

phospholipids of the red blood cell membrane, leading to spherocytosis and lysis of the erythrocytes.^{2,3}

In cases of bacteraemia, 6–20% are polymicrobial. *Clostridium* can be isolated in 0.5–2% of all blood cultures, with *C. perfringens* being the most common and responsible for 20–50% of cases.⁴ Sepsis caused by *C. perfringens* has a 30-day mortality rate of 27–44%. Massive haemolysis can develop in 7–15% of cases. This factor is associated with a worse prognosis, raising the mortality rate to 70–100%, with an average time from admission to death of 9.7 h.²

Growth and identification of *C. perfringens* in blood cultures is required for definitive diagnosis. When sepsis develops, it tends to progress rapidly, and there is no time to obtain culture growth. Therefore, when massive haemolysis is detected, *C. perfringens* should be suspected and treatment started as soon as possible in order to improve the prognosis of these patients. The optimal treatment is based on high-dose penicillin G and local control of the focus by way of surgical debridement.⁵

In our two cases, due to the rapid and difficult-to-manage progression, it was not possible to supplement the antibiotic therapy and support treatment with surgical debridement. In view of the severity of the condition, *C. perfringens* should be considered in all patients with severe sepsis and gas-forming liver abscess (with or without massive haemolysis).

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2444-3824/

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Portal-splenic-mesenteric venous thrombosis in a patient with Klinefelter syndrome[☆]



Trombosis venosa portal y del eje esplénomesentérico en un paciente con síndrome de Klinefelter

Acute portal vein thrombosis (PVT) is defined as the recent formation of a thrombus in the portal vein and/or main branches, which can also involve the mesenteric or splenic veins.¹

The aetiological factors are classified as local (30%), which include cancer and cirrhosis, followed by an intra-abdominal inflammatory focus; and systemic (70%), such as myeloproliferative syndromes, primary antiphospholipid syndrome, paroxysmal haemoglobinuria, factor II or factor V Leiden mutation and deficiency of proteins C, S and antithrombin III. Risk factors related to hormone profile, such as the use of oral contraceptives and pregnancy, are not well

established. In 15% of cases there are several causes, while in 30% the cause remains unknown.^{1,2}

We present the case of a 42-year-old male smoker (10 pack-years), who was not a heavy drinker, with a history of grade I obesity, type 2 diabetes, dyslipidaemia and azoospermia who went to Accident and Emergency complaining of sudden-onset epigastric pain accompanied by sweating and dizziness, rectorrhagia and pyrexia of 38 °C. On physical examination, weight 99 kg, height 177 cm, BMI 31.6 kg/m², central fat distribution, gynaecomastia and hypogonadism. On abdominal palpation, he had left iliac fossa pain, without signs of peritonitis. Bloods showed leucocytes 18,900 with neutrophilia, Hb 14.4 g/dl, prothrombin time 92%, aPTT 31.7 s, fibrinogen 258 mg/dl and CRP 16 mg/l. X-ray of abdomen was normal.

Colonoscopy showed lesions compatible with ischaemic colitis, 28–48 cm from the border of the anus and this was confirmed by the biopsies. Computed tomography (CT) of abdomen with contrast (Fig. 1). showed partial thrombosis of the branches of the portal vein, predominantly in the left branch and the splenic and mesenteric veins, with no evidence of abscesses, neoplasia, pancreatitis or liver disease which might explain it. Thrombophilia study (proteins C, S and antithrombin III; antiphospholipid antibodies, paroxysmal nocturnal haemoglobinuria clone, prothrombin mutation G20210A, factor V Leiden mutation, JAK2 [V617F] gene and calreticulin) was normal.

[☆] Please cite this article as: Larraín M, Castillo-Regalado E, Puig-Jove C, Sala M, Masnou H. Trombosis venosa portal y del eje esplénomesentérico en un paciente con síndrome de Klinefelter. *Gastroenterol Hepatol.* 2018;41:563–565.

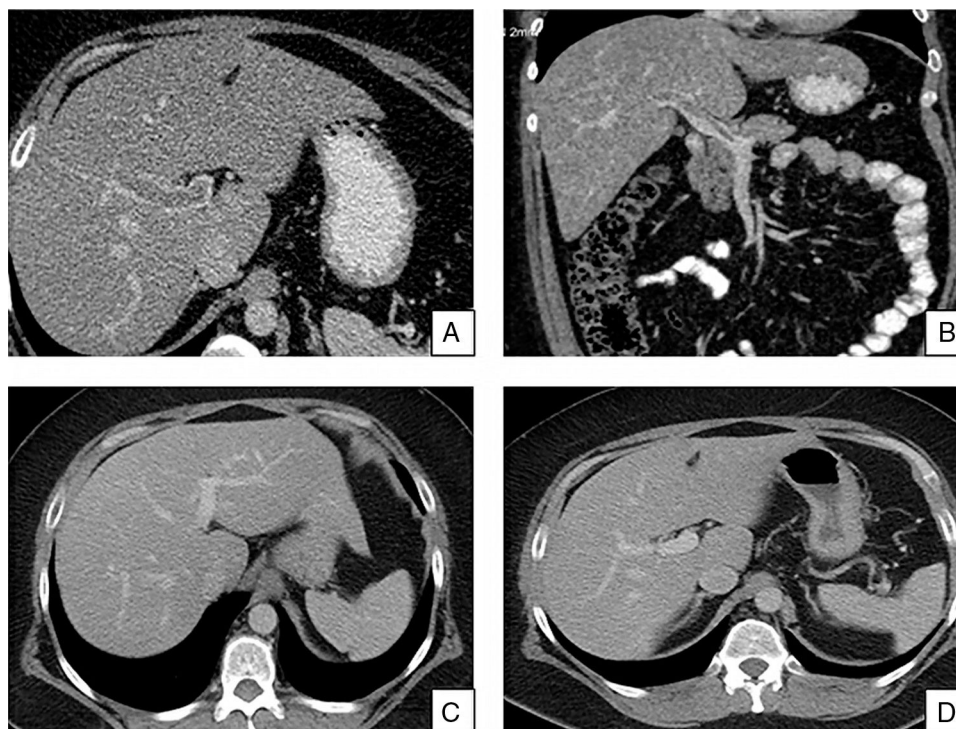


Figure 1 (A and B) Abdominal CT with contrast showing filling defect in the main portal vein and its branches, predominantly in the left portal branch, in the portal-splenic-mesenteric axis, compatible with partial thrombosis of portal vein and main branches. (C and D) Repeat abdominal CT at six months showing complete recanalisation of the main portal vein, branches and the portal-splenic-mesenteric axis.

Study of chronic liver disease was negative (transferrin saturation 22%, ferritin 492 mg/dl, ceruloplasmin 23.6 mg/dl, alpha-1-antitrypsin 156 mg/dl, TSH 3.54 mIU/l, HBsAg –ve, HCV –ve, autoimmunity negative) and liver biopsy, which showed signs of simple steatosis, ruling out cirrhosis. Endoscopy ruled out oesophageal varices.

Chromosomal analysis performed to investigate the azoospermia and testicular atrophy was compatible with a karyotype 47,XXY, with a hormonal profile consistent with hypergonadotropic hypogonadism (LH 14.77 IU/ml, FSH 30 IU/l and testosterone 74.1 ng/dl). The patient was diagnosed with Klinefelter syndrome (KS) and started on treatment with topical testosterone.

In view of the episodes of rectorrhagia, he was treated with subcutaneous enoxaparin 100 mg/day. As his clinical and radiological progress were good, it was decided to maintain the same dose for six months.¹

KS is the most common disorder of the sex chromosomes (47,XXY or mosaic). Phenotypically they have testicular atrophy, gynaecomastia and hypergonadotropic hypogonadism. The estimated prevalence is 153 per 100,000 males.³ Due to the wide phenotypic variation, only 25% of the total are diagnosed, some during a sterility study.⁴

Individuals with KS have an increased risk of venous thromboembolism (VTE), which may be explained by the androgen deficiency that increases the levels of plasminogen activator inhibitor-1 (PAI-1), causing hypofibrinolysis, platelet hyperaggregation, and an increase in factor VIII activity.⁵

The risk of developing venous thrombosis (VT) or pulmonary embolism (PE) is 5–20 times higher than that of

the general population.⁶ A Swedish retrospective study, with 1085 patients diagnosed with KS, showed that the cumulative incidence of VTE was 8.6% at the age of 50 and 20.8% at the age of 70, being comparable to hereditary thrombophilias.⁷ However, the risk of PVT or mesenteric vein thrombosis (MVT) has not been determined.

The first case of MVT in a patient with KS was published in 1988. In that case, MVT manifested as rectal bleeding and intestinal infarction, requiring resection. Anticoagulation was started postoperatively, but the duration was not specified. Aetiological study showed only a slight decrease in protein C levels. At follow-up, there was no evidence of re-thrombosis.⁸

Two cases of PVT and MVT associated with KS were later published in which no local or systemic factors for thrombosis were found. Outcomes were good in both cases, with patency of the PVT and MVT partially restored, probably due to the early administration of anticoagulation.^{9,10}

In conclusion, we have presented the case of a patient with a recent diagnosis of KS in whom no local or systemic prothrombotic factor was identified who developed portal-splenic-mesenteric venous thrombosis. After treatment with low-molecular-weight heparin for six months, he was found to have full recanalisation, with no evidence of new thrombosis at his 12-month follow-up. The optimal duration of anticoagulant treatment in PVT has not been fully established. However, it is recommended that treatment should be maintained for at least six months, and that permanent anticoagulation be considered in patients with an underlying prothrombotic factor.¹ As a high prevalence

of thromboembolic events has been reported in KS, the need for long-term or even permanent anticoagulation should be considered in these patients.

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Syphilis, unusual cause of abdominal pain[☆]



Sífilis, causa poco habitual de dolor abdominal

Until the introduction of penicillin in the mid-twentieth century syphilis, caused by the bacterium *Treponema pallidum*, was one of the most common sexually transmitted infections. The incidence of syphilis decreased significantly following the discovery and subsequent introduction of penicillin, but has begun to increase over the last decade, mainly in the context of HIV. In Chile, in the first quarter of 2016, 846 cases were reported to the Ministry of Health, with an incidence rate of 4.6 per 100,000 population.¹

T. pallidum has a similar pathogenesis to *Mycobacterium tuberculosis*, the pathogen responsible for tuberculosis. Both agents have in common a slow replication time, often asymptomatic primary infection, and silent dissemination which can lead to multi-organ failure.

Syphilis is classically described as having three phases: (a) primary phase in which local multiplication occurs; (b) secondary phase with lymphatic and haematogenous dissemination and skin, eye, central nervous system (up to 40% of cases) involvement observed in the patient, and (c) tertiary phase, mediated by hypersensitivity, which can manifest up to 30 years after infection, and among other problems, can involve bones and central nervous and cardiovascular systems.²

We present the case of a 68-year-old male patient with a history of hypertension who consulted for a week-long history of colic-type abdominal pain in the epigastrium radiating to both hypochondria.

On physical examination, the patient was alert and orientated, haemodynamically stable, afebrile and eupnoeic. Examination of the patient's skin revealed confluent macular exanthema, predominantly on the trunk. Cardiopulmonary examination was normal. His abdomen was soft and depressible, but sensitive to palpation in the epigastrium. There were no signs of peritoneal irritation.

Laboratory tests showed haemoglobin 17.7 g/dl, leucocytes 13,470, 316,000 platelets/ μ l, ESR 13 mm/h, CRP 15 mg/l, lipase 42 U/l, creatinine 0.9 mg/dl and normal liver profile, and abdominal ultrasound was normal.

As the abdominal pain continued to persist, even with high doses of opioids, a computed tomography (CT) angiography study was performed. The CT showed diffuse wall thickening of the left gastric artery with an area of multi-segmental stenosis, with no abnormalities in renal and coeliac arteries (Fig. 1).

An increase in macular lesions and palmo-plantar involvement were also detected, as a result of which a Venereal Disease Research Laboratory (VDRL) test was requested, showing titres of 1:16. Other results included: HIV negative; FTA-ABS positive 1:32; study with ANA, ANCA, ENA autoantibodies negative; serology for HBV and HCV negative; cytology study in cerebrospinal fluid detected presence of 8–10 predominant mononuclear and physicochemical cells showing proteins of 95 mg/dl and normal glucose, with VDRL 1:8.

Magnetic resonance imaging (MRI) of the brain detected leptomeningeal meningovascular involvement compatible with neurosyphilis. Although the skin and laboratory findings

[☆] Please cite this article as: Navea C, von Mühlenbrock C, Cabello N, Echeverría M, Jiménez A, Poniachik J. Sífilis, causa poco habitual de dolor abdominal. *Gastroenterol Hepatol.* 2018;41:565–566.