

Case Report

## Behind the Peritoneum and Back of the Mind: Consideration of Retroperitoneal Sarcomas as a Differential Diagnosis

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Article Info

Article history:

Received: 29 April 2022

Accepted: 12 August 2022

Published: 17 September 2022

Academic Editor:

Sharifah Najwa Syed Mohamad

Malaysian Journal of Science,  
Health & Technology

MJoSHT2022, Volume 8, Special Issue  
eISSN: 2601-0003

<https://doi.org/10.33102/2022284>

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**Abstract** — Retroperitoneal sarcomas (RPS) are rare malignant tumours that originate from connective tissues of skeletal or extraskeletal origin in the retroperitoneal region, which are often missed. This case report highlights a patient with RPS that presented with abdominal swelling. In addition, this case report was written to elucidate the significance of thorough history, physical examination findings, and awareness in considering RPS as a differential diagnosis of abdominal masses in accurately diagnosing RPS for timely intervention.

**Keywords** — Retroperitoneal Sarcoma; Soft Tissue Sarcoma; Retroperitoneal Mass; Abdominal Swelling; Abdominal Mass

### I. INTRODUCTION

Sarcomas are a group of malignancies that arise from the mesenchymal cell. Most sarcomas are soft tissue sarcomas, with a small percentage occurring in the retroperitoneum [1]. Retroperitoneal sarcomas (RPS) may not always be considered as a differential diagnosis when a patient presents with a painless abdominal mass. Often, these masses are thought to be gynaecological or renal in origin [3]. This can be attributed to the rarity of soft tissue sarcomas that occur in less than 1% of all cancers in adults [1],[2]. Failure to consider RPS may result in excessive, incomplete, or delayed workup leading to inaccurate diagnoses, poor prognosis, or non-beneficial interventions. Therefore, it is paramount to recognize and identify the possible presentations and clinical features of patients with RPS.

### II. CASE REPORT

A 53-year-old male with no previous known medical illness was admitted to a tertiary care hospital under the surgical ward on 20 March 2022 to 31 March 2022. The patient presented with one month of progressively worsening right abdominal swelling associated with loss of weight, pedal edema, and lethargy. Nevertheless, the patient did not complain of abdominal pain, loss of appetite, fever, bowel alterations, nausea or vomiting, jaundice, urinary symptoms such as hematuria, changes in stool and urine colour, or any shortness of breath. The patient was a chronic smoker but had no history of alcohol use or recent travel.

On physical examination, a 20 x 10cm mass was palpated in the right abdomen. The mass extended 1cm from the right costal margin vertically and 1cm right lateral to the midline of the abdomen horizontally. The mass was round, smooth, hard,

and immobile. It was not ballotable, did not move with respiration, and can be overcome. Apart from that, there was no hepatosplenomegaly or ascites noted.

Interestingly, the initial differential diagnoses considered by the treating team were sigmoid colon carcinoma and renal cell carcinoma. As such, an abdominal ultrasound was requested. Two large right abdominal masses with a poor plane to the right kidney were noted, likely to be exophytic renal cell carcinoma or retroperitoneal sarcoma. There were no hepatomegaly or dilated biliary ducts noted in the ultrasound report. Subsequently, the treating team ordered a renal mass contrast-enhanced computed tomography (CECT) protocol and CECT thorax, which noted moderate ascites, pericardial effusion, and multiple right perirenal or retroperitoneal mass, likely to be retroperitoneal sarcoma, plasma cell neoplasm, or lymphoma.

From the differential diagnoses suggested by the CECT results, multiple interdisciplinary consults involving the surgical, urological, and radiological departments were made. A urological referral was made inpatient to assess the likelihood of renal cell carcinoma.

Investigations ordered include a full blood count, renal profile, liver function test, calcium, magnesium and phosphate levels, coagulation profile, peripheral blood film (PBF) and C-reactive Protein (CRP). Among which, the patient had low haemoglobin, elevated total white cell count, hypokalemia, raised alkaline phosphatase (ALP), low albumin levels, and elevated CRP. PBF showed hypochromic anaemia, likely iron deficiency anaemia, and no leucoerythroblastic white cells or abnormal lymphoid cells. No tumour markers were ordered.

Upon review by the urology team and a discussion with the radiologist, it was concluded that renal cell carcinoma was unlikely due to the absent enhancement in the mass during arterial phases and washout during corticomedullary phases of the CECT.

However, when reviewed by the surgical team, it was thought that the mass was still likely renal in origin, which resulted in a second referral to the urology team. No active surgical intervention was made at this stage. A right renal and retroperitoneal biopsy was ordered. The patient was discharged on 31 March 2022 and to be followed up outpatient for the review of histopathological examination (HPE) results.

Subsequently, the patient was admitted to another hospital after developing shortness of breath. Unfortunately, the patient passed away on 16 April 2022 due to causes unknown to the authors. As such, the outpatient review was not carried out. Nonetheless, HPE results noted high-grade spindle cell sarcoma.

### III. DISCUSSION

There is a high likelihood of malignancy in a middle-aged male smoker who presents with progressively increasing abdominal mass, weight loss, and lethargy. However, when considering the primary causes of cancer, RPS is often not included by clinicians.

Notably, retroperitoneal pathologies, especially RPS, have non-specific symptoms and clinical signs. Therefore, most patients with RPS remain asymptomatic until the mass is large enough to compress or invade nearby structures in the

peritoneal cavity leading to abdominal pain [4]. As such, other more common differential diagnoses such as renal cell carcinoma, colon cancer, and hepatocellular carcinoma are prioritised.

Nonetheless, relevant negatives may guide us to 'rule out' more common causes and consider rarer causes of abdominal mass, such as RPS. This is depicted in our patient's case as there was an absence of symptoms signalling toward a cardiorespiratory (chest pain, orthopnoea, exertional dyspnoea, cough), hepatobiliary (jaundice, vomiting, dark urine, pale stools, right upper quadrant pain), renal (haematuria, flank pain, changes in urine output), or colonic (altered bowel habits, hematochezia, melaena, tenesmus) malignant pathology. The patient also presented with lower limb oedema, which can be caused by a plethora of retroperitoneal malignancies that have obstructed lymphatic return, such as lymphomas, renal cell carcinoma, or RPS [4].

Features suggestive of other differential diagnoses of renal and hepatic origin were also absent on physical examination in this patient. The patient's flank was not full, and the abdominal mass can be delineated from surrounding organs such as the liver. It was also not ballotable and did not move with respiration.

While history and clinical signs do not explicitly rule out the mass being of renal, colon, or hepatic origin, the lack of significant symptoms in the history and description of the mass on examination in this patient would suggest a lower likelihood of these common pathologies to be the cause of the mass. Retrospectively, the importance of clinical correlation, reasoning, and consideration of rare pathologies like RPS can change the workup of the patient and subsequently diagnose and treat them in a timelier manner.

At this stage, investigations ordered should narrow down the differential list. RPS can only be confirmed with imaging and biopsy. Therefore, the recommended imaging is computed tomography (CT). Although abdominal CTs cannot predict sarcoma cell types [5], it remains an invaluable diagnostic tool despite being unable to pick up RPS in many cases [3]. Magnetic Resonance Imaging (MRI) was found to be similarly efficacious as CT scans, with no significant difference in preoperative assessment. Hence, the cheaper and more readily available CT scan is the preferred modality for evaluating patients with retroperitoneal masses [4].

One of the most prominent benefits of imaging is its non-invasive nature compared to biopsies. While it is recommended to biopsy all soft tissue mass due to difficulties in distinguishing benign and malignant causes based on physical signs [6], a study in Thailand found that preoperative core needle biopsies were not necessary when CT scans were diagnostic [7]. Supporting this is another study in the Netherlands where RPS was diagnosed in cases without non-affirmative biopsies [3]. Thus, the need for biopsies prior to surgery should be evaluated by surgeons. It is crucial to avoid unnecessary invasive procedures for malignancies that are almost guaranteed to require surgical excision. In this patient, the decision to undergo a biopsy was made.

Tumour markers are commonly used to monitor treatment response and disease progression [8]. Not often, tumour markers are used as a diagnostic or screening tool for cancers [8]. We postulate that tumour markers were not ordered in this patient as there are no specific diagnostic biomarkers for renal

cell carcinoma [9]. Furthermore, immunohistochemical markers for primary RPS tumours [10] may not be readily available in government hospitals as it is expensive and rarely used. However, in the context of RPS, common tumour markers may be useful to detect metastatic secondaries from other sites such as, Alpha Fetoprotein (AFP) for germ cell tumours, or Carcinoembryonic Antigen (CEA) for adenocarcinomas [11].

As many organs lie within the abdominal cavity, clinicians should consider that it is not impossible for RPS to invade other structures and mimic other pathologies like renal cell carcinoma [12],[13]. Some signs to identify tumours from retroperitoneal organs or other retroperitoneal structures include the embedded organ sign, phantom organ sign, beak sign, and prominent feeding artery sign [14]. Thus, radiologists must recognize both present and absent signs on imaging if diagnostic biopsies are unavailable or not done.

The radiologist in this case recognized that renal masses do not enhance during arterial phases and washout during corticomedullary phases – ruling out renal cell carcinoma as a differential diagnosis. Hence, multiple referrals to urology may not have been required as it may cause delays in timely surgical interventions. These referrals may have been made due to the lack of awareness of rarer pathologies such as RPS.

#### IV. CONCLUSIONS

In general, retroperitoneal masses are difficult to diagnose due to vague symptoms and clinical signs. While easier to recognize on retrospective analysis, soft tissue sarcomas such as RPS are often missed. As a result, most RPS go undiagnosed until the late stages, where the prognosis is poor. Therefore, awareness of RPS as a differential diagnosis is the first and most critical step to prompt relevant and necessary workup. Clinical reasoning and proper correlation of history and physical signs are also essential in managing patients with vague symptoms [3]. With early identification, referrals to surgeons specialising in treating sarcomas can be made, and further delays can be avoided.

#### CONSENT TO PARTICIPATE

The patient's sibling has provided verbal consent for publishing as the patient has passed away.

#### CONFLICT OF INTERESTS

The authors declare that there is no conflict of interest. There was no specific funding for this study.

#### ACKNOWLEDGEMENT

The authors express their gratitude to the patient and his family for sharing a part of themselves selflessly. The authors would also like to express special thanks to Monash University Malaysia.

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