Primary Mediastinal Hemangiopericytoma: A Case Report

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Primary mediastinal hemangiopericytomas are very rare tumors. We present a patient with this disease and discuss the clinical symptoms, diagnosis, and the correlation of radiographic features. A 53-year-old woman complained of the cough and the dyspnea for months before consultation. The chest X-rays and computed tomography revealed a well-defined intensive enhanced mass in the left middle mediastinum with necrotic changes and calcification within it. The patient underwent left thoracotomy with tumor resection, and a final diagnosis of primary mediastinal hemangiopericytoma was made. To our knowledge, only 20 cases of primary mediastinal hemangiopericytoma have been reported, including the largest original series presented by Feldman and Seaman [3-7]. We present a case of primary mediastinal hemangiopericytoma and discuss the clinical symptoms, the diagnosis and the correlation of radiographic features.

CASE REPORT

We present a 53-year-old woman with an abnormal shadow over left hilar region first noted on her chest X-ray in 1996, while she received routine health check-up at a local hospital. She did not pay much attention to it until the symptoms became worse. She came to our hospital for help on March 31, 2000 with complaints of cough, chest distress, and dyspnea which started several months before the consultation. She was admitted to our hospital on April 26, 2000 for further examination.

The chest X-ray film revealed a clearly demarcated opacity, measuring 7x7 cm, over the left hilar region (Fig. 1). The computed tomographic scan of the chest showed a well-defined enhanced mass over the left middle mediastinum, with necrotic changes and calcification within it (Fig. 2). There were no enlarged lymph nodes in the hilar region, and no
evidence of abnormalities of either lung. Bronchoscopy showed extrinsic compression with engorged vessels over the left main bronchus. Findings from abdominal sonography, whole body bone scan, and serum tumor markers (CEA, AFP) were unremarkable.

The patient received left thoracotomy with tumor resection on May 2, 2000. At operation, a huge encapsulated tumor located anterior to the left hilar region was found. The tumor grew between the left pulmonary artery and superior pulmonary vein extrapleurally, and extended deeply into the left lung and compressed the pulmonary vessels. Severe adhesions between the tumor and peripheral tissues including the lung and pulmonary vessels were noted, and these structures were injured during the dissection procedure. A total of 3400 ml of blood was lost during resection of the richly vascularized tumor.

Macroscopic analysis of the resected specimen revealed a well-demarcated and slightly lobulated soft tissue mass, measuring 9x6x6 cm. Histopathologically, the tumor was composed of a regular distribution of the pericytes in groups separated by capillary networks. Silver stain showed a framework of reticulin by which most of the tumor cells were surrounded (Fig. 4). These findings were characteristic of a hemangiopericytoma. After the operation, the patient was in stable condition and discharged on May 13, 2000.

**DISCUSSION**

Hemangiopericytomas are rare tumors which was first described by Schmidt in 1937 and named by Stout and Murray in 1942 [1]. They have been reported in various sites, and most commonly in the thigh, head and neck regions [2]. Tumors of this type arising in the mediastinum are quite rare. To our knowledge, only 20 cases of primary mediastinal hemangiopericytoma have been reported in the English literature [3-7]. This type of tumor has no predilection for age or sex, and constitutes only 6% of all primary tumors of the mediastinum [8]. The natural history of this tumor may be very long. It is locally aggressive and potentially malignant, which produces late local recurrences and/or metastases [9].

Clinically, the majority of patients with malignant thoracic hemangiopericytoma have complained of cough, pain, dyspnea at some stage of their illness, while such symptoms were comparatively uncommon with benign tumors. Spontaneous bleeding into the pleural space caused by the mediastinal hemangiopericytoma has also been reported [5]. In our patient,
symptoms including cough, chest distress and dyspnea were noted.

On computed tomography (CT), the findings in our patient were of a mass which was well defined, 7x7 cm, markedly enhanced over the left hilum, with central calcification and necrosis. The smooth interface with the adjacent lung and a broad base on the mediastinum indicated the tumor arisen in the mediastinum rather than the lung. Since an untreated lymphoma or untreated metastatic neoplasm in the lymph nodes almost never causes calcification [10], the preoperative differential diagnosis of such a markedly enhanced, calcified mediastinal mass included carcinoid tumor, teratoma, and Castleman’s disease. Bronchial carcinoids typically arise in the larger bronchi and may narrow, deform or obstruct the airway with consequent atelectasis, which was not consistent with our case. Teratomas mostly occur in the anterior mediastinum (in front of the aortic roots and main pulmonary artery) and may show areas of calcification, teeth, fat, or fat-fluid levels. Our patient had a middle mediastinal mass with no fat or fluid attenuation, so the diagnosis of teratoma was not likely. Castleman’s disease is a relatively rare disorder of lymphoid tissue, and the lesions are usually located in the mediastinum and range from 7 to 10 cm [11]. The lesions of Castleman’s disease may have central calcification and may be densely enhanced, which is similar to the results in our patient. However, 70% of patients with Castleman’s disease are younger than 30 years [12], and our patient was much older than that.

The radiographic features of primary mediastinal hemangiopericytoma were of a well demarcated, homogeneous soft-tissue mass of variable size. The computed tomography shows contrast medium enhancement, and sometimes a curvilinear encircling structure suggesting a large feeder vessel will be found. Some authors have stated that the diagnosis of this tumor should be considered when identifying such findings on the computed tomography although it is a rare cause of mediastinal mass [13]. However, this feature was not found in our patient, probably due to inadequate contrast infusion and scanning time.

**Figure 3.** Precontrast CT scan of the chest demonstrates central calcification within the tumor. The measured CT number of the calcification (arrowhead) is 238H.U.

**Figure 4.** Photomicrograph of the resected tumor. a. The tumor is rich in capillaries and small vessels (arrow) with a single layer of endothelium, around which there are tightly packed tumor cells (arrowhead). b. Silver stain: reticulin meshwork outlines the vessels (arrow) and surrounds tumor cells (arrowhead).
Rapid contrast infusion, dynamic scanning, and spiral technique may be helpful to demonstrate the feeder vessel in such a case. Nevertheless, there were some characteristic features in the CT images in our patient. First, the intensive enhancement of the tumor which measured CT number about 120 HU was noted in the postcontrast CT scan (Fig.2); this suggested that the tumor was highly vascularized. This finding was compatible with the histopathological features of hemangiopericytoma. Microscopically, hemangiopericytomas consist of tightly packed tumor cells around thin-walled, endothelium-lined vascular channels ranging from capillary-sized vessels to large gaping sinusoidal spaces [14]. Second, there were some necrosis and calcifications within the tumor. The calcifications were dense and of significant size, with measured CT number about 220 HU (Fig.3). Calcifications in a primary thoracic hemangiopericytoma have not been mentioned in previous reports in the literature [3-7]. To our knowledge, we present the first case of a mediastinal hemangiopericytoma with central calcifications.

Some investigators [15] have reported that magnetic resonance imaging (MRI) have a distinct advantage over CT in imaging hemangiopericytomas because they are vascular tumors and often containing large areas of hemorrhage. These characteristics are easily visible on MRI and help to define the interface between the tumor and contiguous structures, which CT cannot. Unfortunately, MRI scans were not performed in our case.

Although the findings yielded by CT with contrast, angiography, and MRI can be helpful in suggesting the presence of a hemangiopericytoma, an accurate preoperative diagnosis is difficult to reach [16]. The diagnosis of hemangiopericytoma requires pathologic proof. Some authors have emphasized the value of percutaneous biopsy before making a therapeutic decision [17]; however, physicians must bear in mind the risk of massive bleeding during biopsy of this richly vascular tumor [5].

Because experience in the management of this tumor is limited, controversy remains about the best way to manage it. Extended surgery has been the accepted primary mode of treatment [18], and pre-operative embolization has been reported helpful to reduce the menace of operative hemorrhaging [7]. Chemotherapy and radiotherapy regimens for this tumor have not yet been well established.

Approximately 80% of hemangiopericytomas are malignant and almost two-thirds of these eventually develop local or distant metastases [2]. The overall recurrence rate of hemangiopericytomas originating in any organ system is approximately 50%. However, primary mediastinal hemangiopericytomas are distinguished by early recurrence-about 40% recurring within 1 year of resection [18]. Therefore, cautious observation and long-term follow-up of each patient’s postoperative course is necessary.

REFERENCES

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原發性縱膈腔血管外皮細胞瘤：病例報告

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原發性縱膈腔血管外皮細胞瘤是非常罕見的腫瘤。我們提出一個病例並討論其臨床症狀、診斷、與放射學特徵。一位 50 歲女性病患主訴在求診前數個月開始出現咳嗽及呼吸困難的情形。胸部 X 光攝影及電腦斷層掃描發現在她的左側縱膈腔有一界限清楚的腫塊，腫塊內並有壞死及鈣化。此病患接受了腫瘤切除術，最後的診斷為原發性縱膈腔血管外皮細胞瘤。就我們所知，僅有 3 例原發性縱膈腔血管外皮細胞瘤曾被報告於英文文獻中，且尚無一例發現有鈣化的現象。我們在此提出首例觀察到中央鈣化的原發性縱膈腔血管外皮細胞瘤。

關鍵詞：血管外皮細胞瘤，縱膈腔，鈣化，電腦斷層攝影