SUMMARY
Spontaneous retroperitoneal hematoma associated with iliac vein rupture is a rare and life-threatening emergency. We report a case of spontaneous left iliac vein rupture in a healthy 49-year-old woman, with no relevant medical past history or history of trauma. On the day she was admitted to the emergency room she had a syncope episode and was hypotensive and tachycardic on arrival. She suffered from back pain, which started 5 days prior to admission. Workup with contrast-enhanced computed tomography revealed a large retroperitoneal hematoma without evidence of arterial bleeding. Due to hemodynamic instability an emergency exploratory laparotomy was performed. Intra-operatively, retroperitoneal exploration, revealed a large hematoma caused by a left primitive iliac vein rupture. The vein was repaired primarily with control of the bleeding. She was taken to the Intensive Care Unit for postoperative follow-up where she subsided to cardiopulmonary arrest on the same day. This case provides insight to an uncommon cause of retroperitoneal hematoma and haemorrhagic shock that should be taken into account on the emergency room.

Key-words: Spontaneous rupture, Iliac vein, retroperitoneal hematoma, hemorrhagic shock.

RESUMO
A ruptura espontânea da veia ilíaca com consequente hematoma retroperitoneal é uma emergência rara e que coloca em risco a vida do doente. Descreve-se o caso clínico de ruptura espontânea da veia ilíaca esquerda numa doente do género feminino, de 49 anos, saudável, sem antecedentes pessoais conhecidos ou história de trauma. Recorreu ao serviço de urgência após um episódio de lipotimia, encontrando-se taquicárdica e hipotensa à admissão. Descrevendo, associadamente, um quadro de dor lombar com 5 dias de evolução. Imagiológicamente apresentava um hematoma retroperitoneal sem evidência de hemorragia arterial ativa. Por instabilidade hemodinâmica foi realizada uma laparotomia exploradora de emergência que revelou um hematoma retroperitoneal de grandes dimensões causado por uma ruptura da veia ilíaca primitiva esquerda. Foi realizada sutura primária da lesão vascular com controlo da hemorragia. No pós-operatório foi admitida na unidade de cuidados intensivos onde sofreu uma paragem cardiorespiratória sem resposta às medidas de suporte avançado de vida. Este caso relata uma causa incomum de hematoma retroperitoneal e choque hemorrágico que deve ser tido em conta num departamento de emergência.

Palavras chaves: Ruptura espontânea, veia ilíaca, hematoma retroperitoneal, choque hemorrágico.
BACKGROUND

Spontaneous retroperitoneal hematoma (SRH) is a rare entity. Aetiologies as well as the precise mechanism leading to SRH are unclear in most of the reported cases.\(^1\)

Spontaneous iliac vein rupture is an uncommon cause of SRH.\(^2\) There have been less than 40 cases described worldwide since the first documented case in 1961.\(^3\)

The rupture of an iliac vein is life threatening and mostly results from trauma or injury during pelvic surgery. Traumatic vascular injury is an emergent event and usually can be recognized by mechanism and clinical findings and managed early on.\(^4,5\)

However, non-traumatic vascular injury lacks obvious clinical presentations, so the management usually is delayed with most cases being misdiagnosed and treated inadequately.\(^4\)

CASE PRESENTATION

A 49-year-old woman was admitted to the emergency department complaining of sudden lumbar pain. The pain was sharp, stabbing in nature and without radiation.

The patient had a past history of menometrorrhagia and ferropenic anaemia and was on oral iron supplement for this reason. The patient referred no other past medical or surgical history.

Neither the patient nor her family remembered a history of trauma that could be related to the symptoms nor any straining preceding the onset of symptoms.

She was observed and no abnormal findings on physical examination were reported. No blood test or image study was made at this time. She was discharged with the diagnosis of a lumbar sciatic pain and medicated with analgesic.

Two days after this episode, she was re-admitted to the emergency room after a syncope episode. She maintained the lumbar pain and also presented abdominal diffuse pain. She denied any other new symptom.

Physical examination revealed low blood pressure (50/30 mmHg) with a high pulse rate (120 bpm). She was conscious but pale and sweating. The abdominal examination revealed a tender, non-pulsatile mass on the right lower quadrant.

On the emergency room, a point-of-care ultrasound (POCUS) was done as a complementary tool to the medical examination. It was identified a collapsed inferior vena cava with no associated intra-abdominal free fluid.

Volemic resuscitation with intravenous crystalloids was immediately initiated.

Initial blood test showed anaemia, with a haemoglobin level of 10,9 g/dL, a haematocrit level of 34,6%. Other chemistry parameters were within the normal range.

INVESTIGATION

Once the patient was stabilized with prompt resuscitation an urgent contrast-enhanced computed tomography (CT scan) was performed, revealing a large retroperitoneal haemorrhage, extending from the left renal hilum to the third portion of the duodenum and inferiorly involving the right iliac artery and vein. No active arterial haemorrhage was identified (Fig. 1).

During the abdominopelvic CT scan the patient collapsed, with electric instability and worsening of the hemodynamic status.

For this reason, the exam was interrupted, and the patient was immediately taken to the operating room. The massive transfusion protocol was immediately activated.

At this point, one hour after the first blood test, another blood test was taken which showed a haemoglobin level of 5,8 g/dL and a haematocrit level of 18,8%.
Spontaneous left iliac vein rupture

TREATMENT

The massive transfusion protocol was initiated but the patient maintained hemodynamic instability.

Due to her instability, and no direct access do interventional radiology on our institution, an urgent exploratory laparotomy under general anaesthesia was performed.

Prior to the induction of anaesthesia she suffered a cardiopulmonary arrest with recovery of spontaneous circulation after 4 minutes of advanced life support.

Intra-operatively no free hemoperitoneum was identified but the presence of an extensive retroperitoneal hematoma was confirmed.

After performing a Cattell-Brasch manoeuvre the retroperitoneal cavity was exposed and opened over the hematoma. The hematoma was evacuated, and the retroperitoneal space was explored uncovering a 20 mm longitudinal anterior wall tear of the left common iliac vein, close to the iliac bifurcation, with active bleeding.

After the bleeding source was found the tear was immediately compressed.

With the collaboration of a vascular surgeon and after an infrarenal aortic clamp was placed, a primary repair was completed with a purse-string suture of 4-0 prolene. No other injury was found.

After this damage control surgery an intra-abdominal packing with surgical pads was performed and the patient was left in laparostomy with Barker’s vacuum packing technique.

During surgery the patient presented hemodynamic instability needing vasopressor support (norepinephrine and epinephrine) and massive blood products transfusion (5 red blood cells units + 5 fresh frozen plasma units + 4 platelets units + 2g of fibrinogen).

OUTCOME AND FOLLOW UP

The patient was taken to the Intensive Care Unit for postoperative follow-up where she maintained hemodynamic instability, anuria and hypothermia. Besides massive transfusion of blood products and vasopressor support she developed a refractory shock and subsided to cardiopulmonary arrest 8 hours after admission.

At autopsy no traumatic lesions were identified. Histological examination of the vein revealed “haemorrhagic dissection of the left primitive iliac vein wall, no other microscopic alterations identified” (Fig. 2).
Spontaneous rupture of the iliac vein is a rare but life-threatening event that usually occurs in middle-aged or elderly women; Hossne reported the first case in 1961 in a 48-year-old woman who was initially thought to be in haemorrhagic shock.\[4,6\]

Since 1961 less than forty-cases have been reported in the literature.\[3,7\]

According to the literature, the average age of the patients is 60.6 ± 13.4 years with a clear predominance of women. The rupture occurred on the left side in 94% of patients.\[3,8\]

Since this first case, various theories have been suggested to explain the aetiology of the disease.

The mechanical factor derives from the obstruction of the left common iliac vein by the overlying iliac arteries, as described by May and Thurner in 1957 also known as Cockett’s syndrome. Moreover, bending and/or straining can theoretically pressure a segment of the left iliac vein between the right common iliac artery and the inguinal ligament.\[3,9\]

Stretching of the pelvic structure by a previous pregnancy or tumours can be another potential predisposing factor. Furthermore, some reports emphasize vessel weakening due to intimal changes attributed to chronic vibratory irritation and flow disturbances.\[7,9\]

The inflammatory factor stems from the potential loss of vessel wall elasticity due to chronic inflammation. No histological thrombophlebitis was documented in this case.\[3,7,9\]

The hormonal factor relates to the protective effect of oestrogen on vessels, promoting compliance. This factor can potentially be contributory due to the high prevalence of this pathology in postmenopausal woman.\[9,7\]

No theory alone is likely to explain the rupture. The combination of these three factors potentially culminates in the spontaneous tear of the iliac vein.\[3\]

Spontaneous rupture of the iliac vein was often concomitant with deep venous thrombosis and Mat-Thurner syndrome. Whether DVP is a cause or a complication of iliac vein rupture is not yet known.\[6\]

In some cases, the definitive causative origin of the vein rupture remains obscure, and hence, the ruptures have been described as “spontaneous”, “idiopathic” or “no traumatic”.\[10\]

Accurate preoperative diagnosis is usually difficult. According to the literature, some common clinical characteristics include sudden onset of abdominal or lumbar pain, swelling of the limb, anaemia and shock.

Management options include open surgical procedures, endovascular interventions, combined surgical and endovascular treatment and conservative therapy.\[2,5\]

Conservative management can be an acceptable option in selected cases with stable initial vital signs, good response to hydration or transfusion, no specific findings on laboratory results and no evidence of active bleeding on imaging studies.\[11\]

In our case we adopted surgical management taking into account the patient’s instability and the findings on the CT.

The main goal of surgical management is maintaining the continuity of ruptured iliac vein achieved by direct suture or bypass reconstruction.\[1\]

Surgical intervention is associated with high mortality and morbidity. Most reported patients have been treated surgically, with a survival rate of 71%.\[6\]

Spontaneous rupture of the iliac vein is a vascular emergency that should be treated immediately to prevent death. It should be considered as one of the differential diagnoses in patients with hypovolemic shock, who have sudden onset of back or abdominal pain. Early diagnosis, prompt resuscitation and repair of the vein can provide good results.\[11\]

CONCLUSION

Spontaneous rupture of the iliac vein is a rare entity. Clinical suspicion and early diagnosis are very important since it may be life-threatening.
In middle-aged or elderly women sustaining left-sided spontaneous retroperitoneal hematoma and acute deep venous thrombosis, the possibility of spontaneous rupture of the iliac vein should be taken in account. The main clinical features include sudden lower abdominal or lumbar pain, swelling and pain of the lower limb, anaemia and shock.

Therapeutic decisions should take into account the patient stability and image findings. Open surgical procedures are associated with high mortality and morbidity. Conservative therapy seemed to be safer than open surgical procedure, provided that it can be applied. Endovascular intervention may help to improve outcomes.

REFERENCES


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