

Mediastinal Hemangiopericytoma with Neuroforamen Invasion

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Hemangiopericytoma is a rare tumor and accounts for about 1% of the vascular tumor. The most common site is the lower extremities, retroperitoneum/pelvis fossa, and the head and neck. Mediastinum origin is rare especially with neuroforamen invasion. Herein, we presented a case of 71-year-old woman with primary mediastinal tumor mass with progressive enlargement. She was treated by total T4 laminectomy and partial T3 + T5 laminectomy with intraspinal tumor removal and exploratory thoracotomy with resection of the tumor. She was under postoperative radiotherapy at our Oncology Department. The tumor was under well control until now.

Key words: Hemangiopericytoma, mediastinal tumor, spinal

INTRODUCTION

Hemangiopericytoma (HPC) is rare tumor and thought to be derived from fibroblasts. It was first described in 1942 by Stout and Murray. The tumor can be found almost everywhere in the body, but the most common sites are the lower extremities, retroperitoneum/pelvis fossa, and the head and neck. Herein, we report a rare case of posterior mediastinal HPC with neuroforamen invasion.

CASE REPORT

A 71-year-old female denied having any medical diseases. She was under annually physical checkup at the local hospital. An abnormal lung mass was found on the chest film and chest computed tomography (CT) scan disclosed a posterior mediastinal tumor at that time [Figure 1a].

She was then referred to our chest surgery department where surgical intervention was suggested. However, she hesitated due to no symptoms at that time. About 11 months later, she suffered from chest tightness and left chest pain associated with a nonproductive cough for 1-month. She went to a cardiovascular clinic where chest film revealed progressed lung mass. She was then came to our chest surgery department and was admitted to our ward for a surgical intervention.

Physical examination on admission revealed no abnormality. Laboratory findings include tumor markers were all within normal limit. The plain film revealed a 5 cm ×

5 cm mass above the aortic arch [Figure 1b]. Chest CT and magnetic resonance imaging (MRI) scan revealed a posterior mediastinum tumor mass with T4–T5 neuroforamen invasion [Figures 2 and 3]. The operation was firstly performed by neurosurgeon via a posterior approach. T4 lamina invasion was found during operation, and total T4 laminectomy and partial T3 + T5 laminectomy with intraspinal tumor removal was done. Chest surgeon then performed the left thoracotomy with the removal of the posterior mediastinal tumor [Figure 4].

Histological examination showed fascicles of hypercellular spindle cells with mitotic figures measuring up to 22/10 HPF, focal necrosis, focal storiform pattern, and staghorn-like vascular pattern, which compatible with HPC [Figure 5].

We consulted the radiation oncologist doctor for adjuvant radiation because of the microscopic residual tumor. The patient was well after the operation, and a chest tube was removed 3 days after the operation. She was discharged under stable condition 7 days after the operation [Figures 6 and 7].

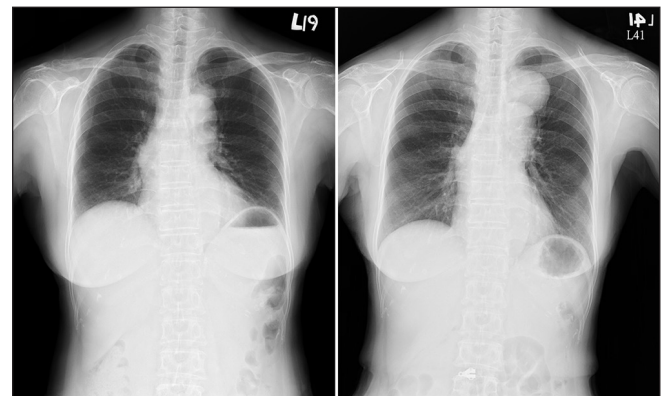


Figure 1: (a) The chest plain film revealed a tumor mass about 3 cm above the aortic arch. (b) The chest X-ray was taken about 11 months later which was showed an enlarged tumor mass

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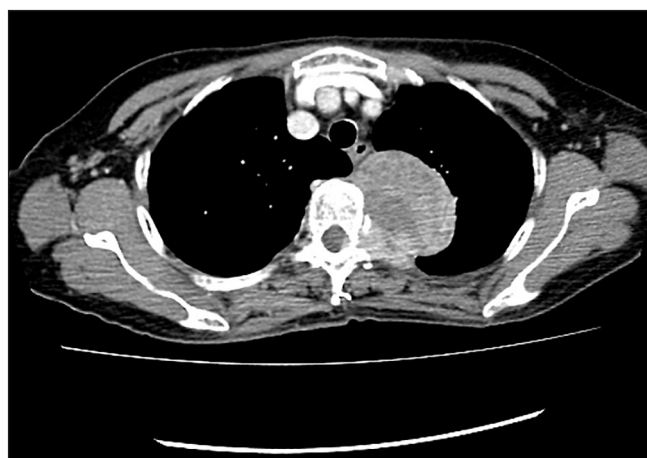


Figure 2: The chest computed tomography here revealed a dumbbell shape tumor mass measuring about 6 cm × 4 cm in size, identified over the posterior mediastinum of right chest with T4-T5 neuroforamen invasion



Figure 3: The magnetic resonance imaging revealed a large well-defined mass lesion with some calcification, cystic/necrotic change, and inhomogenous enhancement



Figure 4: The solid tumor mass taken from mediastinum is 5.3 × 5.0 × 2.3 cm in size. The tumor is grayish yellow, soft, with foci of necrosis

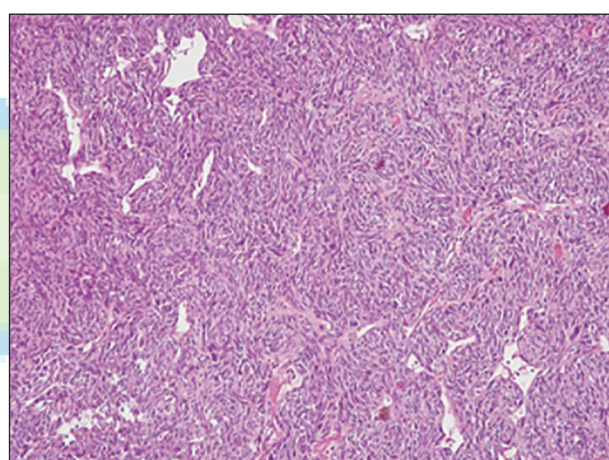


Figure 5: The histologic picture here showed stag-horn pattern

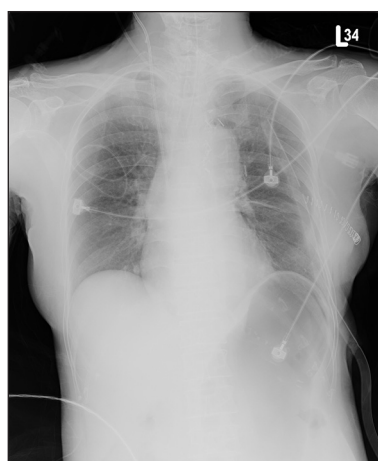


Figure 6: The chest X-ray here was taken before the patient discharged from our ward. Chest tube was removed without complications



Figure 7: The chest X-ray was taken after 3 months during out-patient department follow-up

DISCUSSION

HPC was first described in 1942 by Stout and Murray.¹ It is now thought to be arised from fibroblasts, a small subset of soft tissue sarcomas.² It is a rare tumor and accounts for about 1% of vascular tumors.³ The frequency of the tumor is rare, and no meaningful estimates of incidence had been reported. It primarily affects adults of age 20-70 years, with the median age in the 40 years.² Our patient is older than the mean age of the previous study. Previous study also revealed that it shows an equal sex distribution with similar prognosis.² Either benign or malignant, it can be found virtually at any site in the body. The most common site is the lower extremities, retroperitoneum/pelvis fossa, and the head and neck.^{2,4} Primary mediastinum HPC is very unusual.³ Our patient was leading a normal life before. Incidental tumor mass was found during annually physical checkup. Previous reports suggested that the mediastinum HPC was relative asymptomatic and frequently very large until it was diagnosed. However, some may presented with paraneoplastic syndromes such as hypoglycemia.^{2,4} In our case, the patient was initially asymptomatic and presented with neuropathy as the tumor progressed. The chest MRI confirmed the tumor invaded into the neuroforamen.

For the hemangiopericytoma, bronchoscopy, and percutaneous needle aspiration as the preoperative tools frequently fail to yield a diagnosis. Chest radiograph often showed a lobulated, well-circumscribed, homogeneous noncalcified but nonspecific soft tissue mass consist of our case.⁴⁻⁶ As for the CT scan, it delineates the extent of the tumor more accurately but still adds little diagnostically. The MRI is a distinct advantage over CT because the tumor is vascular origin. Therefore, MRI seems to be the proper tool for the preoperative survey. In our case, preoperative survey is brain CT, chest CT, chest MRI, whole body bone scan, and abdomen sonography and only posterior mediastinum tumor mass with neuroforamen invasion was found.

Surgery is the mainstay of treatment. Complete surgical resection of the tumor, whenever possible is suggested despite the high risk of intraoperative uncontrollable bleeding.^{2,4} Due to the tumor location, neurosurgeon and chest surgeon performed the operation together. However, the tumor size was so large and extent to preform the complete resection. Debulking operation was thus performed. Park and Araujo in 2009 Current Opinion in Oncology suggested that for those who are at risk of recurrent or unresectable ones, radiation is the alternative choice.² Thus, we consulted the radiation oncologist doctor for the adjuvant radiation therapy.

Until now, only limited published data showed the effectiveness of systemic chemotherapy. The most promising one was the doxorubicin-based chemotherapy. Owing to the tumor nature, anti-angiogenic agents are now underdeveloped and somewhat encouraging. Combination therapy with

temozolomide and bevacizumab is a potentially promising regimen.² According to the numerous previous reports, the tumor showed a relative a favorable clinical behavior, with a reported 10 years overall survival of 54-89% after primary surgical resection. About 15-20% display a more aggressive behavior leading to local recurrences or distant metastases despite surgery. In our case, aggressive and high grade picture was showed.² No reliable predicted factor for the prognosis was found until now. A previous study indicated that re-resection and/or metastasectomy for locally recurrent or metastatic disease is suggested by some authors and should be considered if technically feasible, but not always successful.² About 50% of the cases have been reported to recur within 5 years and usually within 2 years. Distant metastases to liver, brain and bone have all been reported. Thus, long-term follow-up is necessary.^{2,3}

CONCLUSION

HPC is a rare tumor. The diagnosed of HPC is controversial and currently concept about HPC is that the tumor is arised from fibroblast rather than pericyte. MRI is a proper preoperative tool in evaluating this tumor. Surgery is the mainstay of the therapy owing to the relatively insensitive to radiotherapy and cytotoxic chemotherapy. Newer therapies aim at inhibition of angiogenic pathways are under investigated. The recurrent disease usually occurs within 2 years and, therefore, a long-term careful follow-up is required.

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