Benign Neurogenic Tumors and Bronchogenic Cysts

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Introduction

Neurogenic tumors and bronchogenic cysts belong to the category of mediastinal compartment growths that leave little room for doubt regarding their diagnosis because of their position, form, and structure.

Neurogenic tumors are the most frequent growths of the posterior mediastinum, accounting for more than 30 % of all mediastinal tumors if infants and children are taken into consideration. Among adults, fewer than 5 % of neurogenic tumors of the posterior mediastinal cavity are malignant, compared with 60 % in children under 16 years of age. The tumors originate from either the sympathetic trunk or the intercostal nerves. Depending on their origin, a differentiation is made between nerve sheath tumors and tumors of the autonomic ganglia, paraganglia, and peripheral neuroectoderm. Neurilemmoma and neurofibroma belong to the benign nerve sheath tumors, which account for up to 60 % of all neurogenic tumors, and may be observed typically in adults in the third to fifth decades of life. These tumors are energy elastic and well-defined. Approximately 10 % develop a so-called hourglass (or dumbbell) formation, which means they possess a link to the intraspinal parts via a narrow channel through the intervertebral foramen (Deslauriers et al. 2005).

Benign tumors of ganglionic origin are called ganglioneuromas. These tumors represent approximately one third of all neurogenic mediastinal tumors, and their clinical as well as radiologic appearance is similar to that of nerve sheath tumors. It is mandatory to exclude an intraspinal extension of the tumor. However, a widening of the intervertebral foramen or the adjacent ribs does not provide evidence of a dumbbell constellation. If there is evidence of such a formation on CT, an additional MRI scan provides the best imaging results with respect to preparing for the operation. In this case, a neurosurgical team should remove the intraspinal parts by performing a laminectomy before addressing the intrathoracic growth in the same or a second operation.

Mediastinal cysts account for approximately 25 % of all primary growths of the mediastinal compartment; however, about 90 % of these are bronchogenic cysts. Despite their (partly) significant size, these cysts are asymptomatic in adults most of the time, and thus are incidental findings. More often, compression symptoms lead to their discovery in children.

Bronchogenic cysts develop mostly along the tracheobronchial tree, most frequently posterior to the main carina. They are mainly unilocular and lined with a ciliated cylindric epithelium, which is responsible for the viscous content ranging in color from gray-white to brown. In form and structure, the cysts are sharply defined round to oval masses, which may vary in shape on dynamic imaging with changes of configuration in breathing, differentiating them from solid tumors. As there is sometimes a direct connection to the bronchial system, the contents of the cyst may become infected, leading to an increase in size. Also, malignant degeneration of the cyst wall has been described (Nichols et al. 2008). These observations, as well as the fact that more than 60 % of all bronchogenic cysts become symptomatic over time, underscore the absolute indication to operate.

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Figures 33.1 and 33.2

If an intraspinal extension of the neurogenic tumor can be safely excluded, it may be assumed that the tumor can be removed completely by thoracoscopy. An exception is the occasional tumor that has broad contact with the pleural apex, requiring an additional axillary incision via the second intercostal space under certain circumstances. The lesion is always covered by the parietal pleura; it rests mostly with a wide base on the intervertebral foramina, thus it is not very mobile. If the surgeon prefers to stand with a ventral view, the patient is placed in the lateral decubitus position and tilted slightly posteriorly. Three to four incisions are made in the midaxillary line for placement of the video-optic, manipulation forceps, and lung retractor. The parietal pleura is cut in circular fashion around the base of the tumor; accurate and thorough hemostasis is of utmost importance to prevent hematoma of the pleura, which would considerably complicate the overall view of the operative field. The blood vessels accompanying the intercostal nerves are clipped anterior and posterior to the tumor. The sympathetic chain proximal and distal to the neurogenic tumor also must be clipped. Then the tumor is mobilized at its base, step by step, by blunt dissection while being manipulated with only a swab; to prevent injury to the capsule, the tumor must not be treated roughly. After complete excision, the tumor is placed into an endosurgical extraction bag and removed from the pleural cavity

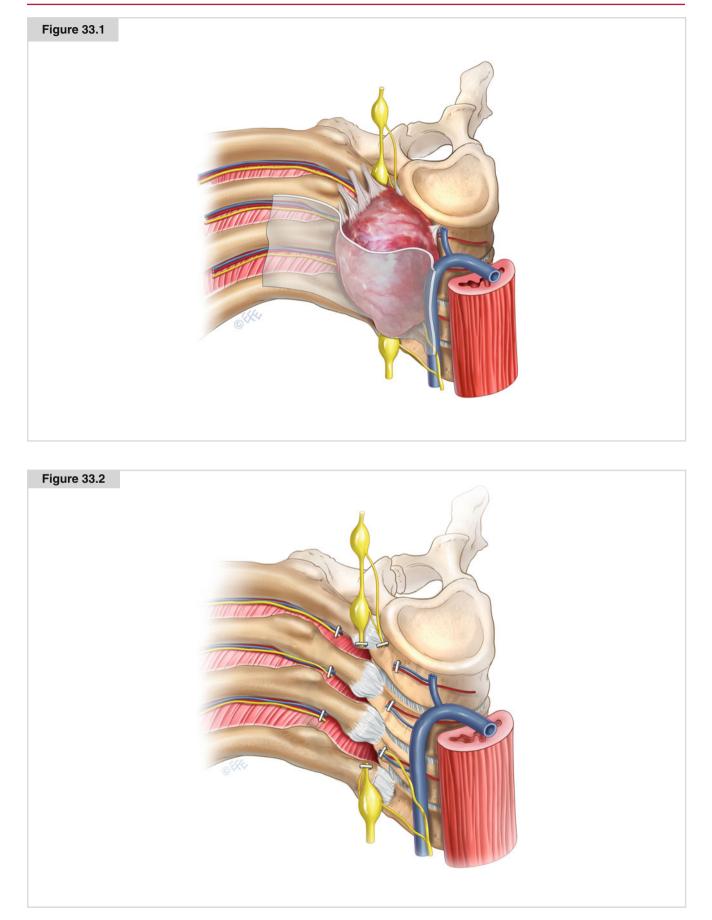
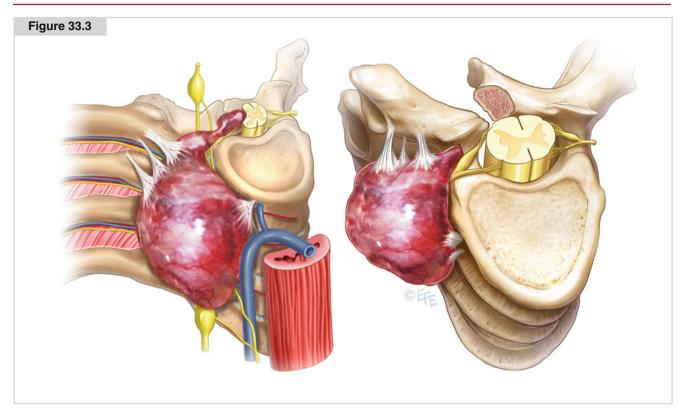


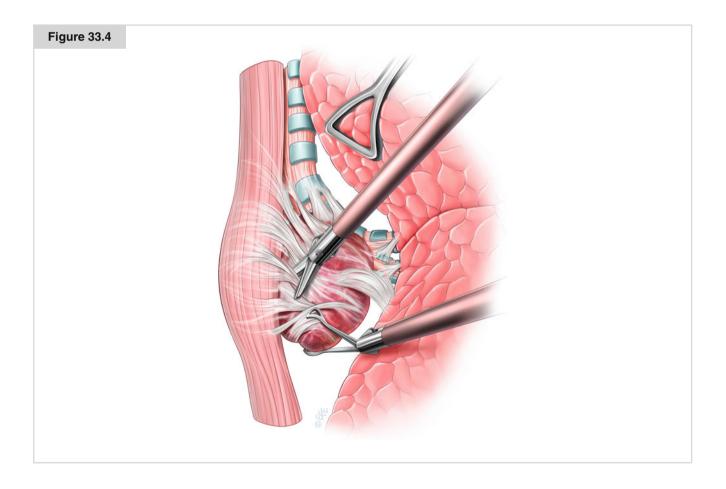
Figure 33.3

Neurogenic tumors may widen the intervertebral foramen without spreading into the canal. This constellation also permits a complete tumor extension alone from the thoracic cavity; however, tearing of the nerve root may have disastrous consequences, namely direct injury to the spinal cord, laceration of the attendant intercostal artery, or dura injury resulting in a loss of cerebrospinal fluid. Placement of a clip before the spinal root is severed prevents the dura from opening. With regard to dumbbell tumors, various strategies have been implemented successfully, including a neurosurgical/thoracic procedure in one or two operations as well as in toto tumor removal by means of a posterolateral thoracotomy (Bousamra et al. 2008). Because it can be determined intraoperatively whether a tumor is completely extradural and can be removed without opening the dura, or whether an intradural extension is exhibited, neurosurgical expertise should always be readily available

Figure 33.4

Bronchogenic cysts, most of which are connected to the tracheal bifurcation and protrude to the right, are exposed via a limited lateral thoracotomy or video-thoracoscopy. It is recommended that the operation be executed dorsally with the patient in a left lateral position and tilted to the left. The treatment objective should be complete enucleation of the cyst, as aspiration of only the cyst contents or an incomplete resection leaving the cylindric epithelium intact favors a relapse. In most cases, the cyst adheres to the attendant structures, especially the bifurcation and esophagus. This circumstance should not prevent complete removal; however, injury to neighbouring organs must be prevented. If, for this reason, a decision is made to leave parts of the wall, at minimum the epithelial tissues lining the cyst should be removed and the interior aspect of the residual cyst devitalized through electrocoagulation. As long as they are not under pressure, small cysts may be pulled with a blunt alligator forceps to release their adherence to the attendant structures and removed stepwise with endoscissors. Sometimes, preparation is easier when the contents of the cyst have been aspired beforehand (Naunheim 2008). Larger cysts should be emptied preliminarily to permit a better view of the attendant structures. Insofar as the cyst is opened, a bacteriologic smear is recommended. If there is doubt as to the integrity of the tracheobronchial tree or the esophagus, this must be clarified intraoperatively with positive pressure ventilation or methylene blue administration via a gastric tube





Conclusion

The thoracoscopic method has become standard procedure for solitary neurogenic tumors without intraspinal extension because of its small degree of invasiveness, although this procedure is more demanding than standard thoracotomy (Han et al. 2002). Absolutely essential at the end of the operation, aside from checking for hemorrhage, is the exclusion of cerebrospinal fluid leakage. Postoperative loss of cerebrospinal fluid requires either local revision or, alternatively, a hemilaminectomy at the corresponding level. A careful preand postoperative survey of the patient's neurologic status, focusing on the cervicothoracic ganglion, recurrent laryngeal nerve, phrenic nerve, and brachial plexus is indispensable, especially for forensic reasons.

The asymptomatic patient is always at risk if there is an infection or perforation of a bronchogenic cyst; here, the indication for extirpation (as complete as possible) is given. As a rule, securing an absolutely certain diagnosis is possible preoperatively with CT or MRI. A relapse might develop within months if vital cylindric epithelium is not removed.

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