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Case Report

Synchronous Whole Sternal Metastasis from Primary Malignant Mediastinal Germ Cell Tumor in a Teenager

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Abstract

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Primary malignant germ cell tumor (GCT) of the mediastinum is a rare entity with an incidence ranging from 1 to 2% of all childhood cancers. Though a few cases of bone metastasis from mediastinal GCTs have been reported, synchronous whole sternal metastasis from primary malignant mediastinal GCTs is very rare. We report such a rare manifestation in a teenager, emphasizing the need for a multidisciplinary treatment approach with neoadjuvant chemotherapy, surgical expertise for en bloc excision of the mediastinal mass along with whole sternal resection, and reconstruction in a dedicated cancer care center for better oncological outcomes.

Keywords

- malignant mediastinal germ cell tumor
- sternal metastasis
- sternal resection and reconstruction
- childhood cancer

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Introduction

Primary malignant germ cell tumors (GCTs) rarely arise from the mediastinum, with the incidence ranging from 1 to 2% of all childhood cancers.[1] Though a few cases of bone metastasis from mediastinal GCTs have been reported,[2] synchronous whole sternal metastasis from a primary malignant mediastinal GCT (PMMGCT) is very rare. Here, we report the clinical details and management of such an unusual presentation in a teenager.

Case Report

A 15-year-old boy presented with painless progressively increasing a 5×5 cm swelling over the sternal region for 3 months. Laboratory parameters were unremarkable except for an elevated serum alpha-fetoprotein (AFP): 47,010 ng/mL. A contrast-enhanced computed tomography (CECT) of the thorax displayed a well-defined heterogeneously enhancing globular mass measuring 13.0×12.5 cm arising from the left anterior mediastinum and left thorax, which was adherent to left hilar structures and chest wall (►Fig. 1A, C). Another distinct heterogeneously enhancing mass lesion was noted encasing the manubrium and the body of the sternum (►Fig. 1B, C). CECT of the abdomen and pelvis did not show significant retroperitoneal lymph nodes or any distant metastasis. An ultrasound of the scrotum ruled out a testicular pathology.

In the multidisciplinary tumor board discussion, a decision was made to proceed with neoadjuvant chemotherapy followed by surgery. The child was given four cycles of PEb (bleomycin: 15 mg/m^2 on day 1 only, etoposide: 100 mg/m^2 day 1 to day 5, cisplatinum: 20 mg/m^2 day 1 to day 5, and etoposide: 100 mg/m^2 day 1 to day 5) with an appropriate log fall of serum AFP after each cycle (►Fig. 1D). Response

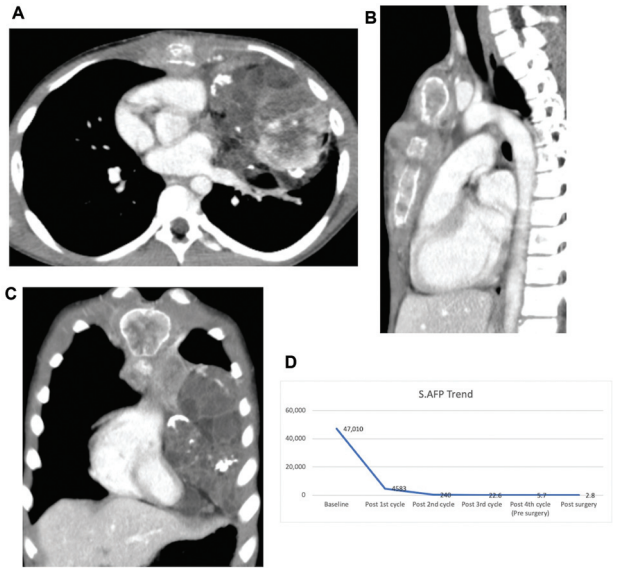

Fig. 1 (A–C) Contrast-enhanced computed tomography (CECT) scan showing the mediastinal germ cell tumor (GCT) with whole sternal metastasis. (D) Serum alpha-fetoprotein (AFP) levels during treatment.

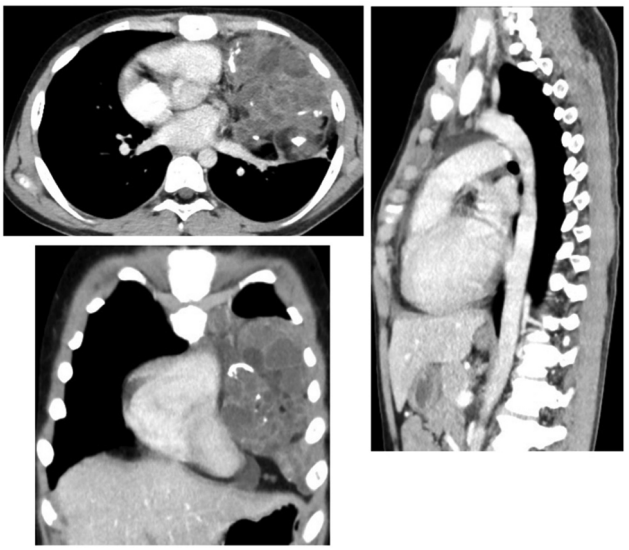

Fig. 2 Response assessment contrast-enhanced computed tomography (CECT) following chemotherapy showing partial response.

assessment CECT showed partial response (►Fig. 2). He was planned for excision of the mediastinal mass with whole sternal resection and reconstruction after adequate optimization. Because of the impact of the coronavirus disease 2019 pandemic, pulmonary functions could not be assessed due to the hospital policy of avoiding tests involving an aerosol generation. He was advised regular chest physiotherapy and incentive spirometry. Six-minute walk test and cardiac evaluation were normal. CECT thorax prior to surgery had no evidence to suggest bleomycin toxicity.

After thoracic epidural catheter insertion, the patient was intubated using a double lumen endotracheal tube and was placed in supine position with a 30-degree tilt to the right side. The mass was approached through a left anterolateral thoracotomy at the 5th intercostal space after preserving the pectoralis muscle. The tumor was removed en bloc with all fibrofatty tissue in the anterior mediastinum following meticulous dissection along phrenic nerve and left hilar structures. For the sternal resection, a midline vertical skin incision was placed and was connected to the thoracotomy incision. The first step was to identify and ligate the left internal mammary vessels. The suprasternal notch was dissected and connected to the substernal space. Brachiocephalic trunk was safeguarded. Anteriorly, whole sternum was exposed including xyphoid process. All ribs attached to left side of the sternum were transected at the costochondral junction. Left clavicle was transected at the manubrio-clavicular joint. The same procedure was repeated on the right side. En bloc resection of the sternum was performed (►Fig. 3A, E).

Left pectoralis muscle flap was harvested and tunneled through the left 2nd intercostal space to cover the sternal bed (►Fig. 3B). Sternal imprint was taken over sterile cloth using the specimen and the corresponding costochondral junctions were marked. An artificial sternum with corresponding holes for future fixation of clavicles and ribs was created using polymethyl methacrylate (PMMA). A layer of bone cement was sandwiched between two layers of polypropylene mesh

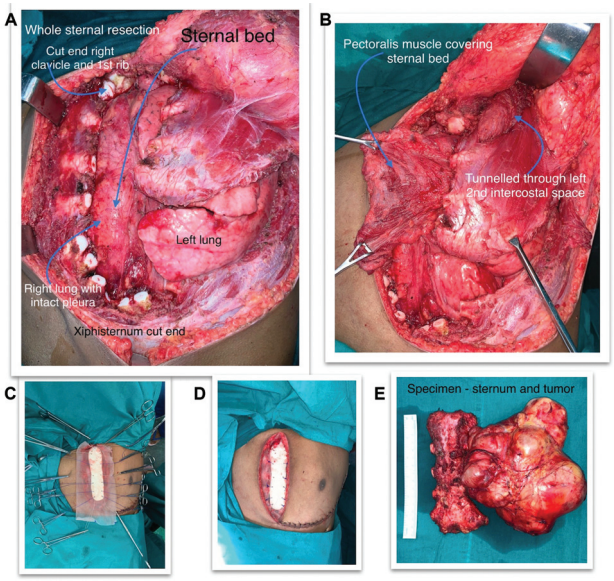

Fig. 3 (**A**–**D**) Sternal resection and reconstruction. (**E**) Specimen.

and fixed to the cut end of the clavicles and ribs with alternate sternal wire and no.1 polypropylene sutures (►**Fig. 3C, D**). Bone cement and mesh were covered with subcutaneous tissue and the thoracotomy was closed. The duration of the procedure was 7 hours with a blood loss of 200 mL.

The postoperative period was uneventful and the patient was discharged on the 8th day. Histopathology of the resected specimen showed a postpubertal/adult extragonadal GCT without any capsular breach with residual viable mature and scant/very focal viable immature teratomatous elements. The sternum had only the residual viable mature teratomatous elements. Posttherapy tumor necrosis was 40 to 50%. He belonged to at least stage III of Children's Oncology Group staging for malignant GCT and as per Moran and Suster staging for mediastinal GCT, he was at least stage IIIA.

He received two more cycles of adjuvant PEb post-3 weeks of surgery (total 6 cycles of PEb). Currently, he is on regular follow-up without any symptoms and a normal serum AFP.

Discussion

PMMGCTs are rare, and the most common histological types include non-seminomatous GCT, mixed type, and seminoma.[1] Distant metastasis from PMMGCT has been reported with the most common site being lung.[3] Bone metastasis from PMMGCT is extremely rare with vertebra being the most common site.[3] There are no reports available on the synchronous whole sternal metastasis arising from PMMGCT to our knowledge.

Mediastinal GCTs have an inferior prognosis compared to gonadal GCTs.[3] In recent times, the survival outcomes have dramatically improved (approaches 70–80%) with the introduction of neoadjuvant chemotherapy and an improved experience with major surgical resection/reconstruction in mediastinal GCTs.[3] Our patient was given a total of six cycles of PEb instead of conventional four PEb cycles as a strategy to obviate the anticipated poor prognosis in this particular age group.

In our case, biopsy was deferred in view of a high baseline serum level of AFP with imaging features indicative of mediastinal malignant non-seminomatous GCT. Even though neoadjuvant chemotherapy makes surgery difficult due to inflammation and fibrosis, it has certain advantages like systemic control, tumor shrinkage, less morbid resection/reconstruction, and prevents capsule rupture, which improves the rate of R0 resection. The most important prognostic factors in PMMGCT are completeness of surgical resection, presence or absence of viable tumor reflected by tumor markers, lung metastasis, and extrapulmonary visceral metastasis.

Sternal reconstruction can be performed by auto- or allografts of bone, muscle, fascia, synthetic mesh, PMMA bone cement, silicon, or titanium plates alone or in combination depending on the extent of the defect. In total sternal resection, aim of reconstruction is to provide rigid prosthetic replacement to prevent paradoxical respiration and to protect the anterior mediastinal structures.[4] In this case, molded PMMA was sandwiched between polypropylene mesh (Marlex sandwich technique)[5] due to its solidity, long-term tolerability, easy availability, and low cost. Use of soft tissue covers prevent mesh and bone cement from coming into direct contact with the pericardium and major vessels of mediastinum.

Conclusion

Whole sternal metastasis from primary malignant GCT is extremely rare. A multidisciplinary treatment approach with neoadjuvant chemotherapy, surgical expertise for en bloc excision of mediastinal mass, whole sternal resection, and reconstruction in a dedicated cancer care center improves the oncological outcomes.

Ethics Approval

The institutional research committee has confirmed that no ethical approval is required.

Note

Written informed consent was obtained from the patient for participation in the study and for publication of their clinical details and images.

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Conflict of Interest

None declared.

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