

Spontaneous Retroperitoneal Hematoma: A Rare Devastating Clinical Entity of a Pleiada of Less Common Origins

Definition of Wunderlich syndrome, also known as spontaneous retroperitoneal hemorrhage (SRH), was first given in 1700 by Bonet and was more completely explained by Wunderlich.^[1] Although SRH is commonly associated with Lenk's triad (acute flank pain, symptoms of internal bleeding, and upper and lower quadrant abdominal tenderness to palpation – costovertebral angle tenderness), the most common signs and symptoms described are abdominal pain (67%), hematuria (40%), and shock (26.5%).^[2,3] It is frequently found in conjunction with hypertension (33–50%) and atherosclerosis (80–87%).

Etiologies as well as the precise mechanisms leading to SRH are unclear in most of the reported cases. Tumors, particularly renal cell carcinoma and angiomyolipoma, are the most common cause of SRH, occurring in 57–73% of cases.^[3] The overall prevalence of SRH as a complication of tumors, however, is low. In renal cell carcinoma, it occurs in only 0.3–1.4% of cases,^[4] although the incidence is much higher in angiomyolipoma, occurring in 13–100% of cases, depending on tumor size. Adrenal myelolipoma, pheochromocytoma, and adrenal hemangiomas have also been reported to cause SRH.

Aneurysms of the visceral circulation as a cause are rare, accounting for 0.1–10.4% in autopsy statistics. The exact mechanism of rupture of branches of splanchnic vessels is unknown, but likely represents weakness of the tunica media, predisposing rupture in the face of abrupt increases in pressure. Pathology specimens regularly exhibit disruption of elastic lamellae. Spontaneous hemorrhage can be seen with inflammatory erosive processes which explain the association with necrotizing arteritis in polyarteritis nodosa and rheumatoid arthritis.

Spontaneous retroperitoneal haematoma among patients receiving anticoagulation therapy has been well described and related to warfarin, low-molecular-weight heparin, unfractionated heparin, or even clopidogrel.

Abdominal compartment syndrome as an acute abdominal emergency after massive retroperitoneal bleeding is also a clinical entity in close correlation

with anticoagulant therapy that we should be aware in such cases.^[5]

A very interesting point in the case that the authors describe is the fact that the young female had undergone an uncomplicated vaginal delivery 10 days prior to presentation with no obstetric concerns. Late postpartum hemorrhage is a rare phenomenon occurring 24 hours to 6 weeks postpartum and may be a cause of a retroperitoneal hematoma.^[6] Infection, subinvolution of the placental bed and retained placental products are the major causes of late postpartum hemorrhage.

Diagnosis via CT scan is the principal method of diagnosis but it is practical only in hemodynamically stable patients. It helps in establishing the site, size, and likely underlying causes.^[7] CT angiography of vessels has proven useful as a screening tool using small amounts of contrast to elucidate sites of active bleeding. Computed tomography can miss segmental arterial mediolysis as a cause of spontaneous retroperitoneal hemorrhage.^[8]

In patients with acute abdominal pain, the evaluation of intraabdominal pathology based on clinical symptoms and signs might be often unreliable. Laparoscopy, as a minimally invasive technique, has unique advantages in such cases and should be regarded as a good diagnostic tool. Percentages of patients who avoided open laparotomy because of this method range from 30% to 65%.

Treatment of spontaneous retroperitoneal bleeding, as with other bleeding phenomena, revolves around resuscitation and restoration of circulating volume – packed red blood cells (PRBCs), fresh frozen plasma, units of platelets, colloids.

This has traditionally been followed by surgical correction in hemodynamically unstable patients.

Radiological intervention with embolization of the feeding vessel is an option in splanchnic aneurysms.

Idiopathic retroperitoneal hemorrhage/hematoma is a relatively uncommon condition with a large variability

and nonspecific presentation in most of the cases, which may lead to shock and even death unless it is promptly recognized and treated appropriately. The thorough presentations from the authors of this rare phenomenon alert physicians to the risk and appearance of this rare grave complication. Prompt diagnosis and treatment may spare the patient surgical intervention and improve outcome.

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