Abdominal apoplexy associated with the levonorgestrel intrauterine system - case report

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Summary

Abdominal apoplexy is defined as intraabdominal haemorrhage without an identifiable pathology. This life-threatening situation is hardly mentioned in the literature. The clinical tableau is non-specific. The onset can be with nausea, mild to severe abdominal pain, a palpable mass in the abdomen or flank and sings of hypovolemic shock. The approach should be resuscitation, look for a likely underlying cause and control the bleeding. The present report will describe a case of abdominal apoplexy in a patient using a levonorgestrel intrauterine system. The management and possible origins of abdominal apoplexy will be discussed.

Key words: Apoplexy; Haemorrhage; Intraabdominal; Bleeding; Intraperitoneal haemorrhage; Shock; Levonorgestrel intrauterine system; Idiopathic; Abdominal apoplexy.

Introduction

Intraabdominal haemorrhage is a life-threatening situation. In the majority of cases a cause can be identified. Rarely, there is no underlying pathology. Probably, Barber made the first description of such an idiopathic intra-abdominal haemorrhage in 1909 [1]. The term abdominal apoplexy was introduced in 1931 by Green and Powers [2]. In 1937, Bruce proposed that abdominal apoplexy in young patients might be due to a structural defect in the vessel wall [3].

Kleinsasser *et al.* in 1970 reported after an extensive literature review covering an era of 58 years that the mortality was 56% and in 37% (31 out of 83 patients) no bleeding source was identified [4]. The number of reported apoplexy cases are decreasing which is undoubtedly due to the more precise imaging.

We report a case of abdominal apoplexy (AA) in a patient with a levonorgestrel releasing intrauterine system (LNG-IUS). The clinical presentation, differential diagnoses, management and possible origins of abdominal apoplexy are discussed.

Case report

A 27-year-old woman entered the emergency department due to severe pain in the lower abdomen. The pain had started suddenly one hour earlier and was constant. Her previous medical history was blank. She had had one normal vaginal delivery and for almost a year she had a LNG-IUS (Mirena®). Since the LNG-IUS had been inserted she had no periods but only some spotting. Her defecation and urine patterns were regular and normal. On physical examination she was sweaty, had a pulse of 120 beats per minute, with a blood pressure of 100/70 mm Hg and a temperature of 36.5°C. The abdominal auscultation was silent and percussion was painful. On palpation there was tenderness and guarding, mainly suprapubic. Vaginal examination revealed pain in the posterior fornix and on rectal examination there was non specified diffuse tenderness, but no mass could be felt. Transvaginal scan showed a normal sized uterus with a LNG-IUS in situ and two normal sized ovaries. The pelvis was filled with a fluid collection (10 x 10 cm). When she entered the hospital her haemoglobin was 12.3 g/dl (normal value 12-15 g/dl), haematocrit 37.2 (normal 37-46%) and white count 13.1 x 10^9/l(normal 4.3-10), platelet count 221 109/l (normal 140-440), CRP < 0.5 mg/dl (normal < 0.5 mg/dl) and hCG on urine and blood were negative.

The number one and two of the differential diagnosis were an extrauterine pregnancy despite the negative hCG levels, or a ruptured appendix.

The patient underwent a laparoscopy within two hours of admission. About one litre of red fluid with a 8 x 10 x 10 cm blood clot in the right fossa ovarica was seen in the lower abdomen. The fluid and the clot were removed and a systematic evaluation of the Douglas pouch, uterus, ovaries, tubes, large and small intestines, appendix, liver, gallbladder, pancreas, stomach, abdominal oesophagus, spleen and major vessels was performed but no bleeding site could be found. The pathology examination of the removed material showed only blood. Postoperatively blood control showed that her haemoglobin had dropped 2 g/dl (10.4) and her haematocrit by 6 (30.9). Iron supplements were prescribed. She made an uneventful recovery and could be discharged after 48 hours. At check-up after six months the patient was well, with a LNG-IUS in situ.

Discussion

Intraabdominal haemorrhages are mainly due to trauma, ruptured aneurysms, tumours or anticoagulant therapy. The clinical presentation is vague with abdominal pain, nausea, vomiting, ileus and tender mass in the abdomen and flank. The onset of an acute haemoperitoneum can be divided in three phases [5]. The first phase is characterised by mild to severe abdominal pain. The second phase is almost asymptomatic and could last for hours or days. The last and third phase is characterised by an increase in symptoms especially abdominal pain and followed by hypovolemic shock.

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In the anamnesis it is important to ask about current medications and predisposing medical conditions, especially hypertension, atherosclerosis, and possible connective tissue diseases like EDS type IV [6]. The date of last menstruation should be known. If the patient has a sudden pain in the second half of her menstrual cycle, ovulation pain or ruptured corpus luteum should be considered. The *golden adagio* "Every woman is pregnant until proven otherwise" should be kept in mind. Even if the patient says that she has had her "periods" an hCG test should be performed in order to exclude pregnancy. Besides a thorough clinical examination, external features of connective tissue disease should be kept in mind. These features are generalised connective tissue laxity, including extremely soft ear cartilage, a long vertical face with a narrow jaw, an asthenic build, mild skin hyperelasticity, and joint hypermobility [6].

The differential diagnosis of intraabdominal haemorrhage includes a variety of diseases (Table 1). This list is actually unlimited and only the general causes are mentioned here. Numerous reports in the literature attribute an uncharacterised vessel wall abnormality as the cause of abdominal apoplexy in young persons [3, 4, 7-11]. Aneurysmal degeneration is probably the most common histopathologic defect [5]. Besides the aorta, aneurysms have been described in nearly all splanchnic arteries: celiac, hepatic, gastric, gastropiploic, splenic, superior mesenteric, renal, and pancreatoduodenal arteries [12].

In young patients the diagnosis of Ehlers-Danlos syndrome (EDS) has to be considered. A total of ten EDS patients have been reported with a spontaneous visceral artery rupture [6, 12]. Of this group 20% died of their haemorrhage [6, 12].

Table 1. — *The major differential diagnosis of non traumatic intraabdominal haemorrhage includes:*

Gastrointestinal
– appendicitis
– diverticulitis, including Meckel
– pancreatitis
Gynaecological
– ectopic pregnancy (tubal, ovarian, intraabdominal)
– haemorrhagic corpus luteum
– ruputured ovarian cyst
– torsion of ovary
– degeneration or torsion of myoma
– ruptured gravid uterus
Cancer
– ovarian cancer
– gastrointestinal cancer
– intraabdominal metastatic disease
- myeloproliferative disorders
Liver disease
– portal hypertension with liver cirrhosis
– ruptured hemangioma
– ruptured varices
Vascular diseases
– aneurysms (aorta or viseral)
Miscellaneous
– anticoagulation therapy
– clotting disorder
– tuberculosis peritonitis
– postcoital

Underlying development of weaknesses of the vessel wall are caused by hypertension and atherosclerosis, rheumatoid arthritis, systemic lupus erythematosus (SLE), and polyarthritis nodosa (PAN) [5]. These potential predisposing factors are however more likely in older patients than in younger patients.

A simple blood analysis with hCG should be performed. Serial measurement of haemoglobin and haematocrit will give an idea if the patient is still bleeding or not. A vaginal ultrasound is very helpful in accessing the amount of intrabdominal fluid. If in doubt whether it is blood or not a culdocentesis can be performed. If the patient is stable computerised tomography (CT) and angiography can be performed.

CT scan is a principal method of establishing the site, size and likely underlying cause of the bleeding.

The high mortality related to active intraabdominal bleeding spurs surgical exploration. Depending on the situation of the patient (stable or not) and the possible cause, either a laparoscopic approach and/or a laparotomy can be performed to identify and control the bleeding site.

The specific role of the LNG IUS in AA is unclear. The association of an IUD and ovarian pregnancy is well known [13, 14]. More then 30 years ago it was already reported that an abortion of an ovarian pregnancy could masquerade as an ovarian apoplexy i.e. rupture of the corpus luteum with haemoperitoneum [15]. Macroscopically they are almost impossible to differentiate. The majority of LNG-IUS users will be in amenorrhea. In prospective randomised controlled trials the 5-year cumulative rates for ectopic pregnancies vary from 0.5 to 0.06 per 100 users [16]. The LNG-IUS type Mirena® releases 20 mcg LNG daily. The first few months after insertion the serum LNG levels are 425 pg/ml and after six months they are 330 pg/ml [17]. These levels exceed the critical value of 200 pg/ml below which ovulation occurs [18]. However a LNG-IUS leads to increased development of ovarian cysts, especially the first year [19]. These cysts are mainly symptomless and show a high rate of spontaneous resolution. A rupture of these newly formed cysts could be a cause of AA. Yet in the current case there were no signs of a ruptured cyst during laparoscopy and also since the introduction of LNG-IUS in 1990 there has been no increase in reported AA.

In conclusion the true incidence of abdominal apoplexy is unknown. Patients with AA present in an acute clinical setting with non-specific findings. It is extremely difficult if not impossible to make a preoperative diagnosis of AA. However in every patient who presents with signs of hypovolemic shock, intraabdominal haemorrhage and no identifiable cause AA should be suspected until proven otherwise. AA is life threatening event. It is important that prompt measurements like laparoscopy or laparotomy are taken to stop the bleeding in these acute situations.

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