

Naturally Occurring Mediastinal Teratoma with Malignant Transformation in an Adult Male

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We report a rare case of rhabdomyosarcoma spontaneously arising in an anterior mediastinal teratoma in a 47-year-old male. The patient was found to have an anterior mediastinal mass on a chest X-ray, which was taken two months before his presentation to Asan Medical Center. A subsequent computed tomography scan revealed an 8.9×7.1×8.0 cm heterogeneous mass in the anterior mediastinum. He underwent an excision via median sternotomy. The histopathologic study identified a mature teratoma with embryonal rhabdomyosarcoma.

Key words: 1. Mediastinal neoplasms
2. Rhabdomyosarcoma
3. Teratoma
4. Teratoma with malignant transformation

CASE REPORT

A 47-year-old male presented with a mass in the anterior mediastinum. The mass was incidentally detected on a chest X-ray during the evaluation of a minor trauma that he had sustained two months prior to his presentation to Asan Medical Center (Fig. 1). As such, he was referred to our hospital for further evaluation. On his admission, a chest computed tomography (CT) scan was taken, which revealed an 8.9 cm×7.1 cm×8.0 cm heterogeneous mass in the anterior mediastinum. The mass was located anterior to the ascending aorta and the main pulmonary artery in the left hemithorax (Fig. 2). The mass was well demarcated without the overt signs of invasion to the adjacent structures. Also, there was neither pleural effusion nor lymph node enlargement. The differential diagnosis for this tumor based on the radiologic findings indicated a thymoma, a germ cell tumor, or a mediastinal

sarcoma. Due to the proximity to the aorta and pulmonary artery, a pre-operative tissue biopsy was not considered. Instead, it was decided that the patient undergo an excision. The procedure was performed via median sternotomy. Intraoperatively, a well-encapsulated mass was identified and noted to arise from the thymus. There was neither adhesion nor invasion to the adjacent structures such as the aorta, pulmonary arteries, or lung parenchyma. The tumor was excised completely with negative margins for a residual tumor. The intraoperative frozen biopsy revealed a rhabdomyosarcoma. The postoperative course did not show any particular issue; the patient was discharged on postoperative day four. The final pathology report identified a mature teratoma with embryonal rhabdomyosarcoma (Fig. 3). Postoperatively, the patient underwent five cycles of chemotherapy and subsequent radiotherapy. However, six months after the primary surgery, the patient was found to have a nodule, which was highly sus-

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pected to be a tumor recurrence, in the upper lobe of the left lung. On the serial follow-up CT scans, we learned that the nodule had grown in size up to 17 mm in the 8 months after primary surgery. After discussions with the medical oncology team, we decided to treat him with another cycle of chemotherapy, which shrunk the size of the nodule down to 15 mm (Fig. 4). Eleven months after the primary surgery, the patient underwent wedge resection of the nodule, which turned out to be a metastatic rhabdomyosarcoma from the primary mediastinal mass. As of the writing of this paper, the patient has finished the seventh cycle of chemotherapy and has not shown another tumor recurrence.

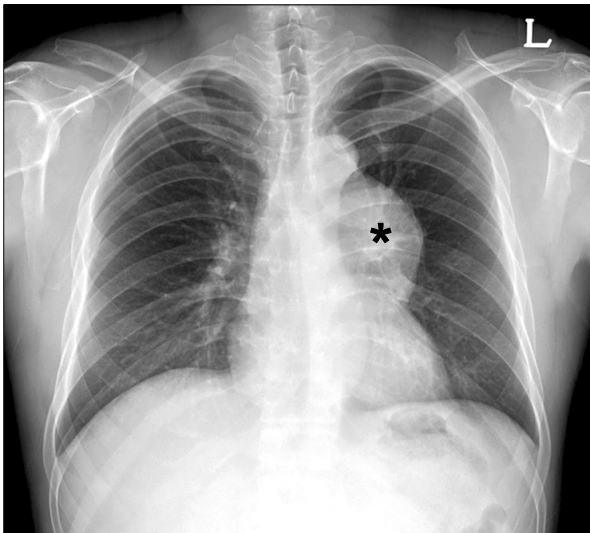


Fig. 1. Initial chest X-ray showing the huge mass in the anterior mediastinum (asterisk).

DISCUSSION

Teratoma can be found frequently in either the gonads or extragonadal organs such as the mediastinum, sacrococcygeum, or pineal region. However, teratoma with malignant transformation (TMT) is a very rare type of malignant teratoma. In addition, TMTs occurring in the mediastinum are known to be extremely rare [1]. TMT can be divided into two subgroups: TMTs induced by chemotherapy or radiotherapy, and spontaneously occurring TMTs [2]. According to our review of the literature, most cases of TMTs occurring in the mediastinum belong to the former subgroup, and TMTs spontaneously arising in the mediastinum are found in only five cases [1,3-6]. Three cases described TMTs with a malignant component of adenocarcinoma [1,5,6], while only two cases reported TMTs with a malignant component of rhabdomyosarcoma [3,4]: one in a child and the other in an adult male.

TMT is known to be very aggressive in its nature and the prognosis is generally pessimistic. Due to the rarity of the cases and the diversity of malignant components of TMTs, there is no widely accepted treatment strategy. Sarcomatous components of TMTs are especially highly resistant to chemotherapy, resulting in the alienation of chemotherapy as a strong treatment option. In contrast, complete surgical resection plays a vital role in the treatment of TMTs, and it is one of the most important prognostic factors [7]. According to previously reported cases of TMTs with malignant components of rhabdomyosarcoma, two patients died of the recurrence of malignancy in spite of undergoing adjuvant and neo-adjuvant chemotherapies in addition to surgical resections (one of them re-

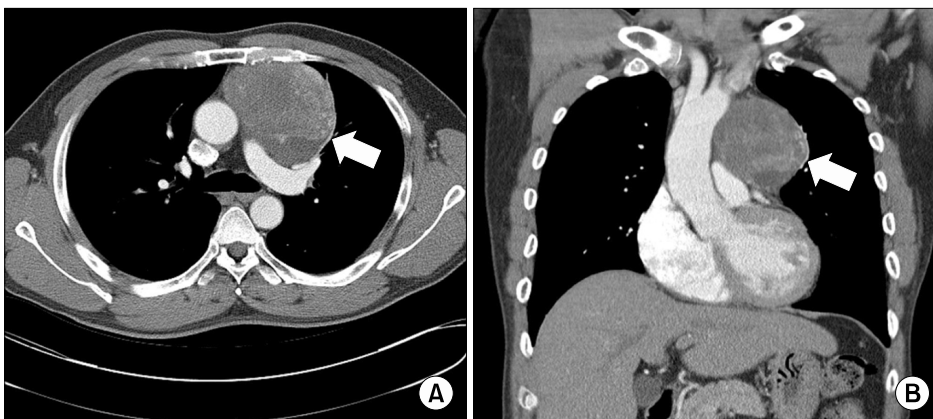


Fig. 2. (A) Axial view and (B) coronal view of contrast-enhanced computed tomography scan showing the well-demarcated large mediastinal mass (arrow) without signs of invasion to the adjacent structures.

ceived incomplete resection). On the other hand, in a case of a different malignant type of TMT, Shimizu et al. [6] reported six months of disease-free survival in a patient who had complete surgical resection and no chemotherapy. In our case, the patient had a combination of chemotherapy, radiotherapy, and surgical resection. Although the patient experienced tumor recurrence, he has survived for a year since the initial surgery. It is too early to draw a conclusion on the overall efficacy of our combined treatment or anticipate the life expectancy of the patient. However, we are, at least, convinced that the surgical

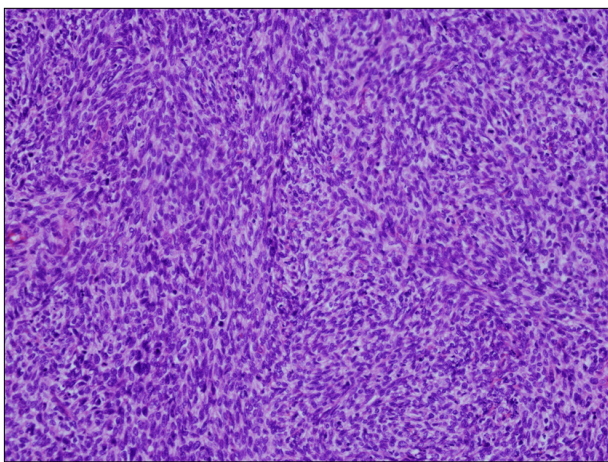


Fig. 3. Histologic findings of the resected rhabdomyosarcoma. A characteristic eccentric nucleus with eosinophilic cytoplasm is noted (H&E, ×200).

resection made a significant contribution to the patient's survival to the present day.

Due to the rarity of cases, little is known about the incidence, gender predilection, or clinical features of TMTs occurring in the mediastinum. However, it is noteworthy that TMTs with embryonal rhabdomyosarcoma in the mediastinum have all been reported in males in their forties, including our case.

To conclude, this report documents a rare case of TMT with embryonal rhabdomyosarcoma, which arose in the mediastinum without previous chemotherapy or irradiation. Due to the extreme rarity of such cases, there is no widely-accepted treatment strategy for this dismal disease entity. However, our experience in the present case and a review of the literature led us to conclude that complete surgical resection plays a vital role in good prognosis, irrespective of the accompaniment of chemotherapy or radiotherapy.

CONFLICT OF INTEREST

No potential conflict of interest relevant to this article was reported.

REFERENCES

1. Jung JI, Park SH, Park JG, Lee SH, Lee KY, Hahn ST. *Teratoma with malignant transformation in the anterior me-*

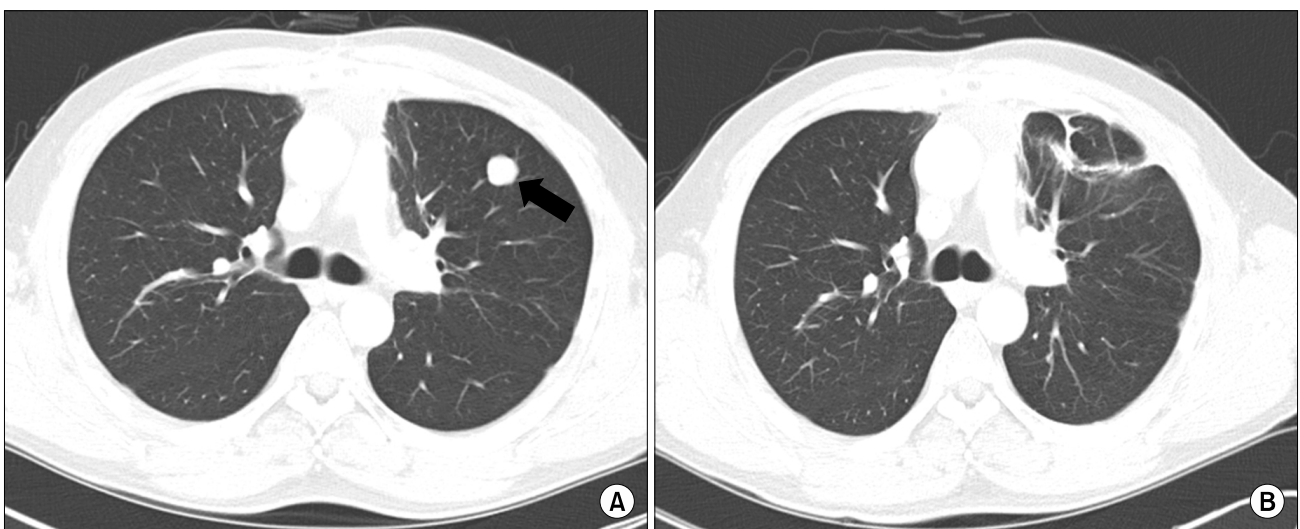


Fig. 4. (A) Computed tomography scan that demonstrates the metastatic nodule (arrow). (B) Scar tissue remains after resection without signs of tumor recurrence.

- diastinum: a case report.* Korean J Radiol 2000;1:162-4.
2. Morinaga S, Nomori H, Kobayashi R, Atsumi Y. *Well-differentiated adenocarcinoma arising from mature cystic teratoma of the mediastinum (teratoma with malignant transformation): report of a surgical case.* Am J Clin Pathol 1994;101:531-4.
 3. Takahashi M, Okumura N, Matsuoka T, Yamashina A, Nakano J, Kameyama K. *Teratoma with naturally occurring malignant transformation in a child.* Ann Thorac Cardiovasc Surg 2011; 17:588-90.
 4. Omezzine N, Khouatra C, Larive S, et al. *Rhabdomyosarcoma arising in mediastinal teratoma in an adult man: a case report.* Ann Oncol 2002;13:323-6.
 5. Chen C, Zheng H, Jiang S. *An unusual case of giant mediastinal teratoma with malignant transformation.* Ann Thorac Surg 2008;86:302-4.
 6. Shimizu K, Nakata M, Hiram Y, Akiyama T, Tanemoto K. *Teratoma with malignant transformation in the anterior mediastinum.* J Thorac Cardiovasc Surg 2008;136:225-7.
 7. Comiter CV, Kibel AS, Richie JP, Nucci MR, Renshaw AA. *Prognostic features of teratomas with malignant transformation: a clinicopathological study of 21 cases.* J Urol 1998;159:859-63.