Resection of a primary mediastinal malignant rhabdoid tumor: A study in multidisciplinary collaboration and 3-dimensional printing

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3D-printed model of tumor (*yellow*), heart (*red/blue*), and trachea (*white*); axial view.

CENTRAL MESSAGE

With the innovative use of a 3D model, a team of pediatric general, congenital cardiac, and adult thoracic surgeons demonstrates the value of multidisciplinary care for a rare pediatric thoracic tumor.

Malignant rhabdoid tumors (MRT) are highly aggressive neoplasms characterized by mutations of the *SMARCB1* gene, most often arising in the kidney or central nervous system of children. extrarenal, noncranial MRTs (eMRTs) are more common in adults and rarely occur in the mediastinum.[1-3] There is no standard pediatric treatment; independent reports describe multimodal therapy of surgical resection, chemotherapy, and radiation.[3,4] eMRTs' aggressive nature often limits surgical management to noncurative, debulking procedures with 5-year overall survival between 15% and 36%.[3]

Tumors in anatomically challenging locations like the mediastinum often benefit from techniques to improve tumor visualization, such as 3-dimensional (3D) printing. 3D printing has been previously described to create surgical guides and anatomical models for cases requiring complex operative planning.[5] Here we present the case of a mediastinal eMRT wherein 3D printing was critical for preoperative and intraoperative planning. Institutional review board approval was not required; the parent provided informed consent for publication.

CASE REPORT

A 6-year-old otherwise healthy boy presented with 5 weeks of cough, abdominal pain, vomiting, and weight loss. Magnetic resonance imaging identified a large posterior mediastinal mass; subsequent biopsy revealed a small round blue cell tumor. Chemotherapy was initiated with topotecan/cyclophosphamide. Pathologic and molecular characterization confirmed eMRT, and chemotherapy was switched to vincristine, doxorubicin, and cyclophosphamide. Despite clinical improvement, follow-up computed tomography imaging after 3 cycles of chemotherapy demonstrated no significant change in tumor size (8.5 × 5.1 × 7.7 cm) with mass effect on the trachea, proximal bronchi, left atrium and bilateral pulmonary arteries, >180° encasement of the descending aorta, and abutment of the esophagus (Figure 1). Given the difficult anatomic location and involvement of adjacent structures, the patient was referred to our center for surgery. The decision to proceed with resection was based on symptom palliation and improving quality of life because the existing respiratory and cardiovascular compromise were expected to worsen with tumor growth.

FIGURE 1. Three-dimensional rendering of tumor (*yellow*), heart (*red/blue*), and trachea (*white*); lateral view.

For preoperative planning, a 3D model of the patient's mediastinum was segmented from computed tomography data using Materialise online 3D printing service and printed on a Stratasys J750 PolyJet printer (Figure 2). A life-sized physical model was chosen over interactive 3D software to provide better fidelity for preoperative planning, intraoperative guidance, and teaching.

Before the procedure, bronchoscopy, esophagoscopy, and transesophageal echocardiography were performed to ensure lumen patency and to assess left atrial compression. Clamshell thoracotomy was performed by thoracic surgeons who specialize in adult surgery, with the expandable view providing improved exposure of the pulmonary hilum and aortic arch with the heart retracted anteriorly. Cardiac surgeons who specialize in congenital conditions opened the pericardium and placed the patient on extracorporeal membrane oxygenation, facilitating anterior heart retraction and exposure of the posterior mediastinum. Cardiopulmonary bypass was not utilized due to a higher activated clotting time and potentially increased raw surface bleeding during dissection. Due to concern for mass concealment of vascular injury, the aortic arch and descending thoracic aorta were circumferentially dissected to obtain a clamp location if needed. General and congenital cardiac surgeons specializing in pediatric patients dissected the tumor inferiorly from the left atrium (Figure 3), then progressed posteriorly to dissect from the esophagus. The posterior pericardium was incised to gain exposure to the esophagus, and it was later removed due to investment. Given the tumor's encasement of the aorta, careful vascular dissection and frequent intraoperative referencing of the 3D model were required. The tumor was dissected posteriorly until freed from the airway, then from the pulmonary vessels and inferior vena cava (Figure 3), ultimately producing a complete macroscopic resection. Pathology demonstrated negative margins, 99% necrosis due to chemotherapy effect, and residual atypical rhabdoid cells with somatic biallelic loss of *SMARCB1*.

The patient tolerated the operation well and was discharged without complications. He continued with adjuvant chemotherapy (vincristine, doxorubicin, and cyclophosphamide with cyclophosphamide, carboplatin, and etoposide) for a total of 7 courses of chemotherapy. Concurrent proton beam radiation therapy (45 Gy) was given based on higher probability of survival.[3] The patient completed his multimodal therapy 6 months ago and currently is without any evidence of disease.

DISCUSSION

Surrounded by vital structures of the cardiovascular, pulmonary, and gastrointestinal systems, the posterior mediastinum is difficult to access operatively and predisposes to surgical damage of these structures. This challenge was exacerbated in our case by the large tumor size, small anatomic space of this pediatric patient, and the tumor's encasement of several critical structures. The use of a 3D model for pre- and intraoperative planning, paired with the expertise afforded by a multidisciplinary team of thoracic, congenital cardiac, and pediatric surgeons at a large academic center, was

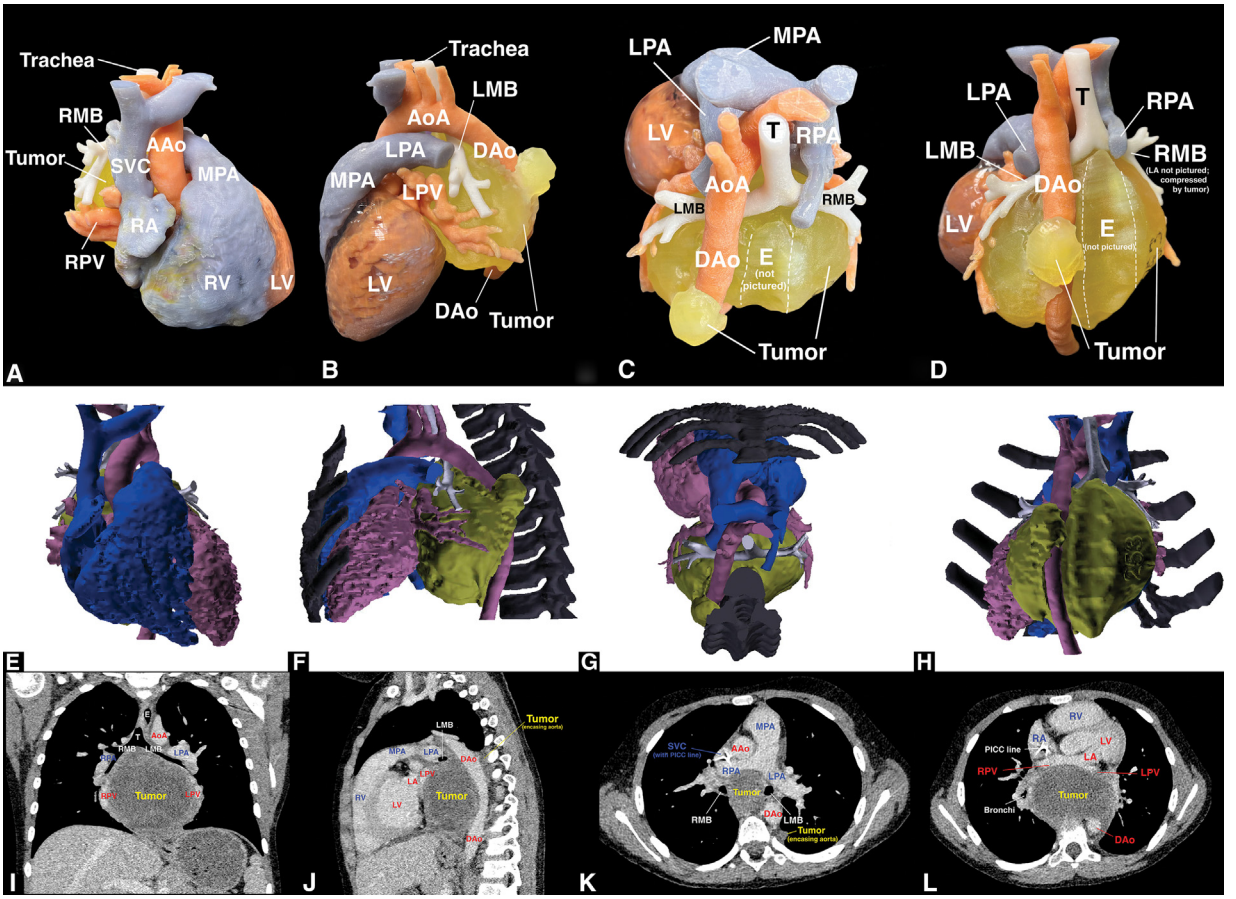

FIGURE 2. Three-dimensional-printed model (*top*, *middle*) of tumor in relation to mediastinal structures and preoperative computed tomography scan (*bottom*). Anterior/coronal view (A, E, I), lateral/sagittal view (B, F, J), superior/axial view (C, G, K, L), posterior view (D, H). *RMB*, Right main bronchi; *AAo*, ascending aorta; *SVC*, superior vena cava; *MPA*, main pulmonary artery; *RA*, right atrium; *RPV*, right pulmonary vein; *RV*, right ventricle; *LV*, left ventricle; *LMB*, left main bronchi; *AoA*, aortic arch; *LPA*, left pulmonary artery; *DAo*, descending aorta; *LPV*, left pulmonary vein; *Tumor*, malignant rhabdoid tumor; *T*, trachea; *RPA*, right pulmonary artery; *E*, esophagus; *LA*, left atrium.

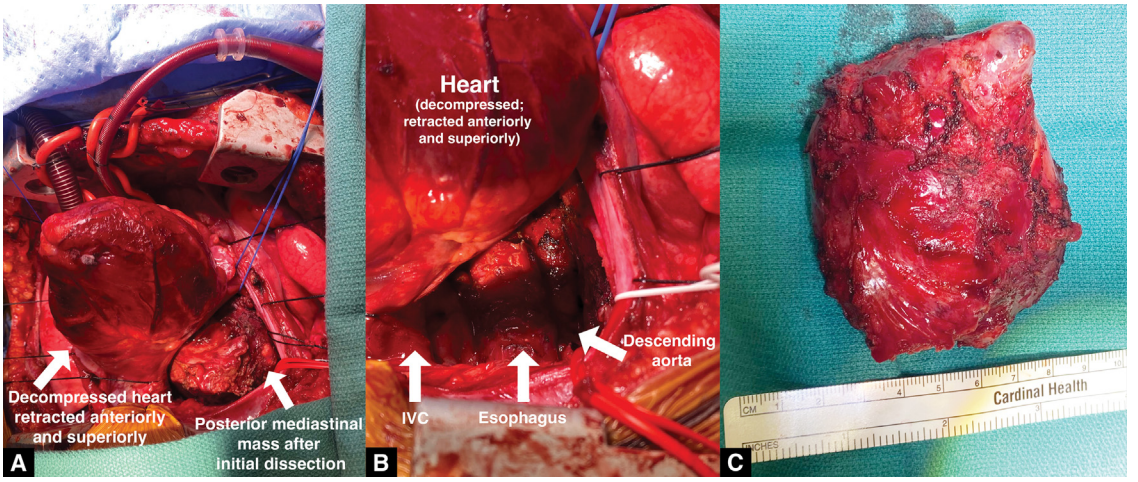

FIGURE 3. Mediastinal malignant rhabdoid tumor (*MRT*), posterior to heart (on extracorporeal membrane oxygenation) during dissection of the tumor from the left atrium (A). Tumor bed after resection of MRT, with preservation of descending aorta, esophagus, trachea, and inferior vena cava (B). Tumor specimen after resection (C).

essential in the planning and execution of a safe and successful R0 resection.

Conflict of Interest Statement

The authors reported no conflicts of interest.

The *Journal* policy requires editors and reviewers to disclose conflicts of interest and to decline handling manuscripts for which they may have a conflict of interest. The editors and reviewers of this article have no conflicts of interest.

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