http://www.thelamfoundation.org>. This worthwhile endeavor has led to a strong patient support group, a collaboration of researchers in this area, and an enthusiastic group of advocates who are raising funds for future research. Uncommon as these tumors may be in the overall population, the condition is personally vital to the affected individual. I am sure you will find this issue of *Cancer Control* worthwhile. Indeed, some of the illustrations might last in your memory for a lifetime.

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