Thoracoscopic Transclavicular Approach for a Large Thoracic Inlet Tumor
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Large lesions of the thoracic inlet represent a significant challenge to surgeons due to their close relationship with important adjacent neurovascular structures. We report a case of neurofibromatosis type 1 presented with a large neurofibroma located in the right thoracic inlet, and a malignant peripheral nerve sheath tumor in the right clavicle, and treated with a hybrid thoracoscopic anterior transclavicular approach to remove the tumors. Combination of thoracoscopic and minimal chest wall resection may serve as a feasible option for resection of the large and benign thoracic inlet tumor in a way of minimal invasiveness and safety.


Neurofibromatosis type 1 (NF1) is an autosomal dominant disorder characterized by noncancerous tumor growth known as neurofibromas. NF1 is caused by dominant loss-of-function mutations of the tumor-suppressor gene neurofibromin 1 located on chromosome 17q11.2, and commonly manifests in the form of cafe-au-lait spots, neurofibromas, Lisch nodules, and axillary or inguinal freckling [1].

We present the case of a 27-year-old male patient who complained of right shoulder pain for 2 weeks. Digital radiology showed a right thoracic inlet mass (Fig 1A). Computed tomography and magnetic resonance imaging scans revealed a 2.3 × 2.0 cm clavicular tumor near the right sternoclavicular, and another 6.4 × 4.7 cm tumor occupying the right thoracic inlet and extending to the root of the neck. The tumor located at the right clavicle was in irregular shape with high signal intensity on T2-weighted image, and enhanced obviously after contrast injection. The tumor occupying the right thoracic inlet showed in soft-tissue-density, round shape, and smooth margin on T2-weighted image. The patient reported a history of neurofibromas resection in the maxillofacial region 1 year previously. Multiple cutaneous neurofibromas and cafe-au-lait macules were found during physical examination (Fig 1C), and 3 Lisch nodules observed on the iris during slit-lamp examination. His family history was notable for NF1 on his maternal side of the family. He was diagnosed with NF1 and referred to curative surgery.

The patient was positioned supine with the neck fully extended, followed by an L-shaped anterior transclavicular approach with supraclavicular extension (Fig 1C). The sternoclidomastoid was dissected away from the clavicle and the medial third portion of the clavicle was divided to remove the clavicular tumor. The subclavian vessels were then dissected free from the 1st first rib. The tumor was tightly attached to the pleural side of the first and extended to the root of the neck. The medial 3 cm of the 1st first rib was sectioned from the sternocostal joint to facilitate the exposure of the tumor’s anterior bound. Next, a 30 degree thoracoscope was introduced in the sixth intercostal space, anterior axillary line to check lower bound of the tumor. The tumor occupied the right thoracic inlet and the right brachiocephalic vein was dislocated anteriorly. Another 2-cm utility incision was made in the fifth intercostal space along the middle axillary line. Progressing from the medial side of the tumor, major vessels, such as the common carotid artery, jugular vein, and subclavian vessels, were freed from the tumor’s medial and anterior aspects through the anterior working window (Fig 1D). After dividing the tumor pseudocapsule, anterior incision, and electronic hook dissection from inside of the chest allowed complete resection of the tumor and delivery out from the anterior working window (Fig 1E). The postoperative recovery was uneventful. Nerve evaluation after surgery revealed short-term decreased sweating of the face ipsilaterally, but ptosis and miosis did not occur. No signs of recurrent laryngeal nerve injury or phrenic nerve palsy were recorded after surgery (Fig 1B), and using the numeric rating scale the average postoperative pain score was 1.5. Pathology demonstrated that the thoracic inlet tumor was a neurofibroma, while the clavicular lesion was a malignant peripheral nerve sheath tumors (MPNST) with negative surgical margins (Fig 1F). The immunohistochemical examination for MPNST showed intense Vim expression with negative expression of S-100, desmin, and CD34. The patient was referred to adjuvant sequential chemoradiotherapy. After a 3-month follow-up, the patients did not report the signs of nerve compression, shoulder dysfunction, or tumor recurrence.

Comment
Patients with NF1 carry a higher risk of developing soft-tissue sarcomas, with a strong link demonstrated between NF1 and MPNST resulting in an estimated lifetime risk of 8% to 13% [2]. The mechanism of malignant transformation is still unknown, but evidence suggests that patients with microdeletions incorporating the entire NF1 gene are more susceptible compared with small truncating mutations [3]. Furthermore, the aggressive clinical features of MPNST have also been observed, including lowered response...
rate to chemotherapy and worse prognosis [4]. The best treatment modality is still complete surgical resection, with margin status and size being the most important predictors of disease-specific survival [5].

The thoracic inlet is an incapsacous area where complex bony structures protect major vessels and nerves underneath. Because large tumors in this region can be challenging, using an anterior route such as a transsternal [6] or transclavicular approach [7] will provide excellent exposure for resection of benign tumors in the thoracic inlet. However, partial sternotomy requires the transsternal approach, which may cause severe postoperative pain and increased risk of infection. Classical transclavicular approach also requires division of several ribs to facilitate dissection [7]. With the development of video-assisted techniques, curative tumor resection and minimally invasive surgery can be achieved simultaneously; in this case by combination of thoracoscopic surgery and minimal chest wall resection without splitting the manubrium.

Because the medial part of the clavicle was involved, a transclavicular approach may be the first choice from an anterior access. A partial resection of the first rib may then enlarge the working window anteriorly without leaving obvious functional implication. Furthermore, the intrathoracic portion of the lesion can be internally dissected safely using thoracoscopic vision while assisted by blunt dissection from the subclavicular space outside. Thus, extended removal of the chest wall may not be necessary. Meanwhile, the vagus nerve and the phrenic nerve can be monitored by direct view within the chest. Mild and temporary postoperative symptoms of sympathetic nerve impairment suggest that this hybrid approach may also minimize the risk of nerve injury and avoid tear to the medial brachiocephalic vessels. The average pain score and early ambulation after surgery demonstrate the advantage of this approach for young patients.

In conclusion, our experience suggests that the hybrid thoracoscopic anterior transclavicular approach is feasible for large and benign thoracic inlet tumors and promotes advantages of minimal invasiveness and safety.

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References


