Pneumonectomy and Contralateral Metastasectomy Through a Single Thoracotomy in a 9-Year-Old Girl With a Giant Tumor

Fotios A. Mitropoulos, MD, PhD, Meletios A. Kanakis, MD, PhD, Andrew C. Chatzis, MD, PhD, Loukas Kaklamanis, MD, PhD, and Achilleas G. Lioulias, MD, PhD

Departments of Pediatric and Congenital Heart Surgery and Pathology, Onassis Cardiac Surgery Center, and Department of Thoracic Surgery, Sismanoglio General Hospital of Athens, Athens, Greece

A 9-year-old girl with a giant tumor of the right lung and an isolated metastasis of the left lower lobe underwent combined pneumonectomy and metastasectomy through means of a right thoracotomy. Her postoperative course was uneventful. The operative approach of a tumor of this scale and the concurrent contralateral metastasectomy are described and discussed.


Malignant peripheral nerve sheath tumor (MPNST) is a rare, yet one of the most common, non-rhabdomyosarcoma soft tissue sarcomas in the pediatric population. These tumors occur most frequently at axial sites, are characterized by local aggressiveness, and have a tendency to metastasize early [1]. Treatment of MPNST represents a major challenge as there is no reliable modality other than radical surgery. Chemotherapy and radiotherapy are occasionally used in selected patients with unresectable tumors or metastatic disease [2, 3].

A 9-year-old girl underwent surgical excision of an MPNST of the cervical spine at the age of 7 years followed by adjunctive chemotherapy and was free of residual disease thereafter. Six months ago a lesion appeared in the right lung, and biopsy revealed metastasis of the primary tumor. She was advised to receive chemotherapy; however, this approach proved ineffective with the tumor increasing rapidly in size. In addition, a metastasis in the lower lobe of the left lung was detected (Fig 1). She was then referred to our unit for possible surgical treatment.

Address correspondence to Dr Kanakis, Syngrou Ave 356, Kallithea, Greece 17674; e-mail: meletis_kanakis@yahoo.gr.
Fig 2. (A) Histologic specimen showing the type of tumor. The tumor consists of fascicles of spindle neoplastic cells, positive for S-100 and neurofilaments (hematoxylin and eosin stain, original magnification \( \times 200 \)). (B) Intraoperative photo showing the excised left lung metastasis.

On admission the patient presented with a moderate rise in temperature (38°C), tachypnea (30 breaths/min) and orthopnea. Arterial oxygen saturation in room air was 84%. The patient underwent a right posterolateral thoracotomy. The thoracic cavity was entered through the fifth intercostal space. The sixth rib was infiltrated by the tumor and was therefore resected. The tumor was invading the right pericardium, including the right phrenic nerve. This portion of the pericardium was resected, and the macroscopically healthy rim of the remainder was suspended anteriorly at the edges of the incision to facilitate exposure. The tumor was totally in contact with and invading the right pulmonary artery, the right superior pulmonary vein, the superior vena cava, and the right hemidiaphragm. The right lung was removed, along with segments of the superior vena cava and the right hemidiaphragm. The right parietal pleura was subtotally resected. The right hemidiaphragm defect (2 × 2 cm²) was repaired by plication. A fenestrated bovine pericardium was used to close the created pericardial defect. The superior vena cava was repaired primarily by direct suturing.

A 5-cm vertical incision on the left pleura behind the posterior pericardium and in front and parallel to the esophagus at the level of the intrapericardial segment of the inferior vena cava was performed, and an entrance into the left thoracic cavity was achieved (Fig 1). The left inferior pulmonary ligament was mobilized, and the left lower lobe came into vision and was delivered through the opening. The metastasis was identified and excised. Further, the created opening allowed thorough palpation of the entire contralateral lung. Meticulous hemostasis was performed, and the left pleura was closed by direct suturing after imposing positive-pressure ventilation in the left lung. During this procedure there were no significant hemodynamic disturbances or arrhythmias. Finally, the thoracic duct was ligated to prevent potential postoperative chylothorax, and the patient was transferred to the intensive care unit in stable condition. Specimens were sent for histologic examination (Fig 2). The patient was discharged home on the sixth postoperative day in excellent clinical status without respiratory distress at rest and reasonable exertion, with an oxygen saturation of 100%. The pathology report confirmed total excision of the tumor and the contralateral metastasis, both identified as MPNST. She went on living a comfortable life for another 9 months, when abdominal dissemination of the tumor led to her demise on the 10th postoperative month.

Comment

We chose a standard right posterolateral thoracotomy for this procedure. The presence of a contralateral metastasis poses certain challenges in selecting the appropriate surgical approach. Alternative options could have been a simultaneous bilateral thoracotomy or staged surgery, with the left side approached by mini-thoracotomy or video-assisted thoracic surgery. A clamshell incision or a midline sternotomy have also been recommended.

Right posterolateral thoracotomy proved an excellent choice: through a single incision it allowed full access to the main tumor and an adequate approach to the left lung [4]. We were able to thoroughly examine the latter by palpation for possible other undetected lesions, and the known metastasis was easily approached, recognized, and removed. The single thoracotomy incision resulted in less distress in a patient with already poor respiratory reserves and therefore less postoperative morbidity.

A midline sternotomy could have provided access to both lungs; however, the right hemidiaphragm and the posterior mediastinum would not have been an easy reach. A clamshell incision provides excellent exposure of both lungs yet carries the worst record of postoperative morbidity. A video-assisted thoracic surgery approach in a second stage could not be safely performed in the context of prior contralateral pneumonectomy. In the presented case, the size of the tumor caused significant respiratory distress owing to massive vENOArterial shunting. Pneumonectomy was therefore mandatory to improve the respiratory status of the patient; nonetheless, the contralateral metastasectomy should also be performed with minimal respiratory encumbrment.

Distant metastases have been shown in 42% of patients with histologically confirmed MPNST [5, 6]. Neurofibromatosis type 1 is found in about half of patients with MPNST and occurs spontaneously in the other half [6]. Observations support the hypothesis that MPNST arising in patients with neurofibromatosis type 1 and
nonneurofibromatosis type 1 are not different per se, and data suggest that the choice of treatment should be independent of neurofibromatosis type 1 status [2]. Improved survival is expected after pulmonary metastasectomy for sarcoma. Tumor resectability, disease-free interval, and number of metastases seem to be important factors in determining patient selection for curative surgical intervention [7].

References


Extracellular Matrix Pleural Tent for Persistent Air Leak and Air Space in a Child After Upper Lobectomy

Patrick I. McConnell, MD

Department of Cardiothoracic Surgery, Nationwide Children’s Hospital, Columbus, Ohio

Creation of a pleural tent is effective in reducing persistent air leaks after pulmonary resection. I report a case of a pleural-like tent being created out of extracellular matrix to treat a persistent air leak in child after upper lobectomy for a large congenital pulmonary airway malformation type II. Over the next year, ipsilateral lung expansion and growth occurred with near complete resolution of the apical air space.


Dr McConnell discloses a financial relationship with CorMatrix Cardiovascular.