TUMORS OF THE MEDIASTINUM

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ARMED FORCES INSTITUTE OF PATHOLOGY

ATLAS OF TUMOR PATHOLOGY

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TUMORS OF THE MEDIASTINUM

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Hans George Schlumberger.

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INTRODUCTION

The mediastinum contains a variety of organs and structures that may be the sites of cysts and neoplasms (fig. 1). The most frequently encountered tumors in this region are thymomas and teratomas in the anterior mediastinum; lymphoblastomas in the anterior and middle mediastinum; and tumors of the intercostal nerves, sympathetic nerve trunks, and ganglia in the posterior mediastinum.

Benign and malignant lesions of the chest wall, particularly of bone and cartilage, may encroach upon the mediastinum, but seldom, if ever, are primary there. No chordoma of the thoracic vertebrae has been reported to involve the mediastinum. Fibrosarcomas and rhabdomyosarcomas that secondarily invade the mediastinum from their primary sites in the chest wall or diaphragm have been reported. Tumors of distant organs may first become manifest as metastases to the mediastinal lymph nodes.

Faulty embryogenesis may lead to the presence of aberrant tissues and to cyst formation. Adenomas of displaced thyroid or parathyroid usually occupy the superior or anterior mediastinum. Cysts may have their origin in the thymus, the pericardium, or the respiratory and gastrointestinal tracts. Cystic teratomas are found chiefly in the anterior mediastinum, pericardial cysts in the anterior and middle regions, bronchogenic cysts in the superior and middle divisions, and gastroenteric cysts in the posterior segment.

The relative incidence and location of tumors and cysts of the mediastinum are shown in Table I. The number of cases reported in the literature often does not accurately reflect the frequency with which a given tumor is found at operation or autopsy. Furthermore, the accuracy of the diagnosis in some instances is open to question. The problem of incidence will be considered again in the discussion of the individual tumor types.

The increasing accessibility of mediastinal structures to the surgeon as the result of improved surgical technique, better preoperative and postoperative care, and more adequate anesthesia has stimulated interest in the neoplasms of this region. In consequence of this and the improved methods of diagnosis, tumors frequently regarded as extremely rare are being reported with increasing frequency. Comprehensive reviews of the literature have been published.

The safety that attends thoracic surgical procedures in competent hands has outmoded the use of the therapeutic test dose of radiation as a means of

diagnosis. Every mediastinal tumor not known to be metastatic should be biopsied before it is irradiated. This not only permits an accurate diagnosis, but prevents the occurrence of tissue changes that often increase the difficulty of subsequent surgical removal.

The tumors of the thymus, esophagus, heart, and pericardium will not be discussed; they are fully considered in Fascicle 19, "Tumors of the Thymus," in Fascicle 20, "Tumors of the Esophagus," and in Fascicle 7, "Tumors of the Cardiovascular System."

Table 1

LOCATION OF TUMORS AND CYSTS IN THE MEDIASTINUM *

ANTERIOR MEDIASTINUM	SUPERIOR MEDIASTINUM	MIDDLE MEDIASTINUM	POSTERIOR MEDIASTINUM
Thymoma	Goiter	Bronchogenic cyst	Neurilemoma
Teratoma	Bronchogenic cyst	Lymphomas	Neurofibroma
Goiter	Parathyroid adenoma	Pericardial cyst	Ganglioneuroma
Parathyroid adenoma	Myxoma	Plasma cell myeloma	Sympathicoblastoma
Lymphomas	Lymphomas		Fibrosarcoma
Lipoma			Lymphomas
Fibroma			Goiter
Lymphangioma			Xanthofibroma
Hemangioma			Gastroenteric cyst
Chondroma			Chondroma
Thymic cyst			Myxoma
Rhabdomyosarcoma			Meningocele
			Paraganglioma

*Arranged in order of incidence and site of relative frequency.

Rhabdomyosarcoma +

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Figure 1. The mediastinum of a newborn male infant seen from the left side.

H—heart; L—root of left lung; N—sympathetic nerve trunk and intercostal nerves; P—pericardium; R—rib; T—thymus.

The most common sites of the primary tumors in this region are the thymus in the anterior mediastinum and the sympathetic nerve trunk and intercostal nerves in the posterior mediastinum. A. F. I. P. Neg. Acc. No. 218687-1.

TUMORS OF NERVOUS TISSUES *

Nearly all neurogenic tumors of the mediastinum arise from the paravertebral sympathetic nerve trunks and the intercostal nerves; therefore, they are usually found in the posterior mediastinum. Rarely, one of these tumors may have its origin in the neck and secondarily encroach upon the superior mediastinum.

The neurogenic tumors are the most frequently encountered neoplasms that are primary in the mediastinum, although they are approached in number by the thymomas and teratomas. The literature up to 1940 listed only about 135 of these tumors, but within the past decade 3 groups of observers have reported 18, 29, and 48 new cases. This would indicate that the older literature is not a trustworthy guide to the true incidence of neurogenic tumors in this region.

TUMORS OF NERVE FIBERS

Neurilemoma.

SYNONYMS AND RELATED TERMS: Neurilemoma (Lat.); angioneurofibroma; false neuroma (Virchow); fibroglioma; "fibromyxoma"; glioma; lemmoma; myoschwannoma; neurilemoblastoma; neurinoma; neurofibromyxoma; neurolemmoma; neuroma fibrillare; palisaded neurinoma; perineural fibroblastoma; perineural glioma; peripheral glioma; schwannoglioma; schwannoma; specific nerve sheath tumor.

This benign neoplasm is the most common neurogenic tumor in the mediastinum and, like others of the group, is characteristically found in the posterior mediastinum; however, it may occur elsewhere, particularly in the superior mediastinum. The origin of these tumors in the nerve sheath of Schwann is now generally accepted. In the mediastinum, the neurilemomas usually attain a larger size than they do in other parts of the body. This may be due to a lack of symptoms; the majority are discovered as an incidental finding on routine examination of the chest.

GROSS. Like other tumors of the mediastinum that are bordered by air-bearing lung, the neurilemomas are sharply defined by roentgenogram (figs. 2, 6, 7, 9). When exposed they are often spherical and well encapsulated (figs. 3, 12). The capsule may be loosely attached to the areolar tissue at the arch of the aorta (fig. 6) or firmly adherent to paravertebral tissues (fig. 7). Rarely, the tumors arise from nerve roots within the spinal canal, grow through

*See Fascicle 6, "Tumors of the Peripheral Nervous System," and Fascicle 29, "Tumors of the Adrenal Gland."

the intervertebral foramina, and continue to increase in size within the mediastinum. Because of their shape, such lesions are labelled "dumbbell" or "hourglass" tumors. These are terms descriptive of a tumor's shape and are not specific for any type of tumor. On section the neurilemomas are mottled gray, yellow, and red, and they may contain several small or large cystic spaces (fig. 12). Occasionally, the cut surface shows definite fasciculation with few, if any, cysts (pl. I–B).

MICROSCOPIC. Two distinct patterns have been identified, both of which are usually found in the same tumor (fig. 5). The first, classified by Antoni as type A, is composed of elongated spindle-shaped cells of Schwann. These are arranged in interlacing bundles with the nuclei frequently aligned in parallel rows (fig. 4). Occasionally, several sections must be examined to discover this nuclear pattern, which may be obscured by extensive degeneration and hyalinization. The regimented nuclei are often grouped in a manner suggesting bizarre tactile corpuscles and are known as Verocay bodies (fig. 8).

The second tissue pattern is identified as Antoni type B. Herein the cells of Schwann are stellate rather than spindle-shaped and form a loose network in which small areas of degeneration (microcysts) are often encountered (fig. 10). After in vitro growth, however, the cells assume the elongated form characteristic of the sheath cells (fig. 11).

Neurofibroma.

SYNONYMS AND RELATED TERMS: Neurofibroma (Lat.); elephantiasis neuromatosa; fibroma molluscum; multiple neurofibromatosis; multiple neuroma; neurinomatosis; neuroblastomatosis; neurofibromatosis; neuromatosis; plexiform neuroma; sheath neuroma; solitary neurofibroma; von Recklinghausen's disease.

The most frequent diagnosis made by pathologists on benign tumors of the posterior mediastinum is that of neurofibroma. In a series of 20 benign neurogenic tumors of this region received at the Armed Forces Institute of Pathology, the submitting diagnosis in 18 was neurofibroma. Examination of the sections revealed that most of these tumors were neurilemomas. This reflects not so much a mistake in diagnosis as the all-inclusive nature of the term "neurofibroma" when employed by the general pathologist.

Unlike the neurilemoma, which represents a neoplastic overgrowth of the sheath cells alone, the neurofibroma is characterized by the presence of all elements of a nerve trunk—the sheath cells, axons, and connective tissue from the endoneurium and perineurium. Although the sheath cells are usually the dominant cell type, they lack the orderly arrangement displayed in the neurilemoma. These tumors are uncommon in the mediastinum, despite their sometimes reputed frequency there. A few cases, particularly those in which there is evidence of the generalized neurofibromatosis of von Recklinghausen, may be accepted. Recently Ackerman and Taylor found 7 cases of mediastinal neurofibroma in a series of 48 primary intrathoracic neurogenous tumors. Each of the 7 patients is free of disease following excision of the tumor.

NEURILEMOMA

Figure 2.* Roentgenogram of a sharply defined mass in the left superior mediastinum that had increased about 1 cm. in diameter during the previous four years. Woman 30 years old. A. F. I. P. Neg. Acc. No. 138927–8.

Figure 3.* Gross of the well encapsulated tumor seen in the preceding roentgenogram. The tumor was readily shelled out from its bed in the superior sulcus beneath the mediastinal pleura. The cut surface reveals a firm cortex 2 to 5 mm. in width and a softer subcortical region, the seat of degeneration and hemorrhage. Small cysts are visible near the center of the mass. A. F. I. P. Neg. Acc. No. 138927–1.

Figure 4. Photomicrograph of neurilemoma, showing palisading of the nuclei—Antoni type A tissue. × 150. A. F. I. P. Neg. Acc. No. 218687–72.

*Courtesy of Blades, B. Mediastinal tumors. Report of cases treated at Army Thoracic Surgery Centers in the United States. Ann. Surg., 123: 749–765, 1946.

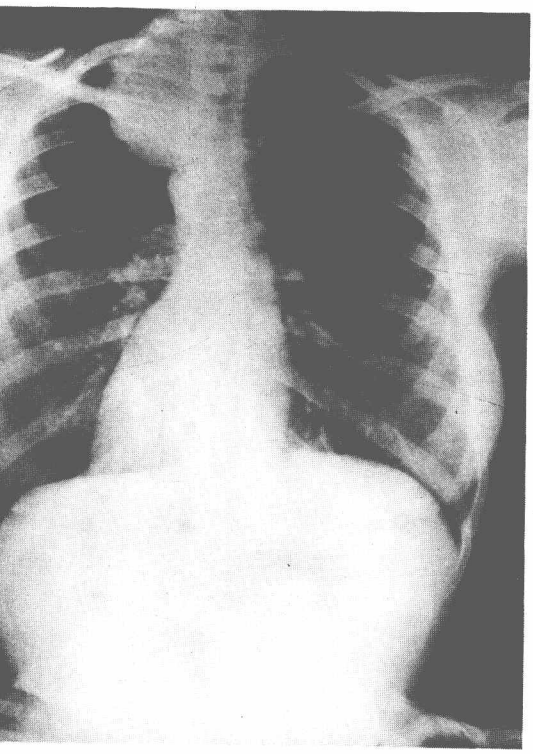

Fig. 2

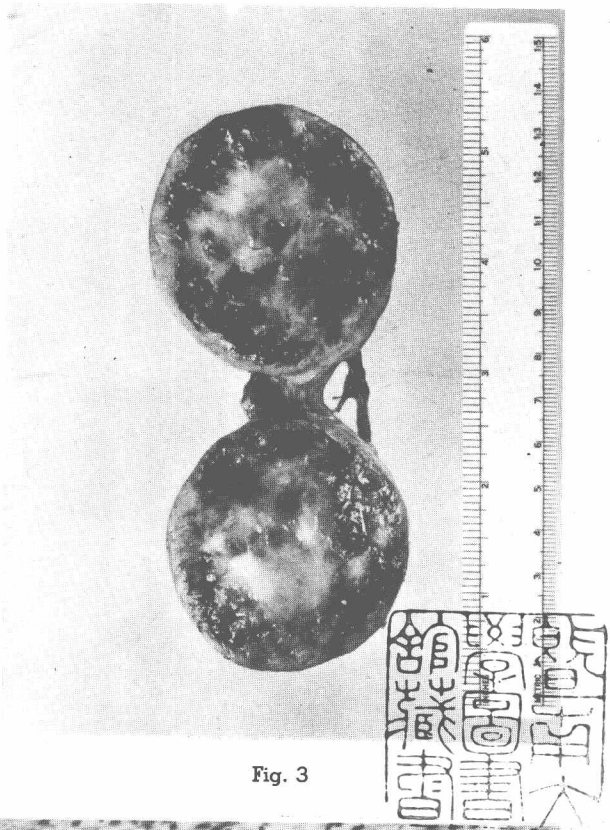

Fig. 3

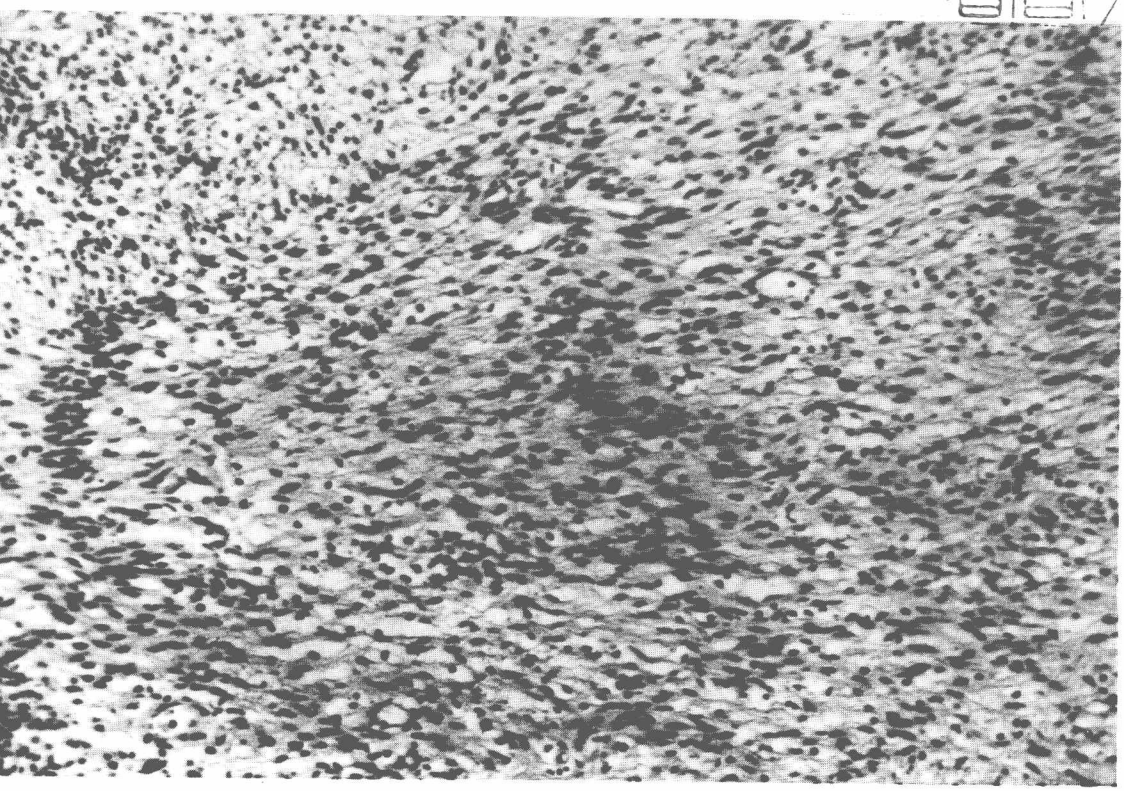

Fig. 4