

## CASE REPORT

# Sacro-anterior haemangiopericytoma: a case report

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### ABSTRACT

Haemangiopericytoma (HPC) is a rare vascular tumor with borderline malignancy, considerable histological variability, and unpredictable clinical and biological behavior. HPC can present a diagnostic challenge because of its indeterminate clinical, radiological, and pathological features. HPC generally presents in adulthood and is equally frequent in both sexes. HPC can arise in any site in the body as a slowly growing and painless mass. The precise cell type origin of HPC is uncertain. One third of HPCs occur in the head and neck areas. Exceptional cases of hemangioblastoma arising outside the head and neck areas have been reported, but little is known about their clinicopathologic and immunohistochemical features. This study reports on a case of a large sacro-anterior HPC in a 65-year-old male.

### KEYWORDS

Sacro-anterior; haemangiopericytoma (HPC); soft tissue tumor

## Introduction

Haemangiopericytoma (HPC) is an extraordinarily rare soft tissue tumor. HPC originates from the pericytes of blood vessels, which is a specific cell type that surrounds the capillary vessels. Therefore, HPC can occur in areas where capillaries are found. HPC was first named and described in 1942 by Stout and Murray<sup>1,2</sup>. The most frequent locations of HPC are the extremities, pelvis, retroperitoneum, head, neck, and meninges. HPCs are usually deeply located within the muscle tissue, but dermal and subcutaneous masses have also been described<sup>3</sup>. According to the literature, approximately one-third of all HPCs occur in the head and neck<sup>4</sup>. No HPC cases were found in the sacro-anterior. The pathogenesis of HPC is unclear and its clinical features are non-specific and vary according to the location of the tumor<sup>5</sup>. The incidence of HPC is highest among individuals aged 50 to 60 years old. The diagnosis of HPC depends on histopathologic and immunohistochemistry

(IHC) findings. HPC can be classified as benign, borderline, and malignant depending on its histopathologic and clinical features<sup>6,7</sup>. Not all HPCs are typical; some cases of HPC present overlapping features with other vascular and mesenchymal tumors. IHC helps in excluding other differential diagnoses, but some cases with unusual findings can pose a challenge to general pathologists<sup>3</sup>.

## Case report

A 65-year-old male underwent a computerized tomography (CT) scan because of abdominal pain. The non-contrast-enhanced CT scan revealed a space-occupying lesion approximately 10.7 cm × 8.1 cm in the sacro-anterior. The tumor was asymmetrical and its boundary was clear. The surrounding fat gap of the tumor was clear and no enlarged lymph gland was found (**Figure 1**). However, the malignancy of the tumor remained undetermined. One week later, a radical surgical excision was attempted in a different hospital. The post-operative pathology revealed that the tumor was a sacro-anterior hemangiopericytoma. Immunohistochemical staining of the tumor revealed CD34 (+++), S-100 (-), SMA (-), EMA (-), CD117 (-), Bcl-2 (+++), CD99 (++), NF-(L+H) (-), MyoD1 (++), and Ki-67 (40%+) (**Figure 2**).

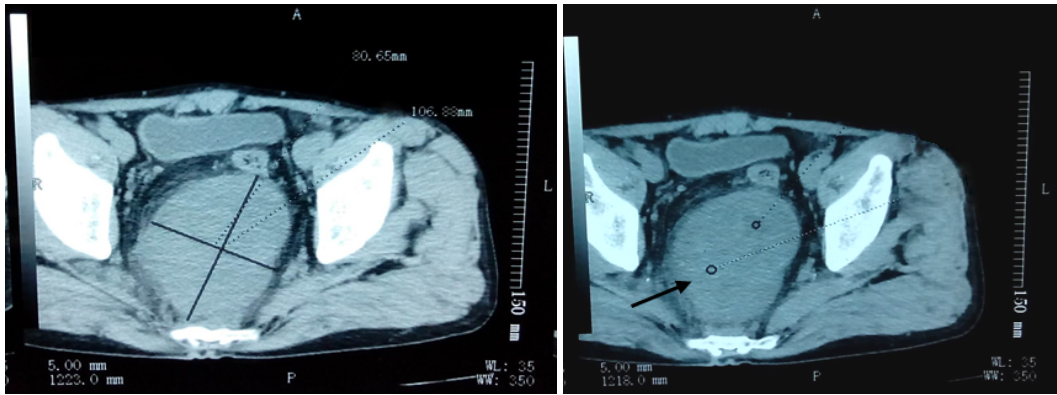
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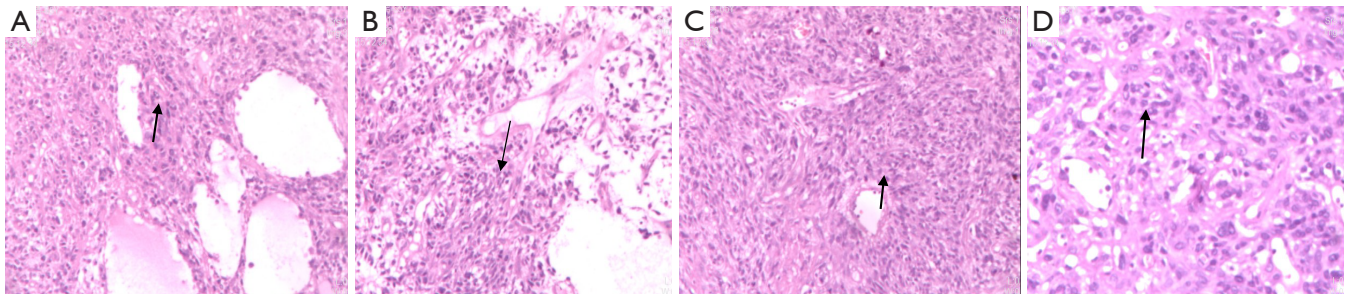
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**Figure 1** Abdominal CT: two pre-operation images of the abdomen. From the images, we can observe a large quasi-circular tumor, which is about 10.7 cm × 8.1 cm in the sacro-anterior. The tumor is intensified asymmetrically and its boundary is clear. The surrounding fat gap of the tumor is clear and no lymph node was found. CT, computerized tomography.



**Figure 2** These four images illustrate the pathology of the sacro-anterior tumor. (A-C) Cancer cells involves spindle cell or oval cell and there are pericyte in perivascular. Cancer cells growth-arranged by radiated, concentric or irregular (H&E staining ×10); (D) there are deckzells which are rounded, ovate or fusiform in perivascular, there are mitotic index in some nuclear (H&E staining ×40).

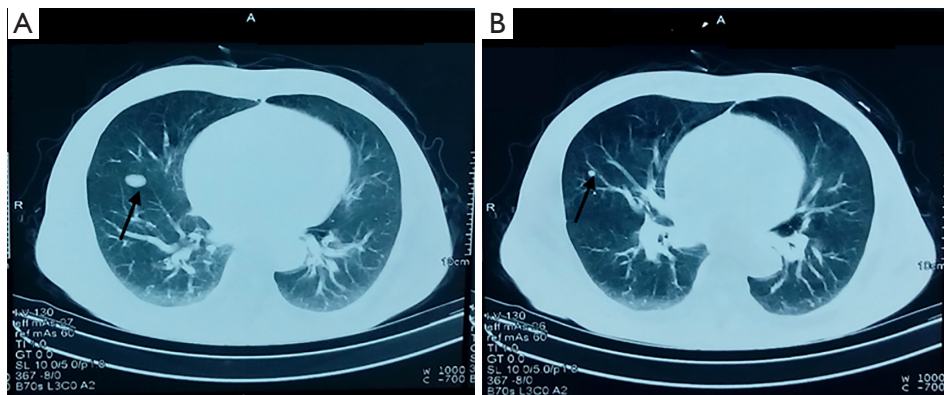
After the tumor was excised, the patient reported no pain or discomfort. Hence, he refused chemoradiotherapy. Ten months later, the patient felt abdominal pain. The patient then went to the hospital and underwent color Doppler ultrasound of the pelvic cavity. The ultrasound results revealed that the HPC recurred, resulting in the patient receiving radical surgical excision again. The pathology and IHC certified that the excised tumor was a recurred HPC. However, the patient still refused chemoradiotherapy. One month later, the patient suffered from frequent coughing and then underwent a chest CT scan. The chest CT scan revealed that the tumor had metastasized into the right lung. The larger metastatic tumor is located in the middle and lower lobe of the anterior basal segment. The tumor was approximately 1.8 cm × 1.1 cm and had a smooth morphology. The smaller tumor was approximately 0.5 cm × 0.6 cm (**Figure 3**). The patient accepted five cycles of EP (VP-16 + DDP) chemotherapy and metastases radiotherapy. After chemoradiotherapy, the patient's condition improved. Six months later, the patient had no complaints and the CT scan showed no

evidence of recurrence (**Figure 4**). The study was approved by the institutional ethics committee.

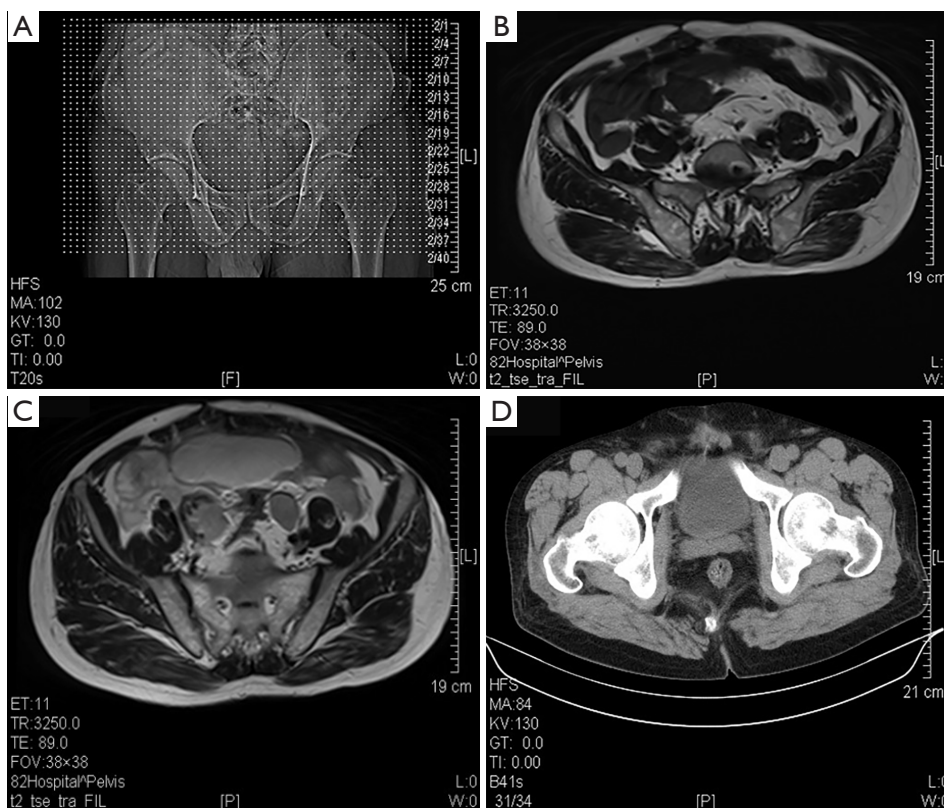
## Discussion

While the classification scheme of the World Health Organization (WHO) for CNS neoplasms considers SFT and HPC as distinct entities with different prognostic implications<sup>8</sup>, SFT and HPC are considered variants of a single entity, with the term “HPC” considered as obsolete<sup>8</sup>. Recent detection of NAB2-STAT6 fusions in both SFT and HPC has provided further support to the notion of a single pathologic entity<sup>9,10</sup>.

HPC is a rare vascular tumor that originates from the pericytes of blood vessels that are also called Zimmurman cells<sup>11</sup>. The Zimmurman cell is a discontinuous spindle cell, which adheres to the reticular fiber membrane. The Zimmurman cell is a juvenile mesenchymal cell that facilitates contraction and potential secretion and can transfer to HPC under pathologic



**Figure 3** Two chest CT images show HPC metastasis in the right lung. The larger metastatic tumor is located in the middle and lower lobe of the anterior basal segment. The tumor is approximately 1.8 cm × 1.1 cm with smooth morphology (A). The smaller tumor is about 0.5 cm × 0.6 cm (B). CT, computerized tomography; HPC, haemangiopericytoma.



**Figure 4** Three abdominal CT images taken 6 months after the last cycle of EP chemotherapy showed no signs of HPC recurrence. CT, computerized tomography; HPC, haemangiopericytoma.

conditions<sup>12</sup>. The peak prevalence age of HPC is 50 to 60 years old, primarily in adult life and at the median age of 55 years regardless of gender<sup>13-15</sup>. HPC has a propensity for recurrence and metastasis and is frequently aggressive. HPC can occur anywhere in the body. About one-third of all HPCs occur

in the head and neck, while others in extremities, pelvis, retroperitoneum, and meninges. Primary sacro-anterior HPCs are very rare tumors and only a few cases have been reported. A painless mass is the most common presentation of head and neck HPCs<sup>7</sup>. The clinical behavior of HPC varies depending on

the tumor grade. A definitive diagnosis of HPC is provided by accurate histopathologic assessment and cannot be made on the basis of clinical and imaging findings [e.g., CT and magnetic resonance imaging (MRI)]<sup>16</sup>. Imaging examinations can only judge the size and boundary of the tumor. Previous studies reported that HPCs observed by CT generally appear as well-circumscribed masses, which are dramatically enhanced after injection of contrast material; meanwhile in MRI images, HPCs are highly intense in T1 and slightly intense in T2<sup>3</sup>. The general morphology of the tumor is round to oval-shaped, and the size of the tumor ranges from 5 to 15 cm<sup>17</sup>, with or without diolame<sup>18</sup>.

HPC has no pathognomonic clinical or radiological signs. Therefore, diagnosis can only be made after careful histopathological examination. The diagnosis of HPC is often made after excluding other tumors that demonstrate HPC-like features. The histologic characteristics of HPC are the tumors that have rich capillary networks and closely arranged tumor cells in the peripheral blood vessels. Tumor cells are intensive skin cells that may be round, oval, or fusiform with unclear boundaries<sup>5,6</sup>. The relative characteristics of HPC immunohistochemically are CD34, Vimentin, CD99, and BCL-2 positive and negative Actin and Desmin<sup>19</sup>. The prediction of the clinical behavior of HPC does not always correlate with the histopathologic features of the tumor. The strict universal histopathologic criteria for malignancy have not been made. The existing malignancy criteria for HPC vary between different studies. The most common metastases sites of HPC are the lungs, mediastinum, liver, and bone. Unfortunately, the aggressive behavior of HPC is unpredictable because benign tumors can become malignant and metastasize<sup>20</sup>. Generally, the large size (>5 cm), increased mitotic rate (>4 mitosis/10 H PF), presence of atypical mitosis, high cellularity, pleomorphic tumor cells, and foci of hemorrhage and necrosis predict a highly malignant course<sup>7,13-15</sup>. A tumor that has a size larger than 2.0 cm and located in a deep site (in the absence of nuclear atypia) demonstrates uncertain malignant potential<sup>20</sup>.

The histopathology of HPCs has been well-documented, but the fine-needle aspiration biopsy (FNAB) results are rarely described<sup>21-24</sup>. FNAB is difficult to use in the diagnosis of primary HPC. However, cytologic analysis may diagnose the recurrence or metastasis of HPC<sup>23</sup>. Incisional biopsy for tumors is not advised because the tumor is considered to be vascular, and the risk of bleeding and consequent fibrosis is present. Furthermore, FNAB is not used in HPC because the primary treatment is always surgery regardless of the pre-operative cytologic findings.

Given the unexpected and potentially aggressive behavior of HPCs, complete excision is the recommended treatment. The common problem with HPC is local recurrence after incomplete

excision, with rates varying from 13% to 40%. In cases of local recurrence, adjuvant radiotherapy and chemotherapy can be offered.

Although the results of adjuvant radiotherapy and chemotherapy vary, adjuvant radiotherapy and chemotherapy are still used as supplementary treatment for HPCs. HPCs are not sensitive to radiotherapy. Radiotherapy is reserved only as adjuvant therapy for cases of incompletely excised tumors, recurrent tumors, and tumors with high-grade histopathologic features. Chemotherapy may have a role in the treatment of distant metastatic disease, but its role in the primary treatment of HPC remains unclear. Vincristine, Adriamycin, and Cyclophosphamide have been used in cases of aggressive HPCs with variable success<sup>25</sup>. Some reports showed that the use of interferon may benefit patients with pulmonary HPC metastases<sup>26</sup>. Some studies reported local recurrence rates of as high as 40%, with metastatic disease in 15% of the cases reported with a latency period ranging from 63 to 107 months<sup>27</sup>. In our case, after operation, radiotherapy, and chemotherapy, the patient's clinical symptoms nearly completely disappeared, and the CT scan (**Figure 4**) revealed that the tumor did not recur.

## Conclusion

HPC is a rare soft tissue tumor with high histological variability and unpredictable clinical and biological behavior. One third of HPCs occur in the head and neck and only a few cases have been reported regarding tumor localization in the sacro-anterior. Pre-operative evaluation must include a thorough imaging evaluation (CT and MRI scan), including the results that may not be specific for HPCs. The treatment of choice is radical excision. HPCs are potentially malignant, regardless of the benign pathology. Given the possibility of recurrence and metastasis in HPC cases, surgical excision may not be sufficient<sup>28</sup>. After tumor excision, adjuvant chemotherapy is necessary. During follow-up visits, imaging examination is used to determine recurrence and metastasis.

## Conflict of interest statement

No potential conflicts of interest are disclosed.

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