

# An Aggressive Large Epithelioid Hemangioendothelioma of the Anterior Mediastinum in a Young Woman

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Hemangioendothelioma is a rare vascular tumor with involvement of the liver, brain, long bones, and lung. Among the 6 histological subtypes, epithelioid hemangioendothelioma (EHE) is the most aggressive. Its occurrence in the mediastinum is quite rare, and very few cases have been documented. The reported cases in the literature have described difficulties in the preoperative diagnosis due to the unusual histological appearance of the tumor. Immunohistochemistry remains the mainstay for a definitive diagnosis. Due to its low incidence, there is no standard treatment for mediastinal EHE, but curative resection is the preferred treatment option where possible, with chemotherapy used as an adjuvant treatment or in cases of widespread inoperable disease. The present case study describes an aggressive EHE occurring in an 18-year-old woman in the anterior mediastinum.

**Key words:** 1. Hemangioendothelioma  
2. Mediastinum  
3. Vascular neoplasms

## Case report

Vascular tumors of the mediastinum are very rare, accounting for fewer than 1%–1.5% of all mediastinal tumors. Vascular tumors of the mediastinum show atypical histological features. Epithelioid hemangioendothelioma (EHE) is a form of hemangioendothelioma. No more than 20 cases of EHE occurring in the mediastinum have been reported in the literature. In 1982, Weiss and Enzinger [1] were the first to describe EHE. They described the clinical and histological features of EHE as intermediate between angiosarcoma and benign hemangioma [1].

An 18-year-old woman from Kabul, Afghanistan attended Thoracic Surgical Clinic with chief complaints of low-grade fever, generalized weakness, and cough

that had lasted for 1 month. There was no history of loss of weight, appetite or night sweats. The patient denied any history of hemoptysis or breathing difficulty. The general physical examination was normal. A systemic examination showed decreased breath sounds on right side of the chest. Contrast-enhanced chest computed tomography (CT) revealed a large anterior mediastinal mass compressing the right lung (Fig. 1). Malignant germ cell tumor markers were negative. The cervical lymph node seen on the CT scan was reactive on an excision biopsy. CT-guided Tru-cut biopsy of the tumor indicated that it was a spindle-cell neoplasm.

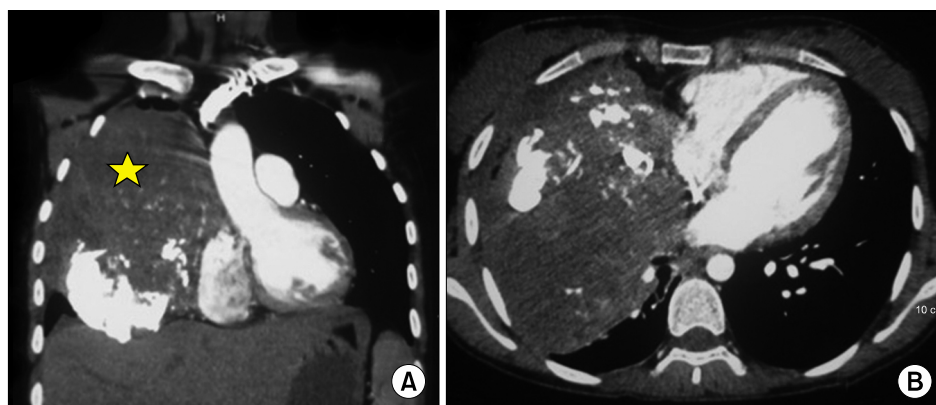
In view of the unifocal nature of the tumor, the patient was offered resection of the tumor. Surgical exposure of the tumor was achieved through right

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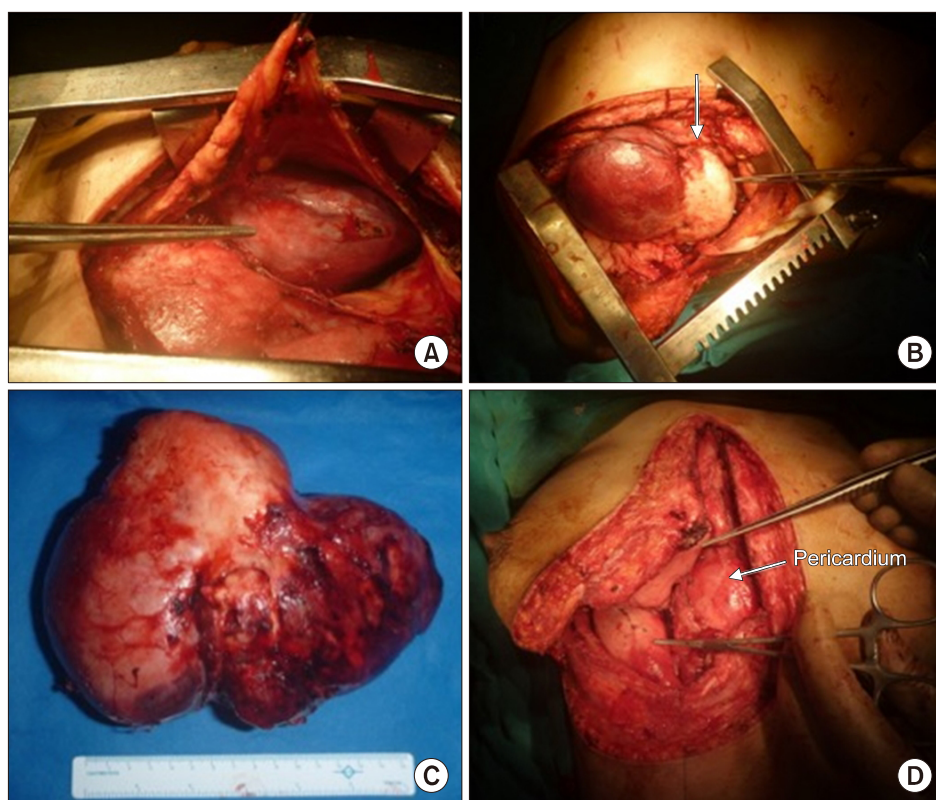
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**Fig. 1.** Chest computed tomography shows large anterior mediastinal mass with calcification and collapsed right lung (star).

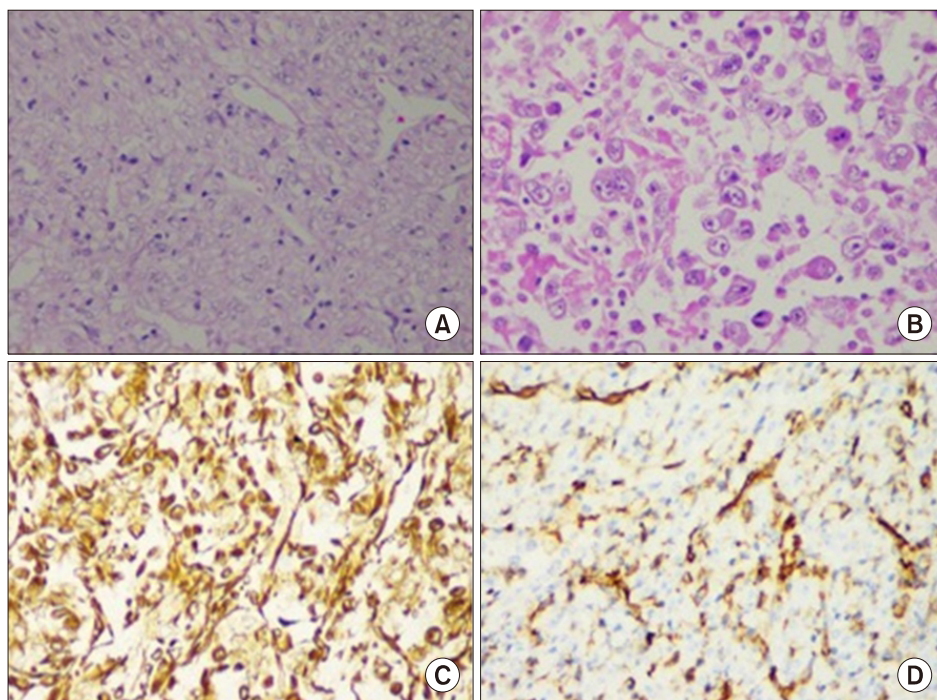


**Fig. 2.** Operative photographs showing large anterior mediastinal mass with venous channel connecting to right internal thoracic vein (A). Tumour with calcified lower half (B). Excised tumour of size 20x15 cm (arrow) (C). Completely expanded lung lobes post resection (D).

thoracosternotomy (hemi-clamshell). Intraoperative findings revealed a large anterior mediastinal mass measuring 20×15 cm that compressed the entire right lung and was adherent to the pericardium. The tumor appeared to arise from the anterior mediastinal tissue and was connected to the right internal thoracic vein (Fig. 2A). The tumor mass was lobulated, with variegated consistency. Areas of bony hard and soft consistency were present (Fig. 2B). No evidence of tumor adhesion to the lung or pleural ef-

fusion was found. The patient had an uneventful postoperative course and was discharged from the hospital on the fifth postoperative day.

Histopathologically, sections showed a tumor with a mixed spindle and epithelioid cell morphology. The tumor cells displayed moderate to marked nuclear pleomorphism, prominent nucleoli, and areas of necrosis. The tumor cells were positive for vimentin and CD31. The Ki67 labeling index was as high as 60%. Smooth muscle actin, S-100, desmin, cytoke-



**Fig. 3.** Photomicrographs (A, B) showing epithelioid cells in nests and cords separated by thin walled blood vessels (H&E, x200, x400). Photomicrographs of high power view (x400) showing the tumor cells were positive for vimentin (C) and CD31 (D).

atin 7, thyroid transcription factor-1, leukocyte common antigen, anaplastic lymphoma kinase-1, human melanoma black-45, and CD30 were negative in the tumor cells. In view of the morphological and immunohistochemical findings, a final diagnosis of EHE was made (Fig. 3).

The case was discussed at a multidisciplinary tumor board and adjuvant chemotherapy was recommended. The patient travelled back to her native country where she received adjuvant chemotherapy. The patient developed multiple distant-site metastasis 7 months after the surgical excision of the tumor and died while receiving follow-up from a medical oncologist. In this case, “written informed consent for academic using photography from the surgery” was provided by patient. So, institutional review board (IRB) review was waived by IRB.

## Discussion

Hemangioendothelioma is a rare vascular neoplasm that may exist in benign or malignant form. Six histopathological variants of it have been described: papillary intralymphatic hemangioendothelioma, retiform hemangioendothelioma, kaposiform hemangioendothelioma, EHE, pseudomyogenic hemangioendo-

thelioma, and composite hemangioendothelioma. EHE may present in soft tissue, bone, lungs, liver, and, on rare occasions, in other locations (central nervous system, lymph nodes, or breast). A mediastinal location is very rare, and few such cases have been reported in the literature. Patients with EHE of the mediastinum may present as asymptomatic or symptomatic.

Lamovec et al. [2] reported EHE of the anterior mediastinum in a 40-year-old female patient that was diagnosed by a chest X-ray and CT scan. An upper partial sternotomy was done. The tumor, measuring 5 cm, was seen to be quite extensive, involving the azygos vein anteriorly and the superior vena cava posteriorly.

Patrini et al. [3] reported EHE of the mediastinum in a 58-year-old male patient who presented with palpitations and chest discomfort. Liu et al. [4] observed EHE of the mediastinum attached to the pleura in a 29-year-old asymptomatic male patient. Isowa et al. [5] reported a case of EHE arising from the left brachiocephalic vein in the anterior superior mediastinum in a 41-year-old male patient. The tumor was resected by the Hemi-Plastron window technique. The patient received regular follow-up for 28 months, and there was no recurrence of the

lesion.

Suster et al. [6] reported 12 cases of EHE of the anterior mediastinum and observed that the mean age of occurrence was 49.2 years, and the patients comprised 9 males and 3 females. They found 5 asymptomatic patients and 7 who experienced symptoms due to compression of surrounding structures. In 5 cases, the tumors were locally infiltrative and in 7 cases, they were well circumscribed and encapsulated. In 9 cases, resection was performed, while in 2 cases, resection along with radiation therapy was performed, and in 1 case, radiation therapy, chemotherapy, and resection were performed [6]. Li et al. [7] reported a case of EHE encompassing the innominate vein in a 38-year-old male. The patient was treated with surgical resection and adjuvant chemotherapy and remained recurrence-free at 18 months of postoperative follow-up.

Histologically, EHE of the mediastinum shows epithelioid cells with intracytoplasmic vacuoles in a hyalinized or mucinous stroma. Spindle cells or osteoclast-like giant cells can occasionally be seen. The prognosis of EHE is uncertain. Some cases show indolent behavior, while others may show a more aggressive character, leading to death within a short time after diagnosis. Although chemotherapy was also initiated after surgical excision of the tumor mass, the patient in our case died within 7 months after surgery. The authors believe that the prognosis might be related to mitotic activity and the degree of cellular pleomorphism [5-8].

The treatment of EHE depends on the site and extent of tumor involvement, the site of metastasis, and specific individual factors. Surgical resection, radiotherapy, and chemotherapy all have been used to treat these masses. If the tumor is unifocal, then it should be entirely removed surgically, and if it exists at multiple sites, several medications, such as anti-angiogenic agents, vincristine, interferon, rapamycin, and radiation should be recommended to slow the growth of the tumor mass [7,8].

In conclusion, the mediastinum is an uncommon site of EHE, but EHE should be considered in the differential diagnosis of mediastinal tumors. In cases of EHE, the tumor can vary in size from very small to huge. The only currently available treatment is surgical excision.

### Conflict of interest

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

### References

1. Weiss SW, Enzinger FM. *Epithelioid hemangioendothelioma: a vascular tumor often mistaken for a carcinoma*. Cancer 1982;50:970-81.
2. Lamovec J, Sobel HJ, Zidar A, Jerman J. *Epithelioid hemangioendothelioma of the anterior mediastinum with osteoclast-like giant cells: light microscopic, immunohistochemical, and electron microscopic study*. Am J Clin Pathol 1990;93:813-7.
3. Patrini D, Scolamiero L, Khirya R, et al. *Mediastinal hemangioendothelioma: case report and review of the literature*. Respir Med Case Rep 2017;22:19-23.
4. Liu Y, Jiang YF, Wang YL, et al. *Epithelioid hemangioendothelioma of the anterior mediastinum: a case report*. Int J Clin Exp Pathol 2016;9:5746-52.
5. Isowa N, Hasegawa S, Mino M, Morimoto K, Wada H. *Mediastinal epithelioid hemangioendothelioma resected by hemi-plastron window technique*. Ann Thorac Surg 2002;74:567-9.
6. Suster S, Moran CA, Koss MN. *Epithelioid hemangioendothelioma of the anterior mediastinum: clinicopathologic, immunohistochemical, and ultrastructural analysis of 12 cases*. Am J Surg Pathol 1994;18:871-81.
7. Li XM, Lin XY, Xu HT, et al. *Mediastinal epithelioid hemangioendothelioma with abundant spindle cells and osteoclast-like giant cells mimicking malignant fibrous histiocytoma*. Diagn Pathol 2013;8:103.
8. Ferretti GR, Chiles C, Woodruff RD, Choplin RH. *Epithelioid hemangioendothelioma of the superior vena cava: computed tomography demonstration and review of the literature*. J Thorac Imaging 1998;13:45-8.