Between January 1992 to December 1996, four patients with pathology-proven malignant retroperitoneal hemangiopericytomas were seen at our hospital. The clinical presentation, CT and pathological findings, treatment and outcome were reviewed. Two of the four hemangiopericytomas were found in abdominal retroperitoneum and the others in the pelvic retroperitoneum. The tumors were shown to have smooth margin, tended to be large (three of them were larger than 14 cm in its longest dimension) enough to displace but not invade the adjacent organs. The two abdominal hemangiopericytomas and one of the pelvic hemangiopericytomas showed heterogeneous enhancement with necrotic components in enhanced CT scan, the other one of pelvic hemangiopericytomas showed strong homogeneous enhancement without obvious necrotic component. In none of the patients, regional lymph node or intraabdominal metastasis was shown in the CT study. No recurrence was found in all of the patients followed up to 5 years after surgery.

Although hemangiopericytoma is a rare disease, it should be considered when a huge encapsulated retroperitoneal mass with strong enhancement and necrotic components is shown in CT scan study.

**Key words:** Computed tomography, Hemangiopericytoma, Retroperitoneum

---

Computed Tomography Evaluation of Retroperitoneal Hemangiopericytomas

SIU-CHEUNG CHAN¹ YUN-CHUNG CHEUNG¹ SHEUNG-FAT KO² LIANG-TZU CHANG³ KOOK-KUAN NG¹
KAR-WAI LUI¹ HO-FAI WONG¹ SHU-HANG NG¹

Department of Diagnostic Radiology¹, Chang Gang Memorial Hospital, Linkou Medical Center and Keelung Hospital
Department of Diagnostic Radiology², Chang Gang Memorial Hospital, Kaohsiung Medical Center
Department of Pathology³, Chang Gang Memorial Hospital, Linkou Medical Center and Keelung Hospital

Hemangiopericytoma is a rare vascular tumor representing approximately 1% of all vascular neoplasm [1] and approximately 25% of the hemangiopericytomas arise from the retroperitoneum and pelvic cavity [1-3]. To increase sensitivity of the diagnostic imaging modalities in the detection and characterization of retroperitoneal tumors needs detailed understanding of the pathologic-radiologic correlation of each type of tumor that arises in this region. However it is difficult to do so because of the rarity of these tumors. Only in a specialized referral institution is it possible to accumulate a series of cases rather than anecdotal experience. Herein, we present the computed tomography (CT) findings of this entity, and stress the common features of hemangiopericytoma found in the retroperitoneal spaces of the abdomen and pelvis.

**MATERIALS AND METHODS**

The cases of this retrospective review were obtained from the pathological data bank in our hospital from January 1992 to December 1996. Four patients with the diagnosis of retroperitoneal hemangiopericytoma by pathological examination were found, all of them had the CT examination before surgery. We reviewed all the cases with respect to clinical presentation, imaging data, pathological findings and outcome.

CT examinations were performed on Picker 1200 units (Picker International, Highland Heights, OH, USA) in two cases, and the other cases were performed on GE 9800 scanner (General Electric Medical Systems Group, Milwaukee, WI, USA) and Siemens Somatom (Somatom Emotion, Siemens, Germany) respectively. Contiguous 1.0-cm-thick axial sections were obtained from the diaphragm to the symphysis pubis in all patients. Contrast medium was given by intravenous injection of 100ml of Loversol 74% (Optiray, 350 mg/mL, Mallinckrodt, Canada) to all patients. The CT features were analyzed and focused...
on tumor size, location, shape, margin, calcification, attenuation value, contrast enhancement, and tumor staging (local invasion and distant metastasis).

For purpose of tumor localization, retroperitoneal space is defined as abdominal and pelvic. The abdominal retroperitoneum includes the anterior and posterior pararenal spaces as well as the perirenal spaces. The pelvic retroperitoneum is considered to be the continuation of the joined anterior and posterior pararenal spaces caudal to the inferior fusion of the anterior and posterior renal fasciae. All of the tumors in our study were confirmed in the abdominal and pelvic retroperitoneal spaces by surgical exploration. The diagnosis of hemangiopericytoma was based on histopathologic features: a tumor composed of a proliferation of pericytes surrounding branching, thin-walled vascular channels. Tumor cells had hyperchromatic nuclei with mild to moderate pleomorphism (Figure 1).

**RESULTS**

There were one man and three women with their age ranging from 34 to 50 years. The average age was 44 years. Two of the patients presented with a palpable abdominal mass and vague abdominal discomfort. The other two presented without palpable mass in abdomen and only complained of non-specific abdominal discomfort (Table 1). There was no complaint concerning other gastrointestinal symptoms in all patients. Blood examination was normal in all patients.

All patients received exploratory laparotomy after CT examination. During operation, the masses were found to be soft, well encapsulated and with adhesion but without direct invasion to the adjacent anatomic structures. Two tumors located in the abdominal retroperitoneum, the other two masses were in the pelvic retroperitoneum. The longest dimension of the masses ranged from 5 to 30 cm. Total excision of the tumors was done. Microscopically, all tumors showed tightly packed cells around thin-walled vascular channels ranging from capillary-sized vessels to large sinusoidal spaces. Spindle cells were noted in many places. There were focal hemorrhage, necrosis and a few mitotic figures. The histopathologic picture of all the tumors was compatible with hemangiopericytoma.

CT showed mass effect of huge tumors upon the adjacent anatomic structures in three of the four cases (Case 1, 2 and 3). In case 1, the bulky mass was disregarded as pancreatic lesion, because it attached closely to pancreatic tail, and displaced bowel loops outwardly. In case 2, the tumor located in the right upper abdomen and displaced the right kidney.
upwardly. In case 3, the tumor situated in the pelvic retroperitoneum, abutting to the superior border of the uterus and compressing the urinary bladder. In case 4, the tumor located between the abdominal aorta and the left psoas muscle, 3 cm below the lower pole of the left kidney, no mass effect was found. Obviously, the tumors were shown, on CT scan studies, to have a clear margin without direct invasion to the adjacent anatomic structures.

The mass in case 1 was measured 30 cm in its longest dimension. It showed heterogeneous enhancement in the soft-tissue portion with multiple low attenuation areas (Figure 2). The mass in case 2 was measured 25 cm in size, enhanced heterogeneously and contained multiple low attenuation areas (Figure 3). The pelvic retroperitoneal mass in case 3 was measured about 14 cm in the longest dimension and showed relatively homogeneous enhancement (Figure 4). The mass in case 4 was situated in the pelvic retroperitoneum, measured about 5 cm in diameter and presented heterogeneous enhancement (Figure 5). Regional necrotic areas were shown in all of the tumors (except case 3) in enhanced CT scans. None of the tumors revealed calcification.

There were no enlarged lymph nodes in the paraaortic, bilateral iliac chains or inguinal regions.
There was no evidence of intraabdominal metastasis in the CT scans of the abdomen and pelvis. All patients discharged in stable condition after surgical operation, and were tumor-free up to 5 years.

**DISCUSSION**

Hemangiopericytoma is a rare vascular tumor representing approximately 1% of the vascular tumors. It is derived from pericytes and was first described by Stout and Murray in 1942 [4]. Pericytes are cells with long processes that surround the capillaries and serve to alter the caliber of the capillary lumen. Hemangiopericytomases can occur anywhere in the body but are most commonly found in the lower extremity and the pelvis. Other sites in decreasing frequency are: head, neck, trunk, and upper extremity [5]. About 25% of hemangiopericytomas arise from the retroperitoneum and the pelvic cavity [1-3]. Retroperitoneal hemangiopericytoma tend to be encapsulated and growing slowly. It usually presents as a painless mass bulky enough to displace the adjacent structures. The median size of the tumors in reports was 6.5 cm, and the largest one was 30 cm in dimension [6]. Tumor size may be related to malignant potentiality, larger hemangiopericytomas are more frequently malignant than its smaller counterpart [2].

Hemangiopericytoma occurs equally in both sexes with a predilection to early middle age [3, 7]. In our series, there were three women and one man. Their age ranged from 34 to 50 years.

All the retroperitoneal hemangiopericytomases in our series were found to have a clear margin without calcification in CT scan. Three of them (Case 1, 2 & 4) had CT characteristic of soft tissue density with multiple hypodense areas. After the administration of contrast medium the masses shown heterogeneous enhancement with multiple low attenuations. Areas of poor enhancement with greater conspicuity in case 1 indicated necrosis and hemorrhage, and malignancy were proven pathologically [8].

The tumor of case 3 was shown to be relatively well enhanced without obvious low attenuated area in the enhanced CT scans, because it was found to have abundant vascular structures, lack of hemorrhage and less necrotic content in histopathology. Calcification occurs in less than 1% of reported in hemangiopericytomas [9]. Literature review did not reveal that there is an increased incidence of calcification in malignant hemangiopericytomas [8]. Retroperitoneal soft-tissue mass of mesenchymal origin, such as liposarcoma, malignant fibrous histiocytoma and leiomyosarcoma should be considered in the list of differential diagnosis. Liposacroma is generally hypovascular and usually not well enhanced on post-contrast CT scan. Occasionally fat content can be detected by CT scan. Malignant fibrous histiocytomas typically appear as lobulated masses with ill-defined margin and inhomogeneous contents on post-contrast CT scan. These features were absent in our cases. Amongst the CT images of liposarcoma, malignant fibrous histiocytomas, leiomyosarcoma and hemangiopericytoma, hemangiopericytoma usually shows the greatest degree of contrast enhancement [10].

The treatment of choice for hemangiopericytoma is extensive surgical excision. Adjuvant postoperative radiation therapy can improve local control rates in patients initially treated with surgery, but complete remission of hemangiopericytoma from radiation alone is rare [11]. Chemotherapy has not been proven useful for the management of hemangiopericytoma [3,12]. Ten-year survival rate of resectable malignant hemangiopericytoma is only 29% [2]. Local recurrence with metastases occurs in more than 50% of the reported cases [11]. Regular CT scan follow-up is necessary because of the high recurrent rate and possible metastasis, even after complete excision of the tumor initially. All of our patients were followed up by CT scan annually and there was no recurrence for 5 years or longer. In our experience, we found that even with tumor as long as 30 cm in diameter, if there is no adjacent viscera, vessels or lymph nodes involvement, the patients may have a favorable prognosis of 5-year-survival after a successful complete excision.

In fact, a definitive diagnosis of hemangiopericytoma was made only upon histopathology after surgical exploration. The presentation of a large, well circumscribed, lobulated soft tissue mass with low attenuated necrotic zones, and enhancement of solid areas in CT scan study may suggestive of, although not specific for, malignant hemangiopericytoma.

In conclusion, hemangiopericytoma is prominently enhanced in post-contrast CT scan due to its hypervascularity, particularly in the periphery of the lesion, which is helpful for the diagnosis. Heterogeneous enhancement with necrotic and hemorrhagic areas indicates malignancy. CT scan could be a good modality in demonstrating the tumor size, location and the relationship of hemangiopericytoma to the adjacent viscera, treatment planning and useful in follow-up examinations for early detection of recurrent disease.

**REFERENCES**

1. Stout AP, Murray MR. Hemangiopericytoma: vascular
後腹腔血管外皮細胞瘤之電腦斷層評估

陳肇長

張潤忠

高常發

張良慈

吳冠群

呂嘉偉

黃浩輝

吳樹銳

長庚紀念醫院 內科醫學中心及基隆分院 放射診斷科

本院共發現了四位患有後腹腔血管外皮細胞瘤的病人，其中三位女性及一位男性，年齡在34至50歲。我們回顧及分析這四位病人的臨床症狀、電腦斷層影像、病理診斷、治療方法及其預後追蹤等。四例中，有兩例來自腹部後腹腔，兩例來自骨盆後腹腔。發覺這些腫瘤有整齊的邊緣，體積巨大（其中三例的最大長度超過14公分）而導致排擠但沒有侵犯其週邊的器官，沒有腹腔骨盆腔內器官及淋巴結的轉移；兩例腹部後腹腔及一例骨盆後腹腔血管外皮細胞瘤在注射造影劑的電腦斷層影像中，有不均勻的顯影及壞死組織，其餘一例骨盆後腹腔血管外皮細胞瘤則有強烈的顯影而沒有明顯的壞死組織。四例腫瘤在電腦斷層影像中均沒有鈣化。四位病人都接受外科手術切除腫瘤，經病理檢查確定為惡性血管外皮細胞瘤。其後經過五年的手術後追蹤，均沒有發現復發跡象。

後腹腔血管外皮細胞瘤是一罕見的疾病，在電腦斷層掃描檢查中若發現有一邊緣完整，注射造影劑後有良好顯影及壞死組織的腫瘤時，血管外皮細胞瘤應列入鑑別診斷，此外，電腦斷層掃描也是決定治療計劃及預後追蹤的重要工具。

關鍵詞：後腹腔，血管外皮細胞瘤，電腦斷層掃描