Resection of large mediastinal germ cell tumor in an adolescent

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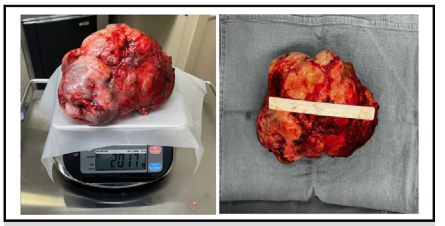

Giant mediastinal germ cell tumor.

CENTRAL MESSAGE

En bloc removal of giant mediastinal tumor is possible even in pediatric patients, and this technique may contribute to the prevention of residual tumor.

Germ cell tumors (GCTs) are neoplasms that arise from the pluripotent cells of the embryo, typically originating in the gonads. However, in approximately 2% to 5% of cases, they develop in extragonadal locations such as the mediastinum, retroperitoneum, or pineal gland.[1] The clinical presentation of primary mediastinal GCTs can vary widely, ranging from incidental detection on imaging studies to severe, life-threatening symptoms, including respiratory distress or cardiovascular compromise. Large intrathoracic masses pose significant challenges both for surgical resection and anesthesia management. Surgeons often face difficulties related to tumor accessibility, whereas anesthesiologists must contend with issues such as compression of the contralateral lung or obstruction of venous return, which can complicate positioning during surgery.[2] In this Case Report, we describe a case of a large primary mediastinal GCT that was successfully removed through a total en bloc surgical approach. Institutional review board approval was not required for this case report, and informed written consent was waived by the institutional review board/ethical review board for the publication.

CASE PRESENTATION

A 12-year-old boy with Klinefelter syndrome and attention-deficit hyperactivity disorder presented to our center with a history of right-sided chest pain with associated difficulty breathing. He underwent a radiographic examination, which showed a large mass in the mediastinum (Figure 1, *A*). The large mass (155 mm × 137 mm × 99 mm) was confirmed on computed tomography scan of the chest (Figure 2), and 3-dimensional construction (Figure 3) showed a compression on right atrium.

For further treatment, he was referred to the oncology department, and malignant GCT was diagnosed by mediastinal biopsy. Laboratory investigations showed alpha fetoprotein (AFP) and β-human chorionic gonadotropin (β-HCG) were elevated (AFP = 394.9 ng/m; β-HCG = 5870 mIU/mL). Therefore, combined chemotherapy with etoposide, bleomycin, and cisplatin was subsequently initiated. However, when 4 cycles of chemotherapy were finished, he underwent a computed tomography scan of chest again, which indicated that there was no obvious improvement. Therefore, the patient underwent surgical resection of the tumor after informed consent was obtained.

The surgery was performed via sternotomy. A plane of dissection was accomplished anterior to the pericardium and the innominate vein, primarily by blunt digital dissection. Bleeding was controlled using electrocautery. Blunt dissection was used to dissect the tumor from the right lung, superior vena cava, left innominate vein, and aorta. The right chest was cleared of a tumor weighing 2017 g via total en bloc resection (Figure 4). After removal of the tumor, the right lung was reinflated completely to fill the chest cavity. At the end of the resection, no tumor was evident. A right-sided chest tube was placed to the pleural space.

Postoperatively, the patient was extubated on first day. The postoperative course was completed successfully. No abnormal findings were detected on a postoperative

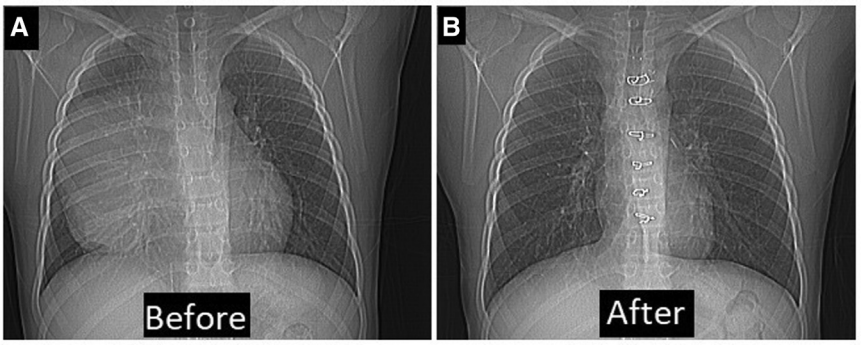

FIGURE 1. Radiographic images showing (A) before and (B) after surgical removal of the tumor.

radiograph of the chest (Figure 1, *B*). The patient was discharged on eighth postoperative day. The pathologic examination of the mass specimen revealed GCT with teratomatous elements.

DISCUSSION

GCTs are a heterogeneous group of tumors that primarily arise from the gonads. However, extragonadal GCTs, which account for approximately 5% of all GCT cases, are increasingly recognized in clinical practice.[3] These tumors can occur at a variety of extragonadal sites, including the mediastinum, retroperitoneum, brain, and less commonly in the lungs, liver, prostate, and omentum. Although the origins of extragonadal GCTs remain incompletely understood, it is hypothesized that abnormal migration of germ cells during embryogenesis may contribute to their ectopic presence in these sites.[4] These tumors can act as reservoirs for significant genetic, hematologic, and immunologic functions, potentially impacting surrounding organs as well. Despite their low frequency, their clinical implications are profound, often leading to significant diagnostic challenges and requiring an individualized therapeutic approach.

The clinical manifestations of extragonadal GCTs vary considerably, depending on tumor size, location, and organ involvement. Symptoms often arise from the compressive effects of large tumors, which can cause respiratory or cardiovascular compromise, including cough, shortness of breath, and chest pain. Rupture of the tumor can lead to pleural or pericardial effusions, further complicating the clinical picture.[5] These tumors can grow to enormous sizes, exacerbating symptoms and requiring urgent medical intervention. Konstantinov and Fricke[6] presented an intrathoracic mass occupying the mediastinum and compressing the superior vena cava, causing facial edema in their study. In our case, the tumor's large size and mediastinal location posed a unique set of diagnostic and therapeutic challenges; however, it did not cause clinical facial edema.

Preoperative biopsy remains crucial in confirming the diagnosis of GCT and distinguishing it from other mediastinal masses, particularly lymphoma. Radiologic imaging

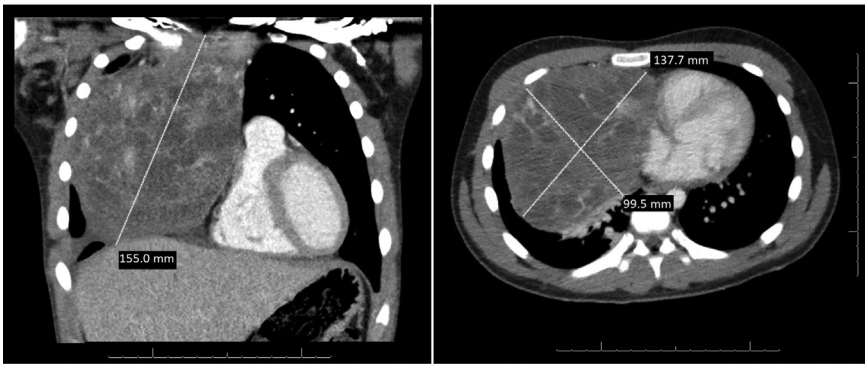

FIGURE 2. Tumor size is shown in axial and coronal planes on contrast-enhanced computed tomography.

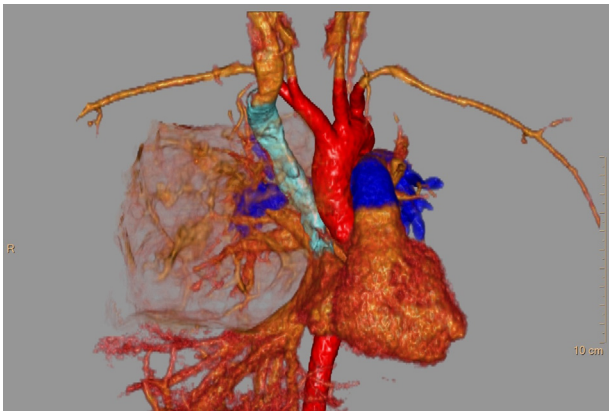

FIGURE 3. Compression of the tumor on the right atrium in a 3-dimensional configuration is shown on contrast-enhanced computed tomography.

plays a critical role in localizing the tumor and guiding biopsy procedures. Fine-needle aspiration or core biopsy, under imaging guidance, allows for tissue confirmation of the tumor type and ensures that treatment decisions are determined on the basis of accurate pathology.

Tumor markers are instrumental in diagnosing GCTs and assessing their response to chemotherapy. In particular, AFP and β-HCG are commonly elevated in nonseminomatous GCTs and serve as vital indicators of disease activity and treatment response.[7] In our case, there was no marked reduction in AFP and β-HCG levels after chemotherapy, and therefore the patient had to undergo surgical removal.

Surgical management of GCTs, especially those located in the mediastinum, often requires a tailored approach depending on the tumor's size and extent. Although minimally invasive techniques such as laparoscopy have been successfully used for smaller tumors, large mediastinal masses necessitate more invasive approaches, such as median sternotomy or clamshell incisions.[8] The choice of surgical approach is guided by the need for optimal exposure, as well as the tumor's proximity to critical structures such as the heart, lungs, and great vessels. In some cases, lower sternotomy or anterior thoracotomy can be chosen to provide better cosmetic results. However, in this case of tumor located high on the superior vena cava, we had to perform a full sternotomy to ensure adequate access and tumor removal, as it provides better exposure compared with other methods for large tumors.

In a review of the literature, Huang and colleagues[9] reported a successful en bloc resection of a mediastinal tumor in an adult patient in their article. Our case presentation shows that total en bloc resection is possible for tumors occupying the entire mediastinum and hemithorax even in pediatric patients.

CONCLUSIONS

In conclusion, mediastinal GCTs present unique diagnostic, and therapeutic challenges. Although chemotherapy remains the primary treatment modality, surgery plays a crucial role in managing large, residual tumors. En bloc removal is possible even in pediatric patient and this technique may contribute to the prevention of residual tumor. Given the complex nature of these tumors, a multidisciplinary approach is necessary to optimize patient outcomes and address the wide range of potential complications associated with both the tumor and its treatment.

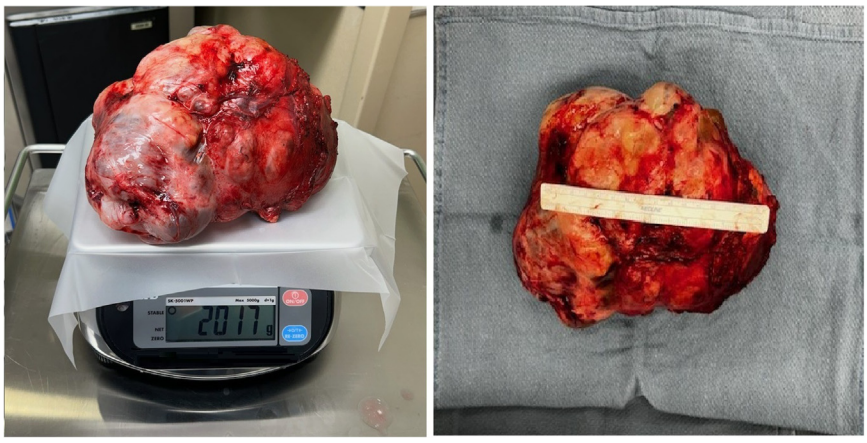

FIGURE 4. Macroscopic image after total en bloc removal of the tumor.

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 or reviewing 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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