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Mediastinal schwannoma: A clinical, pathologic, and imaging review

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Dear Editor,

Neurogenic tumors represent approximately 20% of all adult tumors and 35% of all pediatric mediastinal tumors and are the most common cause of a posterior mediastinal mass.[1] Forty-five percent of schwannomas occur in the head and neck, with 9% occurring in the mediastinum. Schwannoma is the most common (about 50%) mediastinal neurogenic tumor. It is also known as neurilemmoma, neurinoma, or perineural fibroblastoma. It is a rare benign neural tumor, arising from the neural sheath Schwann cells of the peripheral, cranial, or autonomic nerves. This article reviews the clinical, pathologic, and imaging findings of this rare entity.

Some mediastinal tumors and other masses have been most often found in particular mediastinal locations; therefore, many authors have artificially subdivided the area for better descriptive localization of specific lesions. Most commonly, the mediastinum is subdivided into three spaces or compartments (i.e., anterior, middle, and posterior) when discussing the location or origin of specific masses or neoplasms. Historically, in adults, the most common type of mediastinal tumor or cyst found is the neurogenic tumor (21%-usually in posterior mediastinum), followed by thymic tumors (19%-anterior mediaastinum), lymphomas (13%), and germ cell tumors (10%). Foregut and pericardial cysts are the next most frequently occurring abnormality within this group. More recent data from several large series indicate that thymomas have become the most common mediastinal tumor.

Several tissues, including the neural tissue, neural sheath tissue, and associated fibrous connective tissue of mesodermal origin, can be the source of these neoplasms.[2] More than 50% of the malignant schwannomas are found in patients with neurofibromatosis.[1]

Approximately two-thirds of the mediastinal tumors and cysts are symptomatic in the pediatric population, whereas only approximately one-third of the tumors produce symptoms in adults. This is most likely related to the fact that a mediastinal mass, even a small one, is more likely to have a compressive effect on the small and flexible airway structures of a child. When considering all age groups, nearly 55% of patients with benign mediastinal masses are asymptomatic at presentation, compared with only approximately 15% of those in whom masses are found to be malignant. On gross examination of the surgical specimen, it is usually solitary, encapsulated, and sharply demarcated from the adjacent soft tissue. Nerve of origin is often present in the periphery and does not penetrate the substance of tumor; thus, the tumor may appear to "hang" from large nerves. Large tumors may be cystic. It is also famous as "dumbbell tumor" as in posterior mediastinum, which originates from or extends into the vertebral canal. The tumor has usually yellow cut surface and is rarely dark red/black due to hemorrhage. Microscopically, it is composed of Schwann cells within a background of loose reticular tissue. Areas designated as Antoni A are more organized and are composed of cellular spindle cells arranged in short bundles or interlacing fascicles. Antoni B areas are hypocellular and contain more myxomatous loosely arranged tissue.[3] Mitoses are rare in benign schwannomas. Immunohistochemical staining of the cytologic specimen is positive for S-100 and negative for pan-cytokeratin, cluster of differentiation (CD) 34, CD117, calcitonin, smooth muscle actin, and desmin. A characteristic feature of malignant schwannomas is their ability to exhibit other cellular components such as clusters of epithelial cells, mucin-secreting glands, and even mesenchymal features such as bone, cartilage, or skeletal muscle.

Malignant transformation can occur even without neurofibromatosis.[4] Transformation usually occurs to malignant peripheral nerve sheath tumor (MPNST), angiosarcoma, or

epithelioid malignant change. There is a tendency of schwannomas to show epithelioid cytomorphology when they undergo malignant change. Most common sites are limb, limb girdles, head, and neck. Five-year survival in such cases is <20%.

Imaging studies are an important tool in diagnosis and predicting the behavior of the mediastinal tumors. Chest radiography[5] posteroanterior (PA) for an unrelated cause is the usual means by which an asymptomatic mediastinal mass is identified. Lateral view is particularly helpful for determining the compartment of the mediastinum involved. Computed tomography (CT) scan can greatly assist in determining the exact location of the mediastinal tumor and in determining its relationship to adjacent structures. It is also useful in differentiating masses that originate in the mediastinum from those that encroach on the mediastinum from the lung or other structures. Magnetic resonance imaging (MRI)[5] provides superior vascular imaging and can help better delineate the relationship of an identified mediastinal mass to nearby intrathoracic vascular structures. MRI can help differentiate between a possible mediastinal mass and a vascular abnormality such as an aortic aneurysm. Video-assisted thoracoscopy (VATS) technique has been used successfully for biopsy of various mediastinal masses and is often used for the sampling of perihilar lymph nodes. VATS can also be used to resect benign posterior mediastinal tumors. Shorter hospital stay and faster return to work have been demonstrated with this technique. VATS is also one of the commonly used methods for the evaluation of mediastinal lymphoma. Posterior mediastinal tumors are typically approached through a thoracotomy, if not with VATS.

Treatment of choice for benign schwannomas is surgical resection. This includes the plexiform varieties and melanotic schwannoma. For malignant lesions, surgical resection and adjuvant radiation therapy is usually used to control residual disease, but the benefit of this is unknown. No known chemotherapeutic regimens are effective against these tumors. In cases of benign neoplasms, complete excision of the lesion itself is generally sufficient. Benign neurofibroma requires some resection in addition to the lesion itself, that being resection of the associated nerve. All benign neoplasms that are encapsulated should be resected without violation of the capsule. VATS resection is now commonplace for these benign tumors. When surgical resection of malignant neoplasms of the mediastinum is the primary treatment, bloc resection of the tumor should be performed whenever possible.

Mediastinal schwannoma is a lesion not so often encountered in clinical practice. Though it is usually benign, malignant transformations have been documented. A clinician should be alert to identify them at an early stage.

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