

## Case Report

# Sporadic hemangioblastoma of the retroperitoneum

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**Abstract:** Hemangioblastoma is a rare neoplasm occurred in soft tissue. Herein, we present an intriguing case of sporadic hemangioblastoma in the retroperitoneum occurring in a 59-year-old male. The tumor was characterized by an alternation of cellular and paucicellular areas surrounded by a thick fibrous capsule. The stromal cells showed a pale cytoplasm exhibiting occasional lipid droplets and hyperchromatic, irregularly nuclei. CD56, NSE, a-inhibin and S100 were positive in tumor cells. This is the third reported case of a hemangioblastoma in the retroperitoneum. Based on clinical examination, computed tomography imaging, surgical operation, and histopathologic studies, a definitive diagnosis was made. Further characteristic images and pathology of this disease are discussed.

**Keywords:** Hemangioblastoma, soft tissue, retroperitoneum, immunohistochemistry

### Introduction

Hemangioblastoma (HB) occurred in soft tissue is so rare. To date, less than 10 such cases have been reported [1-6], and among them only 2 cases of sporadic HB arising in the retroperitoneum [2, 8]. Here we report the case of sporadic hemangioblastoma of the retroperitoneum (HBR), and its clinical presenting features, radiologic findings, histopathologic findings and immunohistochemical stainings are described, and their differential considerations were also discussed in detail.

### Case report

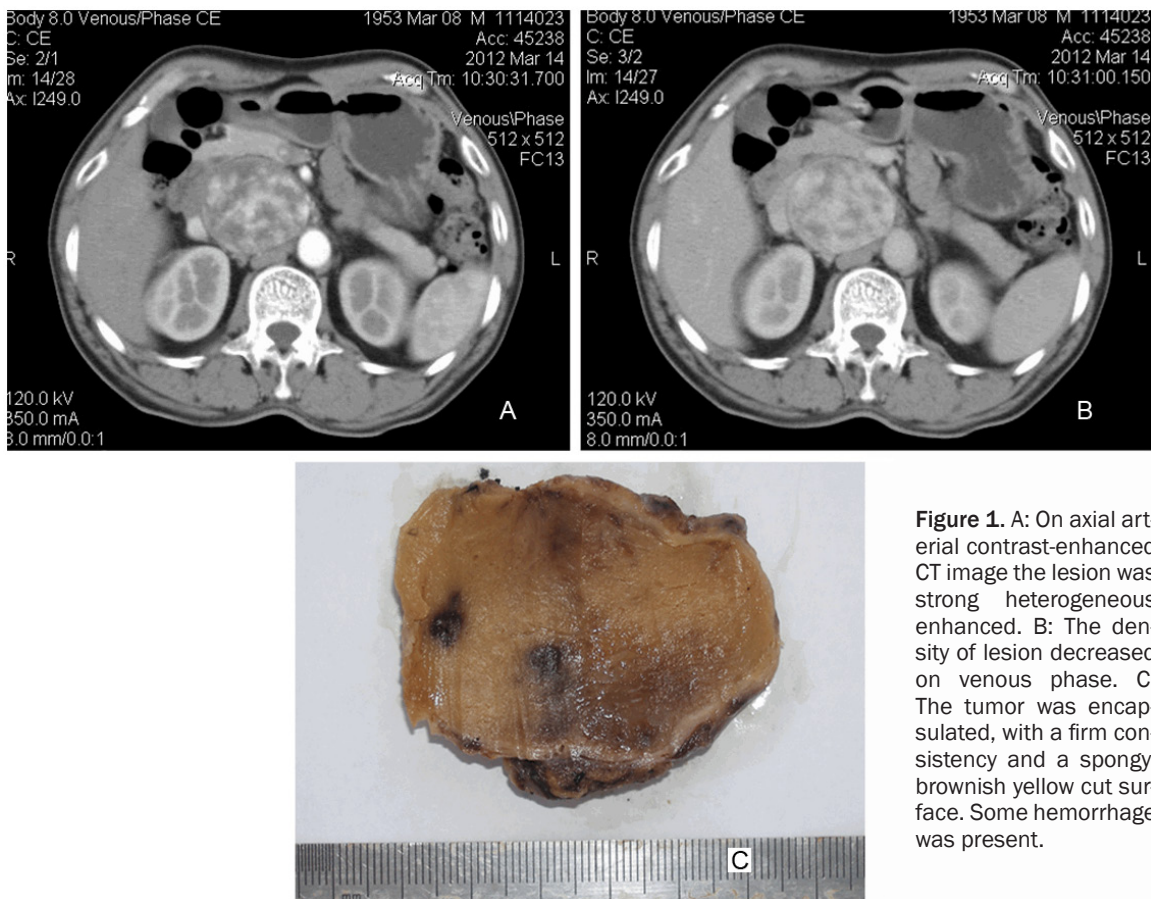
An otherwise healthy 59-year-old man was referred to our hospital for clinical evaluation, and he was found to have a solid, slight low density and oval retroperitoneal mass behind the head of pancreas, left rear of the duodenum, measuring 6 cm in diameter. Axial unenhanced CT image showed a well-defined heterogeneous mass occupying nearly the abdominal aorta and displacing the pancreas with patchy low density areas. On axial arterial contrast-enhanced CT image, the lesion was strong heterogeneous enhanced (**Figure 1A**), and the density of lesion decreased on venous

phase (**Figure 1B**). The density of lesion also decreased on excretory phase. The irregular multifocal non-enhanced areas in the lesion suggested cystic degeneration and necrosis. The case we presented here was initially diagnosed extragastrintestinal stromal tumors or solitary fibrous tumor by radiologists. No remarkable symptoms were reported by the patient. There was no clinical evidence of von Hippel-Lindau disease, and no other tumors were detected. During the operation, a 6 cm × 6 cm × 4 cm mass was located in the upper site behind the pancreas, well-encapsulated and brownish, which was firm and nodular and was clear distinct from peripheral tissues.

### Pathological findings

Gross examination of the resection specimen revealed a well circumscribed, encapsulated, 6-cm mass with a firm consistency and a spongy, brownish yellow cut surface. Some hemorrhage was presented (**Figure 1C**). The mass was completely surrounded by a thin fibrous pseudocapsule. Microscopically, the tumor was well circumscribed, solid, without any cystic component, which was characterized by an alternation of cellular and paucicellular areas surrounded by a thick fibrous capsule

## Sporadic hemangioblastoma of the retroperitoneum



**Figure 1.** A: On axial arterial contrast-enhanced CT image the lesion was strong heterogeneous enhanced. B: The density of lesion decreased on venous phase. C: The tumor was encapsulated, with a firm consistency and a spongy, brownish yellow cut surface. Some hemorrhage was present.

(**Figure 2A**). Nerve bundles were found within the capsule. The cellular areas were composed of arborizing capillary-sized vessels and also thick blood vessels enclosing stromal cells (**Figure 2B**). The vessels were thin-walled varied with thick-walled along with intervening epithelioid stromal cells. The stromal cells showed a pale cytoplasm exhibiting occasional lipid droplets and hyperchromatic, irregularly nuclei. Some stromal cells nuclei were pleomorphic and bizarre (**Figure 2C**). No mitotic rate or necrosis was identified. The paucicellular areas were mainly composed of edematous and hyalined fibrous stroma traversed by a vascular network, and a few stromal cells were intermingled with the vessels (**Figure 2D**). At low-power magnification, hemangiopericytoma-like arrangement was identified. Scattered mast cells were identified in the tumor. No extramedullary hematopoiesis was found. Immunohistochemical analysis revealed that the tumor stromal cells labeled for inhibin-a (**Figure 3A**), neuron-specific enolase (**Figure 3B**), CD56 (**Figure 3C**), S-100 (**Figure 3D**) and Vimentin. But they were

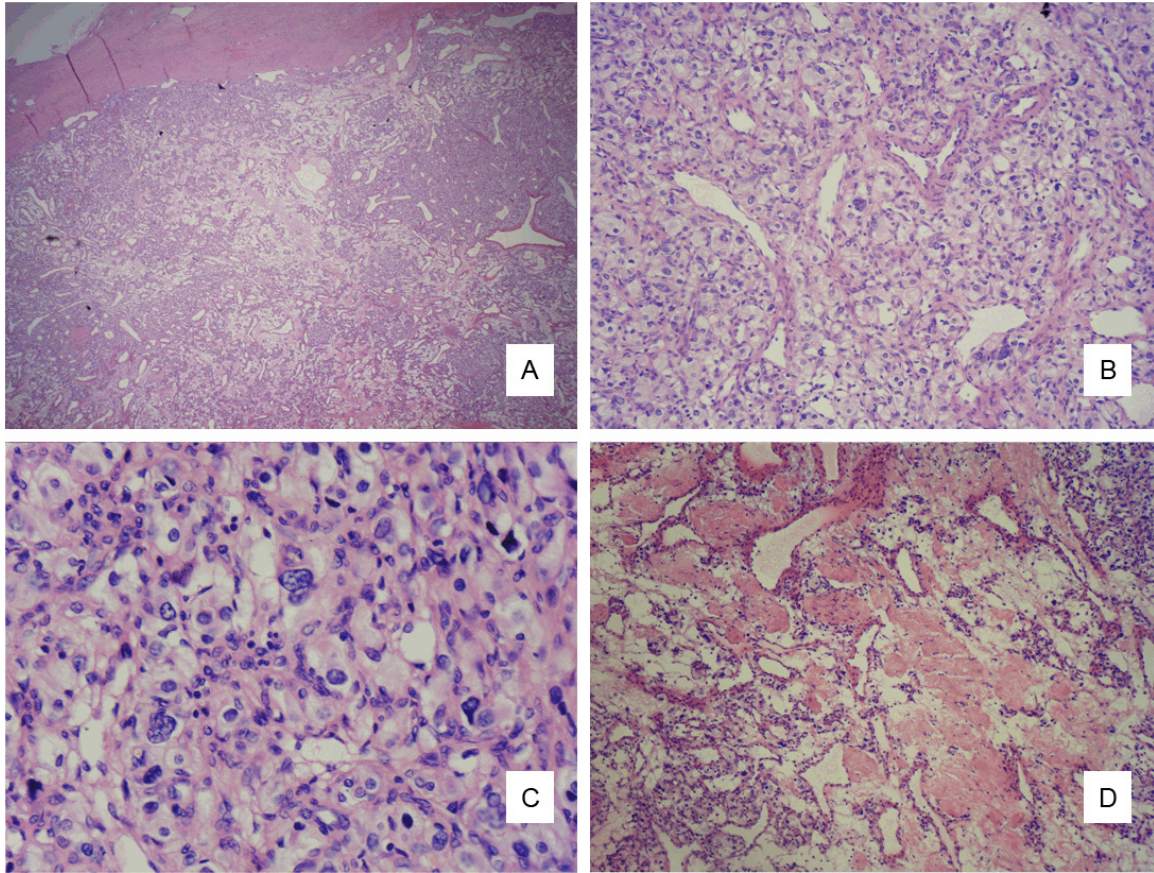
negative for D2-40, AE1/AE3, EMA, SMA and HMB45. CD34 immunostaining highlighted the arborizing and complex vascular network, whereas the tumor cells were negative. Ki-67 index was less than 1% of the tumor cells.

### Discussion

In addition to the age of the patient, the most important radiologic features in evaluation of soft tissue tumors are location and intrinsic imaging characteristics which include size, morphology, shape and extent. Location is one of the most important clues to diagnosis. Neoplasms arising in the retroperitoneum predominately come from soft tissues, vessels, nerves, lymph node and some are derived from sequestered embryonic tissue.

HBR is extremely uncommon since only two cases were definitely reported previously (**Table 1**). The characteristics of HBR included circumscribed borders, often solid, lack of mitotic figures, fine vacuoles and intracytoplasmic lipids,

## Sporadic hemangioblastoma of the retroperitoneum



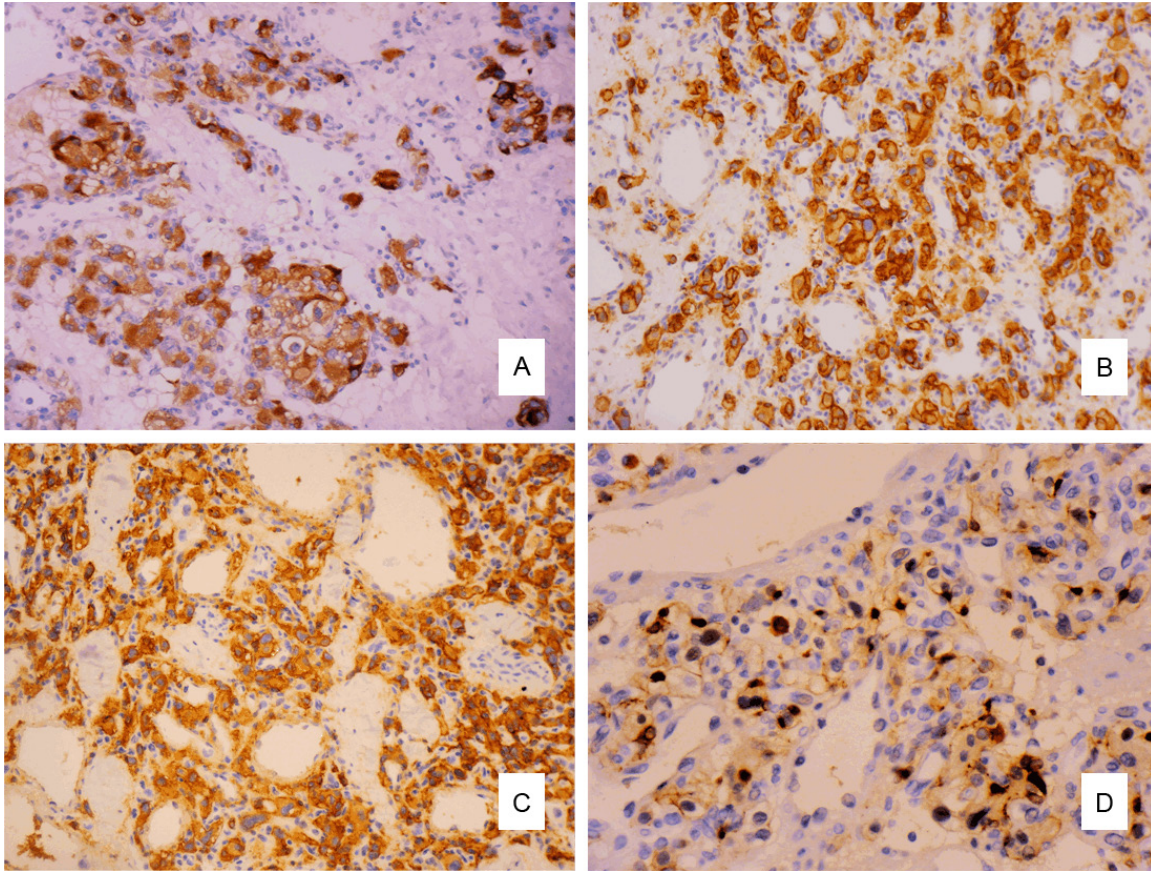
**Figure 2.** A: The tumor was characterized by an alternation of cellular and paucicellular areas surrounded by a thick fibrous capsule. B: The cellular areas were composed of arborizing capillary-sized vessels and also thick blood vessels enclosing stromal cells. C: Some stromal cells nuclei were pleomorphic and bizarre. D: The paucicellular areas were mainly composed of edematous and hyaline fibrous stroma traversed by a vascular network, and a few stromal cells were intermingled with the vessels.

alternation of cellular and paucicellular areas, nuclei pleomorphism and arborizing capillary-sized vessels. The immunoprofiles (S100+, NSE+, inhibin-a+) also conformed to those of hemangioblastomas.

Hemangioblastomas are well-demarcated, highly vascular tumors with varying proportions of capillary proliferation (predominant in reticular variant), fibrosis, and epithelioid clear to foamy stromal cells (predominant in cellular variant). The diagnosis of HBR might be difficult, which should be differentiated with some entities occurred in the specific location, such as paragangliomas, solitary fibrous tumors, metastatic renal cell carcinomas, adrenal cortical carcinoma, and even extragastrointestinal stromal tumors. Because of the distinct components, such as prominent vessels and peripheral vacuolated cells around the vessels, HB

must be differentiated with primary perivascular epithelioid cell tumor, renal cell carcinoma, paraganglioma, hemangiopericytoma, capillary hemangioma, and so on. Adrenal cortical carcinoma was positive for neuron-specific enolase and inhibin-a, but negative for S-100. What's more, the patient's adrenal gland was not involved and no mass was found in adrenal glands. Hemangioblastomas have foamy cytoplasm and irregularly nuclei with degenerative-type atypia, whereas metastatic renal cell carcinomas often have large vesicular nuclei with prominent nucleoli, along with other high-grade features, such as necrosis and mitotic activity, and are typically positive for cytokeratins, EMA, CD10, and RCC, whereas hemangioblastomas are positive for inhibin-a, neuron-specific enolase, and S-100 protein. Primary perivascular epithelioid cell tumor was positive for Melan A, HMB45 and smooth muscle actin, but negative

## Sporadic hemangioblastoma of the retroperitoneum



**Figure 3.** A: Positive staining for  $\alpha$ -inhibin. B: Positive staining for CD56. C: Positive staining for neuron-specific enolase. D: Positive staining for S100 protein.

**Table 1.** Clinicopathological characteristics of the reported sporadic hemangioblastomas in the retroperitoneum

Case	Age (y)/ Sex	Gross features	VHL	Histological features	Immunohistochemical staining (positive)	Follow-up	References
1	47/M	solid and cystic component	-	spindled to plump stromal cells, admixed with epithelioid, lipid-filled multi-vacuolated cells; nuclear pleomorphism; lack of mitoses	vimentin, calponin, Leu7, S100, NSE	NED/NA	[2]
2	71/F	solid, without a large cystic component	-	cytoplasm amphophilic to clear; round with delicate chromatin; lack of mitoses	inhibin-a-(focal), S100, Vimentin, CD57, CA IX, NSE; brachyury (weak)	NED/4 y	[5]
3	59/M	solid, without a large cystic component	-	pale cytoplasm exhibiting occasional lipid droplets; irregularly nuclei; delicate chromatin; nuclear pleomorphism; lack of mitoses	inhibin-a, S100, NSE, CD56, Vimentin	NED/2 y	present

F, female; M, male; NA, not available; NED, no evidence of disease; VHL, von Hippel-Lindau disease; y, years.

for  $\alpha$ -inhibin, S-100, neuron-specific enolase and CD56.

The histogenesis of HB is debatable. The suggested origin of tumor stromal cells includes glial, endothelial, arachnoid, neuroendocrine, fibrohistiocytic, and neuroectodermal cells [7]. Recent studies have also postulated that an

embryonic progenitor cell with hemangioblastic differentiation may be a cytologic equivalent of tumor stromal cells [7].

Reviewing the literature, compared with cerebellar HB, soft tissue HB shares with some common features, such as arborizing capillary-sized vessels and stromal cells, but soft tissue

## Sporadic hemangioblastoma of the retroperitoneum

HB manifest with some different features, such as usually presenting as a solid mass. The pleomorphic and bizarre nuclei frequently present in soft tissue HB and also renal counterpart tumors, are usually not seen in cerebellar HB. Stromal hyalinization in soft tissue HB is also found in renal HB, which is also more prominent than that in cerebellar HB.

In conclusion, we report a sporadic HBR not associated with VHL disease. This entity should be considered in the differential diagnosis of many soft tissue tumors which had the features of vacuolated epithelioid cells intermingled with prominent vascular components. Attention to the distinct histological features of the tumor, such as vacuolated lipid-like cells, varying proportions of capillary proliferation, epithelioid clear to foamy stromal cells, together with the stromal cells coexpression of neuron-specific enolase, S-100 and inhibin-a could make the proper diagnosis.

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### Disclosure of conflict of interest

None to declare.

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