CHAPTER 44

PLEURAL TUMORS

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Pleura-associated neoplasms develop from the thin, double layer of tissue that surrounds the lungs, mediastinum, chest wall and diaphragm. Primary tumors of the pleura are rare. Malignant or diffuse tumors occur more frequently than benign or localized tumors and comprise mesothelial cells. The diagnosis or the pleural tumors is clinched by combined radiologic examination and clinical presentation. They may cause uncomfortable symptoms and are difficult to treat.

The benign and malignant forms of the pleural tumors are listed in Table-1.

BENIGN PLEURAL TUMORS

Benign tumors of the pleura comprise approximately 5% of all the pleural neoplasms. Radiologic properties may mimic malignant pleural cancers, and it is crucial to distinguish them from each other. Therefore, histological diagnosis by a *surgical excision* is essential before planning the treatment. It is essential to obtain greater quantity of tissue samples during the biopsy procedure. Total excision of the tumor is often curative; however recurrence or transformation to malignancy may occur during or after the follow-up period.

Table-1. Benign and Malignant Pleural Tumors.

Benign	Malignant
Solitary Fibrous Tumor	Malignant Mesothelioma
Lipoma	Malignant Solitary Fibrous Tumor
Lipoblastoma	Localized Malignant Mesothelioma
Adenomatoid Tumor	Vascular Sarcoma
Calcifying Fibrous Tumor	Pleuropulmonary Synovial Sarcoma
Mesothelial Cyst	Primitive Neuroectodermal Tumor (PNET) / Askin Tumor
Multicystic Mesothelial Cyst	Desmoplastic Small Round Cell Tumor
Schwannoma	Epitheloid Hemangioendothelioma
Thymoma	Primary Effusion Lymphoma (PEL)
Nodular Plaque	Diffuse Large B-cell Lymphoma with Chronic Inflammation
Desmoid Tumor	Desmoid Type Fibromatosis
	Pleuropulmonary Blastoma
	Liposarcoma
	Metastatic Malignant Tumor

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RARE MALIGNANT TUMORS OF THE PLEURA

MPM consists of the majority of primary pleural malignant tumors. However, some rare malignant pleural tumors can also be detected [39]. Complete surgical resection, if possible, is recommended for cure.

Malignant Solitary Fibrous Tumor: 10% of SFTPs may show malignant formation. Malignant forms demonstrates more cellularity. Dedifferentiation can be seen in rare samples of malignant SFTP.

Localized Malignant Mesothelioma: Although it shows ultrastructural, immunohistochemical and microscopic properties as diffuse MPM, it does not spread diffusely.

Vascular Sarcoma: It is a malignant tumor of endothelial origin and characterized by a vascular pattern. Rare cases with asbestos exposure and pyothorax have been reported. Hemothorax is a serious complication.

Synovial Sarcoma: Although they usually appear as localized solid tumors, they can also cause diffuse pleural thickening.

Primitive Neuroectodermal Tumor (PNET): It is a small, round blue cell tumor and is a member of the Ewing sarcoma family. It is also known as Askin tumor. Thoracopulmonary location of PNET is very rare.

Desmoplastic Round Cell Tumor: This is a malignant mesenchymal tumor composed of cells in round cell morphology. It is commonly encountered in young adolescents and especially in men.

Epithelioid Hemangioendothelioma: Cases may present with pleural thickening, effusion and/or pleuritic pain. In addition to prgressive focal pleural insemination, metastases to the lung, liver and regional lymph nodes are also common.

Primary Effusion Lymphoma (PEL): PEL presents with effusion in serous cavities. Kaposi sarcoma associated herpes virus (KSHV), also called human herpesvirus 8 (HHV8), are rare tumors consisting of large, atypical B cells. While there is usually no solid mass at the time of diagnosis, it may occur subsequently.

Diffuse Large B-cell Lymphoma with Chronic Inflammation: B cell lymphoma mostly associated with *Epstein-Barr* virus EBV. It is usually associated with long-term chronic inflammation in serous cavities or other anatomical regions close to restricted vascularization areas. The interval between initial inflammation and the appearance of lymphoma can be more than 10 years.

Desmoid Type Fibromatosis: It is a locally aggressive but non-metastatic myofibroblastic tumor. It typically originates from the deep soft tissue. It can be classified in this section due to its frequent and aggressive recurrence.

Miscellaneous: Pleuropulmonary blastoma and liposarcoma are other malignant tumors of the pleura.

METASTATIC PLEURAL TUMORS

Malignancies from extrathoracic structures may metastasize to the pleura frequently. Malignities from breast, lung, ovary, stomach and lower gastrointestinal system, kidney, as well as lymphoma, various sarcomas and melanoma are the most encountered tumors that exhibit metastases to the pleura. Sometimes, the primary focus may not be detected.

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