Spontaneous retroperitoneal hemorrhage: where's the culprit?

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Learning objectives

• To review the most common causes of spontaneous retroperitoneal hemorrhage (non-traumatic/non-iatrogenic), focusing on retroperitoneum fascial anatomy.

• Describe the imaging findings that can provide clues to establish a correct diagnosis and track the source of hemorrhage.

• To point out what are the most important questions that the radiologist is expected to answer, which will determine the therapeutic approach.
Background

ETIOLOGY

Spontaneous retroperitoneal hemorrhage is a rare entity occurring in the absence of a traumatic or iatrogenic event. Excluding aortic ruptures, it is most commonly seen in coagulopathic states relating to vessel abnormalities, but also in vascular rupture of retroperitoneal organ expansive lesions, most often spontaneous rupture of renal carcinomas and angiomyolipomas and adrenal expansive lesions. [1],[2].

CLINICAL PRESENTATION

Clinical presentation can be protean depending on the etiology, the site of bleed and the extent. Symptoms are most commonly nonspecific and difficult to classify in the beginning. Patients may refer generalized abdominal pain or pain located to the epigastric region, the hips or radiated to the lumbar region. In the setting of massive bleeding, signs of hemodynamic instability and a dwindling hematocrit may suggest the diagnosis [4].

RETROPERITONEAL ANATOMY

Knowledge of retroperitoneum spaces and fascial planes is important to locate the site of origin and thus identify a potential culprit.

Traditional tricompartment model of the retroperitoneum (anterior pararenal space (APS), perirenal space (PS), and posterior pararenal space (PPS) circumscribed retroperitoneum processes to a specific compartment.

Recent understanding of the variability of the perirenal fascia fusion, led to a "modified tricompartment model" (Fig. 1), that contemplates interfascial connections between the spaces. The retromesenteric, retrorenal, and lateroconal planes are potential routes of interfascial communication between the retroperitoneal spaces. Perinephric septa run between the renal capsule and the perinephric fascia, allowing subcapsular fluid to communicate with the retrorenal space or retromesenteric plane [9].

Retroperitoneal hemorrhage or rapidly expanding fluid collections can spread via these interfascial connections. Interfascial spread is a method of communication between the
abdomen and pelvis and also allows communication across the midline through the retromesenteric space.

**Figure 1.** Fascial planes and spaces of the retroperitoneum. (a) Traditional tricompartment model of the retroperitoneum, divided into the anterior pararenal space (APS), perirenal space (PRS), and posterior pararenal space (PPS). The anterior renal fascia (ARF), posterior renal fascia (PRF), and lateroconal fascia (LCF) divide the spaces. (b) Modified tricompartment model, which reflects the understanding that the perirenal fascia is variably fused and there are interfascial connections between the spaces. The retromesenteric plane (RMP), retrorenal space (RRS), and lateroconal space are potential interfascial communications. APS = anterior pararenal space, PPS = posterior pararenal space, PS = perirenal space.[9].
Findings and procedure details

For patients with mild unspecific abdominal pain, abdominal x-ray and ultrasonography are often the first imaging modalities performed. They are aimed essentially at excluding gastrointestinal perforation and other common etiologies of acute abdominal pain such as cholecystitis or renal calculi. Ultrasonography evaluation is limited by body habitus (obesity) and interposing abdominal gas, especially in the ER setting where there is no previous patient preparation and patients are often unable to cooperate. Abdominal X-ray can provide a clue when it shows an effacement of the psoas shadow (Fig. 2).

For patients presenting with hemodynamic instability, a drug-resistant pain with consistent clinical findings or a clinical suspect of bleeding, CT is often the first exam in order to quickly establish a diagnosis of a potential life-threatening intra/retroperitoneal condition that may need urgent or emergent intervention. CT plays a primary role because of its ability to identify, with high sensitivity and specificity, the presence and cause of hematomas into the retroperitoneal space, significantly influencing the subsequent management. The goal is to identify the retroperitoneal hemorrhage, its location, and its possible source and to assess its relative stability on the basis of the size and presence (or absence) of active extravasation of intravascular contrast material.

Digital subtraction angiography is recommended in cases where intermittent bleeding is suspected (active bleeding not demonstrated on CT but increase of hematoma in follow up CT).

Conservative management is adopted when there are no signs of active bleeding or only low-flow active bleeding; urgent embolization is indicated in significant arterial bleeding without hemodynamic instability and surgical treatment is reserved for actively bleeding hematoma with hemodynamic instability.

OPTIMAL CT PROTOCOL

A baseline acquisition is useful for depicting the spontaneous hyperattenuating bold clot (Fig. 5 and 8).

CT protocol should be tailored to depict an optimized arterial phase, ideally using a "bolus tracking" technique with high flow injection of contrast medium (4-5ml/sec), so active bleeding won't be missed and to differentiate arterial phase from venous bleeding. Active extravasation is seen in a minority of patients in whom CT reveals a hematoma. Its detection with enhanced single phase CT, is limited when nonspecific areas of hyperattenuation are encountered; active extravasation is better distinguished with at least a dual phase CT protocol [7]. The classic pattern of active extravasation
at dual phase CT is a jet or focal area of hyperattenuation within a hematoma on initial images that fades into an enlarged, enhanced hematoma on delayed images, indicating significant bleeding.

Furthermore, CT can help detection of contained vascular lesions such as aneurysms or pseudo-aneurysms within a hematoma.

**CLUES TO THE SOURCE**

Understanding the spatial distribution of the collection in the retroperitoneal spaces can help orientate a diagnostic algorithm.

<table>
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<th>Table 1</th>
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<td><strong>Anatomic spaces of the retroperitoneum</strong></td>
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<tr>
<td><strong>Anterior</strong> pararenal space</td>
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<tr>
<td><strong>Perirenal</strong> space</td>
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<td><strong>Posterior</strong> pararenal space</td>
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- The "**sentinel clot sign**": it refers to the clot organized in the first hour after bleeding. Clots tend to form first near the site of bleeding; thus, the identification of a heterogeneous and relatively higher attenuation clot allows localization of the site of hemorrhage. Narrowing CT windows may help detect this clot (Fig. 5 and Fig. 8).

- The "**hematocrit sign**": corresponds to the presence of a cellular-fluid level caused by the settling of cellular elements in the dependent portion of a hematoma, and is a highly sensitive (87%) and specific sign of coagulopathic hemorrhage [8].

- The "**signal flare sign**": when active bleeding is present in the context of a hematoma with cellular-fluid level, contrast medium settles as a layer between cellular and fluid component, because of the different gravitational weight.
• **Hemorrhage** is defined as free extravasation of contrast media that persists and enlarges on delayed images and **pseudoaneurysm** as a round or ovoid cavity, communicating with an injured vessel wall, that shows wash out on delayed phase. **Arteriovenous fistula** is defined as early, simultaneous vessel enhancement of both artery and vein.

**SPECIFIC CAUSES**

Main causes can be essentially divided into parenchymal bleeding and vascular bleeding:

**Parenchymal bleeding** is often related to renal or adrenal lesions.

When there is no trauma context, spontaneous renal bleeding is named Wunderlich syndrome. The major culprits are renal cell carcinoma and angiomyolipomas, but other benign neoplasms and cysts (Fig. 6) can also be responsible.

In most cases, CT permits the radiologist to clearly differentiate a mass from the surrounding hematoma.

The diagnosis of an underlying angiomyolipoma is based on the identification of low-attenuation areas of fat in a large heterogeneous mass (Fig. 3 and Fig. 4). On contrast-enhanced CT, the presence of a solid mass with less contrast enhancement than the adjacent renal parenchyma suggests RCC. However, small tumors may initially be obscured by the hematoma; therefore, follow-up imaging after resolution of the initial hematoma is essential.

In adults, spontaneous adrenal hemorrhage is an uncommon condition that is usually bilateral and associated with anticoagulation therapy, severe stress or sepsis. Cases of spontaneous rupture associated with an adrenal masse include pheochromocytoma, myelolipoma, cortical adenoma, adrenocortical carcinoma, and metastases.

CT of adrenal hemorrhage appears as a round solid adrenal mass with attenuation comparable to soft tissue, without appreciable enhancement after the IV administration of contrast material, which decreases in size during follow-up (Fig. 7). MR is very sensitive and specific for diagnosing adrenal hemorrhage and determining if blood is the sole component of the hematoma.

**Vascular bleeding** is related to rupture of splanchnic arteries aneurysms, arteriovenous malformations, or disease resulting from atherosclerotic or inflammatory small vessels injuries being most prevalent in patients with coagulopathic states like anticoagulation therapy (with a reported incidence at 0.6-6.6% ) [1], bleeding abnormalities (such as
factor IX or X deficiency, von Willebrand disease or antiphospholipid syndrome), and chronic haemodialysis. [1],[2]. In this setting, bleeding commonly involves multiple sites, and especially the body wall muscle compartment such as the rectus sheat or the iliopsoas muscle [8]. When contrast enhanced CT detects coagulopathy-associated active extravasation, this is more frequently venous than arterial, usually not requiring surgery or embolization. Treatment is mainly conservative and based on withholding of anticoagulant medications.

More frequently described etiologies are rupture of aneurysms and pseudo-aneurysms of the superior mesenteric artery branches, renal artery aneurysms, pancreatic-duodenal artery aneurysms (often associated with stenosis of the celiac trunk) and vasculitis such as polyarteritis nodosa and Wegener's granulomatosis (notably in the kidney). Lumbar, ileo-lumbar and obturator artery ruptures have also been reported.
Fig. 1: Figure 1. Fascial planes and spaces of the retroperitoneum. (a) Traditional tricompartment model of the retroperitoneum, divided into the anterior pararenal space (APS), perirenal space (PRS), and posterior pararenal space (PPS). The anterior renal fascia (ARF), posterior renal fascia (PRF), and lateroconal fascia (LCF) divide the spaces. (b) Modified tricompartment model, which reflects the understanding that the perirenal fascia is variably fused and there are interfascial connections between the spaces. The retromesenteric plane (RMP), retrorenal space (RRS), and lateroconal space are potential interfascial communications. APS = anterior pararenal space, PPS = posterior pararenal space, PS = perirenal space.[9].

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**Fig. 2**

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**Fig. 3**

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**Fig. 4**

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**Fig. 5**

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**Fig. 6**

57-year-old man with a single kidney, presents with sudden onset acute abdominal pain. Axial unenhanced CT, shows hyperdense collection spreading in the perirenal and posterior pararenal space, caused by rupture of several renal cysts. Note stranding in the lateroconal plane reflecting interfascial connections of the retroperitoneum.

b) MPR reconstruction in unenhanced CT, depicts intracystic hemorrhage in a patient with dominant polycystic disease.

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**Fig. 7**

Axial unenhanced (a) and enhanced portal phase (b) CT in a patient presenting with left iliac fossa pain and nausea. There’s a nodular lesion in the left adrenal area, spontaneously hyperdense in unenhanced acquisition, pointing to an hematic content. In portal phase there is no enhancement, globally suggesting a left adrenal hematoma. Note peri-renal stranding pointing to a process originating in this compartment.

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Fig. 8

Axial unenhanced (a) and enhanced arterial phase (b) CT, shows left common iliac artery aneurysm rupture with adjacent blood clot (arrow) signaling the source of rupture (“blood clot sign”) and local peri-aneurysmatic fluid with no signs of active bleeding (no contrast medium extravasation).

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Conclusion

Spontaneous retroperitoneal bleeding is an entity of difficult clinical diagnosis due to its insidious and fuzzy-symptom presentation. Early diagnosis is crucial to prevent a severe clinical progression and unfavorable outcome.

Multiphasic CT is the gold standard for the identification of a spontaneous retroperitoneal hematoma. Familiarization with retroperitoneum anatomy and knowledge of specific signs in the context of hemorrhage, help establish the diagnosis and identify the source of bleeding.

Recognition of CT signs of active bleeding is the most important element influencing the timing of therapeutic treatment [2]. When active bleeding is present, decision of surgery or urgent embolization depends on whether there is hemodynamic instability or not, respectively. An initial more conservative approach can be adopted in patients without signs of contrast extravasation or low-flow active bleeding.
References


