

Fitz-Hugh-Curtis syndrome

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Abstract: *Fitz-Hugh–Curtis syndrome is a rare complication of pelvic inflammatory disease (PID) involving liver capsule inflammation leading to the creation of adhesions. The condition is named after the two physicians, Thomas Fitz-Hugh, Jr and Arthur Hale Curtis who. It also known as “gonococcal perihepatitis” or “perihepatitis syndrome.” Most cases of Fitz-Hugh-Curtis syndrome are caused by infection with the bacterium Chlamydia trachomatis and Neisseria gonorrhoeae. Fitz-Hugh-Curtis syndrome is marked by an acute onset of right upper quadrant (RUQ) abdominal pain aggravated by breathing, coughing or laughing, which may be referred to the right shoulder. CT of the abdomen with IV contrast may show subtle enhancement of the liver capsule. Testing for gonorrhoea and chlamydia should be performed to make the diagnosis. Treatment involves a course of antibiotics to cover the appropriate organisms, typically ceftriaxone plus azithromycin. Laparoscopy for lysis of adhesions may be performed for refractory pain.*

Keywords: Fitz-Hugh–Curtis syndrome, gonococcal perihepatitis, perihepatitis syndrome, gonorrhoea, guitar string, Gardnerella, peritoneal carcinomatosis

1. Introduction

Pelvic inflammatory disease is an infection of a woman’s reproductive organs. Most often it’s caused by sexually transmitted infections (STIs) like chlamydia and gonorrhea. It usually causes inflammation of the uterus, ovaries, fallopian tubes, cervix, or vagina. Sometimes, this inflammation spreads to the covering of the liver or the tissues surrounding the liver in the abdomen. It can also spread to the diaphragm, the muscle that separates the abdominal cavity and the chest.

Fitz-Hugh–Curtis syndrome is a rare complication of pelvic inflammatory disease (PID) involving liver capsule inflammation leading to the creation of adhesions. The condition is named after the two physicians, Thomas Fitz-Hugh, Jr and Arthur Hale Curtis who first reported this condition in 1934 and 1930 respectively. It also known as “gonococcal perihepatitis” or “perihepatitis syndrome.”



Figure 1

“Violin-string” adhesions of chronic Fitz-Hugh–Curtis syndrome

Epidemiology

The prevalence of Fitz-Hugh–Curtis syndrome in women with mild to moderate pelvic inflammatory disease may approximate 4%. The prevalence may be higher in genital tuberculosis. It most commonly occurs in women of childbearing age; however, there have been rare cases reported in males. The actual incidence of Fitz-Hugh-Curtis syndrome in the general population is unknown. In extremely rare cases, Fitz-Hugh-Curtis syndrome was first described in the medical literature in 1920.

Causes

Most cases of Fitz-Hugh-Curtis syndrome are caused by infection with the bacterium Chlamydia trachomatis, which causes Chlamydia or the organism Neisseria gonorrhoeae, which causes gonorrhoea. Chlamydia and gonorrhoea are common sexually transmitted diseases (STDs). Researchers believe that more cases of Fitz-Hugh-Curtis syndrome are caused by infection with Chlamydia trachomatis than with Neisseria gonorrhoeae.

The exact process by which such infections cause Fitz-Hugh-Curtis syndrome (pathogenesis) is not completely understood. Some researchers believe that it occurs because of infection of the liver and surrounding tissue, which may result from bacteria traveling from the pelvis directly to the liver or via the bloodstream or lymphatic system.

Some researchers have speculated that Fitz-Hugh-Curtis syndrome may occur because of an improper immune system response (autoimmunity) to infection with Neisseria gonorrhoeae or Chlamydia trachomatis. Autoimmune disorders are caused when the body’s natural defenses (antibodies, lymphocytes, etc), against invading organisms suddenly begin to attack perfectly healthy tissue. Several studies have demonstrated that individuals with Fitz-Hugh-Curtis syndrome have high levels of antibodies against Chlamydia trachomatis. More research is necessary to determine what role autoimmunity plays in the development of Fitz-Hugh-Curtis syndrome.

Fitz-Hugh-Curtis syndrome is characterized by the developed of string-like, fibrous scar tissue (adhesions) between the liver and the abdominal wall or the diaphragm.

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2. Signs and symptoms

Fitz-Hugh-Curtis syndrome is marked by an acute onset of right upper quadrant (RUQ) abdominal pain aggravated by breathing, coughing or laughing, which may be referred to the right shoulder. There is usually also tenderness on palpation of the right upper abdomen and tenderness to percussion of the lower ribs which protect the liver. Surprisingly there is often no or only minimal pelvic pain, vaginal discharge or cervical motion tenderness, which may lead to the diagnosis being missed. This may be due to infectious bacteria bypassing pelvic structures on the way to the liver capsule. Moving usually makes it worse.

Other symptoms might include:

- Fever
- Chills
- Night sweat
- Nausea and vomiting
- Hiccups
- Headaches
- A general feeling of poor health (malaise)

Risk factors

Women of childbearing age who have PID have the biggest chance of developing Fitz-Hugh-Curtis syndrome. Teens are high-risk, too, because they're more prone to infections. In very rare cases, men can also get it.

Pathophysiology

Fitz-Hugh-Curtis syndrome occurs almost exclusively in women. It is usually caused by *Chlamydia trachomatis* (Chlamydia) or *Neisseria gonorrhoeae* (Gonorrhoea) though other bacteria such as *Bacteroides*, *Gardnerella*, *E. coli* and *Streptococcus* have also been found to cause Fitz-Hugh-Curtis syndrome on occasion. These bacterial pathogens cause a thinning of cervical mucus and allow bacteria from the vagina into the uterus and fallopian tubes, causing infection and inflammation. Occasionally, this inflammation can cause scar tissue to form on Glisson's capsule, a thin layer of connective tissue surrounding the liver

Diagnosis

CT of the abdomen with IV contrast may show subtle enhancement of the liver capsule. Testing for gonorrhoea and chlamydia should be performed to make the diagnosis. An endocervical or low vaginal swab should be taken to test for these organisms. Antibody testing is rarely required but may be considered if other tests are non-diagnostic and suspicion is high. Abdominal will typically be normal. Liver function tests will typically be normal or unchanged from baseline as the infection does not involve the liver parenchyma. If a D-dimer is ordered, which it often is when there is pleuritic torso pain, it will usually be markedly elevated but other testing for pulmonary embolism will be normal. Laparoscopy is also rarely required, but may be performed when the diagnosis is uncertain and may reveal "guitar string" adhesions of parietal peritoneum to liver.

Differential diagnosis

Imaging differential considerations include:

- peritoneal carcinomatosis: shows more peritoneal nodularity and a solid component, overt pelvic malignancy on imaging and has a different clinical presentation
- appendicitis: interestingly, both as differential diagnosis and possible complication

Treatment

Treatment involves a course of antibiotics to cover the appropriate organisms, typically ceftriaxone plus azithromycin. Laparoscopy for lysis of adhesions may be performed for refractory pain.

If treating the underlying STI doesn't ease abdominal pain, laparoscopy is performed to remove scar tissue around liver. During the procedure, a small, thin tool inserted through a small cut made in abdomen and cut away the dead tissue ("adhesions"). This is rarely done. Treating PID to cure Fitz-Hugh-Curtis syndrome

Since Fitz-Hugh-Curtis syndrome emerges from PID, symptomatic therapy is the key. Judlin et al. published recent data from the MONALISA study, according to which a fourth-generation synthetic fluoroquinolone antibacterial agent, moxifloxacin, is an ideal monotherapy for the treatment of PID. Nevertheless, antibiotic therapy should be directed to the most likely pathogens, such as *N. gonorrhoeae* and *C. trachomatis* and microorganisms found in the endogenous flora of the vagina and cervix, such as anaerobic bacteria and facultative bacteria, many of which are associated with bacterial vaginosis. Parenteral and oral therapies appear to have similar clinical efficacy in treating women with PID of mild or moderate severity. The gold standard seems to be cefotetan 2 g IV every 12 h or cefoxitin 2 g IV every 6 h with doxycycline 100 mg orally or IV every 12 h for parenteral treatment and ceftriaxone 250 mg IM in a single dose with doxycycline 100 mg orally twice a day for 14 days, with or without metronidazole 500 mg orally twice a day for 14 days, for per os treatment. Clinical experience should guide decisions regarding transition to oral therapy, which usually can be initiated within 24 h of clinical improvement. Oral therapy can be considered for women with mild to moderately severe acute PID, as the clinical outcomes among women treated with oral therapy are similar to those treated with parenteral therapy. Women who do not respond to oral therapy within 72 h should be reevaluated to confirm the diagnosis and should be administered parenteral therapy on either an outpatient or inpatient basis.

Prevention

Since this condition is linked to PID, the best way to prevent it is to not get PID. To reduce risk the following measures can be taken

- Use condoms and limit your number of sex partners
- Regularly get tested for STIs if you're sexually active
- Ask any sexual partners to get tested
- Avoid douching, which can make you more prone to vaginal infections

3. Summary

Fitz-Hugh–Curtis syndrome is a rare complication of pelvic inflammatory disease (PID) involving liver capsule inflammation leading to the creation of adhesions. It also known as “gonococcal perihepatitis” or “perihepatitis syndrome.” The diagnosis and management of HFCS are not easy and require an interprofessional team that ideally includes a gynecologist, radiologist, emergency department physician, specialty nurse, infectious disease expert, and laboratory professionals. Treatment of HFCS coincides with the management of PID. The primary care provider and nurse practitioner need to treat the partner and educate the patient on safe sex practices.

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