

## Surgical Treatment of Neurogenic Tumors of the Chest

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**Background:** Neurogenic tumors are commonly found in the mediastinum, especially in the posterior mediastinum or in the chest wall, and have a variety of clinical and histological features. We reviewed our experience with these types of tumors and assessed diagnostic and therapeutic approaches.

**Patients and Methods:** A series of 60 consecutive patients with a neurogenic tumor of the chest, all seen at a single institute, was retrospectively reviewed.

**Results:** The mean age of the 60 patients was 40 years, including 32 males and 28 females. Preoperative symptoms were present in seven patients (11.7%). Median tumor size was 5.3 cm, ranging from 1.4 to 20 cm. The major location of the tumor was the posterior mediastinum in 38 cases (63.3%) and the chest wall in 16 cases (26.7%). The operative procedure performed was tumor extirpation in 58 cases (96.7%), in which video-thoracoscopic procedure was performed in nine patients and chest wall resection in two cases. The major histological type was neurilemmoma in 51 patients (85%), and malignant tumor was only diagnosed in one patient with malignant schwannoma. Tumor related death occurred in only one case with malignant schwannoma. Ten patients with neurilemmoma were precisely diagnosed by magnetic resonance imaging.

**Conclusion:** Almost all cases of intrathoracic neurogenic tumors were benign in nature. Therefore, surgical indications may be carefully determined in cases with no symptom and with imaging that indicate benign neurilemmoma. (*Ann Thorac Cardiovasc Surg* 2004; 10: 148–51)

**Key words:** neurilemmoma, magnetic resonance imaging, video-assisted thoracic surgery (VATS)

### Introduction

Thoracic neurogenic tumors usually originate from the neural crest during the development of peripheral nerve systems such as those of the autonomic or paraganglionic nerves. Nerves and ganglia of both somatic and autonomic nervous systems are found throughout the thorax but are concentrated in the paravertebral sulcus region, commonly described as the posterior mediastinum.

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Most of the peripheral nerves in the thorax are situated in the posterior mediastinum, and 12% to 21% of all mediastinal tumors are neurogenic.<sup>1,2)</sup> Such tumors, often asymptomatic, are occasionally found on chest roentgenograph or chest computed tomography (CT).

At present, due to the uncertainty of preoperative diagnosis, the increasing size of the tumor, and the possibility of malignancy, early surgical excision is considered to be the most acceptable strategy.

We herein report on our 30-year experience with neurogenic tumor of the thorax.

### Patients and Methods

From August 1974 to August 2002, 60 consecutive patients with neurogenic tumor of the thorax were surgi-

cally treated in our department. The patient group included 32 males and 28 females, with a mean age of 40 years (range, 5 to 72 years). The majority of these patients were referred to us because of clinical symptoms or radiographical abnormalities on chest radiograph or chest CT. The mean follow-up period was 83 months (range, 3 to 294 months).

We retrospectively reviewed the medical records of these patients and investigated the indications of surgical resection for neurogenic tumor of thorax based on tumor location.

## Results

Preoperative symptoms were observed in seven cases (11.7%), as follows: back pain in three, chest pain in one, dyspnea in one, dysphasia in one, and hemoptysis in one patient. The preoperative CT showed the tumor as a solid lesion in 58 cases (96.7%) and as a cystic lesion in two cases (3.3%). The tumor was located on the right side in 38 (63.3%) patients and on the left side in 22 (36.7%) cases. The site of tumor location was the posterior mediastinum in 38 cases (63.3%), the middle mediastinum in five cases (8.3%), the anterior mediastinum in one case (1.7%), and the chest wall in 16 cases (26.7%) (Table 1). An anterior mediastinal tumor originated from the sympathetic nerve, five middle mediastinal tumors from the vagus nerve in three cases, the sympathetic nerve in one case, undetermined in one case, 38 posterior mediastinal tumors from intercostal nerves in 14 cases, the vagus nerve in one case, the sympathetic nerve in 10 cases and undetermined in 13 cases and 16 chest wall tumors were all from intercostal nerves, respectively.

The operative procedure was tumor extirpation in 58 cases and chest wall resection in two cases. A case with malignant schwannoma required combined resection of the esophagus. Among these cases, nine (47.4% of cases from June 1992 to August 2002) were treated under video-assisted thoracic surgery (VATS) and 51 were under conventional thoracotomy including 32 posterolateral thoracotomies, 17 axillary thoracotomies and one median sternotomy. Of cases in which the patient underwent VATS, tumors were located in the posterior mediastinum in five and on the chest wall in four. The median duration of operation was 136 minutes (range, 50 to 350 minutes) for conventional thoracotomy and 134 minutes (range, 40 to 245 minutes) for VATS. The median amount of bleeding was 312 g (range, 0 to 1,200 g) for conventional thoracotomy and 91 g (range, 0 to 410 g) for VATS. In

**Table 1. Profile of patients and diseases**

|                       |       |                 |
|-----------------------|-------|-----------------|
| Number                | 60    |                 |
| Age (years)           | 40    | (range, 5-72)   |
| Male/female           | 32/28 |                 |
| Symptoms              |       |                 |
| Present/absent        |       | 7/53            |
| Right/left            | 38/22 |                 |
| Location              |       |                 |
| Anterior mediastinum  | 1     | (1.7)           |
| Middle mediastinum    | 5     | (8.3)           |
| Posterior mediastinum | 38    | (63.3)          |
| Chest wall            | 16    | (26.7)          |
| Size (cm)             | 5.3   | (range, 1.4-20) |
| Operative procedure   |       |                 |
| Extirpation           | 58    |                 |
| Thoracotomy           | 49    |                 |
| VATS                  | 9     |                 |
| Chest wall resection  | 2     |                 |
| Histology             |       |                 |
| Neurilemmoma          | 51    | (85.0)          |
| Neuroganglioma        | 6     | (10.0)          |
| Neurofibroma          | 2     | (3.3)           |
| Malignant schwannoma  | 1     | (1.7)           |
| Origin                |       |                 |
| Sympathetic           | 21    | (35.0)          |
| Vagus                 | 4     | (6.7)           |
| Intercostal           | 30    | (50.0)          |
| Undetermined          | 5     | (8.3)           |

Percent in parenthesis

VATS: video-assisted thoracoscopic surgery

none of our cases did the tumor extend into the spinal canal (a so-called "dumbbell" tumor).

The histological type of the tumors was neurilemmoma in 51 cases (85.0%), neuroganglioma in six cases (10.0%), neurofibroma in two cases (3.3%), and malignant schwannoma in one case (1.7%). In one case, a patient with neurilemmoma had von Recklinghausen's disease. The tumor originated from the sympathetic nerve in 21 cases (35.0%), from the intercostal nerve in 30 cases (50.0%), from the vagus nerve in four cases (6.7%), and the origin could not be determined in five cases (8.3%).

The only tumor-related death occurred at 9.8 months after surgery in the case of the patient with malignant schwannoma. There were five deaths related to other diseases.

For the preoperative examination, 11 of 60 cases underwent magnetic resonance imaging (MRI), including dynamic curve examination in three cases (Table 2). Neurilemmomas showed a low intensity to isointensity pattern on T1-weighted imaging and a high intensity pattern on T2-weighted imaging, whereas the ganglioneurofibroma showed a low intensity pattern on

**Table 2. Result of magnetic resonance imaging of the neurogenic tumors**

| Case | T1  | T2   | Enhancement | DS  | Histology           | Origin      |
|------|-----|------|-------------|-----|---------------------|-------------|
| 1    | Low | High | (+)         | ND  | Neurilemmoma        | Intercostal |
| 2    | Low | High | (+)         | ND  | Neurofibroma        | Intercostal |
| 3    | Iso | High | (+)         | ND  | Neurilemmoma        | Sympathetic |
| 4    | Low | High | (+)         | ND  | Neurilemmoma        | Sympathetic |
| 5    | Low | High | (+)         | ND  | Neurilemmoma        | Intercostal |
| 6    | Low | High | (+)         | ND  | Neurilemmoma        | Intercostal |
| 7    | Low | High | (+)         | ND  | Neurilemmoma        | Intercostal |
| 8    | Low | High | (+)         | GIP | Neurilemmoma        | Sympathetic |
| 9    | Iso | High | (+)         | GIP | Neurilemmoma        | Sympathetic |
| 10   | Low | High | (+)         | GIP | Neurilemmoma        | Intercostal |
| 11   | Low | Iso  | (+)         | NC  | Ganglioneurofibroma | Intercostal |

DS: dynamic scan, Low: low intensity, Iso: isointensity, High: high intensity, ND: not done, GIP: gradual increase pattern, NC: no change.

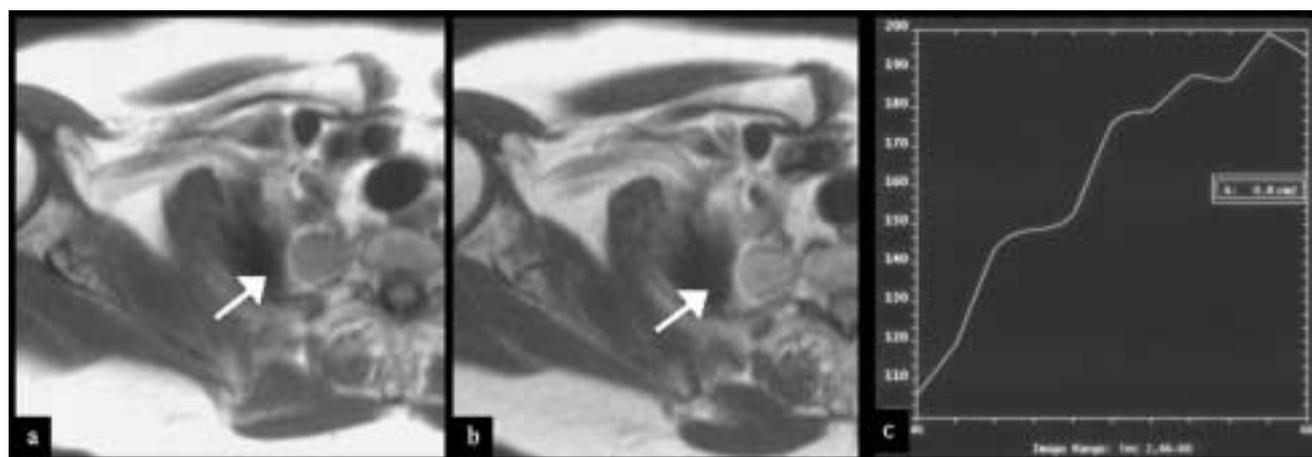
T1-weighted imaging and isointensity on T2-weighted imaging. Dynamic curve examination using gadolinium-diethylene triamine pentaacetic acid (Gd-DTPA) showed a pattern of gradual increase (Fig. 1).

**Comment**

Intrathoracic neurogenic tumors were reported to represent 75% to 95% of tumors in the posterior mediastinum.<sup>3,4)</sup> Histologically, a benign tumor arising from Schwann's cell is called a neurilemmoma, and that arising from the nerve sheath is called a neurofibroma. Ganglioneuromas arise from nerve cells, most frequently in the autonomic nervous system. The majority of posterior neurogenic tumors are asymptomatic. However, as

they become larger in size, they can produce symptoms related to local compression, bone erosion, and spinal cord involvement. Moreover, both benign and malignant neurogenic tumors are sometimes asymptomatic.

Surgical excision is applicable if there is no interval increase in size, an absence of symptoms, or the location indicates very low risk of a dumbbell tumor. However, the problem is how to make an accurate preoperative diagnosis of malignant tumor. The frequency of malignant tumor among intrathoracic neurogenic tumors is relatively low; it is reported to be from 4% to 13%.<sup>5,6)</sup> Actually, only one case (1.7%) among those of neurogenic tumors of the thorax in our series was malignant (malignant schwannoma). On the other hand, the most common histological type was benign neurilemmoma (84%), which



**Fig. 1.** a: T1-weighted imaging of the neurilemmoma. The tumor showed low signal intensity. b: T2-weighted imaging of the neurilemmoma showed high signal intensity. c: The results of dynamic scan of the neurilemmoma by Gd-DTPA showed a pattern of gradual increase.

can be diagnosed by MRI. Therefore, this imaging modality should be useful for distinguishing malignancy in a neurogenic tumor. The chest wall was the second-most frequent location of a neurogenic tumor (26.7%) in the patients in our series, and the chest wall neurogenic tumors were all benign neurilemmoma. MRI findings have been reported to enable histological comparison of mediastinal neurogenic tumors. Sakai et al.<sup>7,8)</sup> reported that typical MRI findings of neurilemmoma include an inhomogeneous low to intermediate signal intensity on T1-weighted images and high signal intensity on T2-weighted images, and that those of neuroanglioma include homogeneous intermediate signal intensity on T1-weighted images and inhomogeneous high T2-weighted images. Neurofibroma, which has an invasive nature and is more aggressive in nature than neurilemmoma, shows a low to intermediate signal intensity on T1-weighted images and high signal intensity with a peripheral region of the tumor on T2-weighted images. Recently, dynamic curve study on MRI appears to offer higher efficacy for accurate diagnosis of neurogenic tumor of the thorax.

In our opinion, the best approach would be the standard posterior thoracotomy, located one or two intercostal spaces above or below the tumor in order to avoid directly injuring it. However, standard thoracotomy often requires a large incision that impairs the respiratory muscles and/or causes postoperative pain. Moreover, this approach sometimes provides a less effective operative field. VATS minimizes the access trauma and often provides an appropriate view, especially for the posterior mediastinum.<sup>9-11)</sup> We introduced VATS for neurogenic tumor of the thorax at our facility in June 1992. There were no major complications in patients undergoing this procedure, and even tumors as large as 7 cm in diameter could be excised. Postoperative hospital stay was shorter for cases undergoing VATS compared to those undergoing standard thoracotomy.

In conclusion, the VATS seems feasible as the approach

for the thoracic neurogenic tumor since it is less invasive and provides an appropriate view for the operation. The surgical indications may be carefully determined in cases with no symptoms and with imaging that indicate benign neurilemmoma due to the benign nature of the tumor. MRI would facilitate therapeutic determination by distinguishing the most common histological type of neurogenic tumor (neurilemmoma).

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