

ACUTE CHYLOUS PERITONITIS SIMULATING ACUTE APPENDICITIS: A CASE REPORT AND REVIEW OF THE LITERATURE

Fiaz Maqbool Fazili, MBBS, FICA, FICS; Fazal Imtiaz Khawaja, MD, FACC

Chylous effusions into pleural and peritoneal cavities are unusual, and are often a manifestation of an underlying disease process. A sudden outpouring of chyle into the peritoneal cavity may, on rare occasions, produce acute chylous peritonitis. Such patients usually present with features of acute abdomen, however, the diagnosis is rarely suspected preoperatively. Very few cases of acute chylous peritonitis have been described in the literature, and to the best of our knowledge, this is first of its kind in Saudi Arabia. Due to its rarity and presentation as compared to a common surgical emergency such as acute appendicitis, the clinical features and management of acute chylous peritonitis are discussed.

Case Report

A 25-year-old Nigerian male was referred to the emergency room of King Fahad Hospital, in Medina, Saudi Arabia, because of generalized abdominal pain, vomiting and anorexia of three days' duration. The pain had started as a dull ache just above the umbilical region, had gradually increased in severity and become generalized. A day prior to his admission, the pain had localized to the right lower quadrant of the abdomen. The pain was continuous, severe and aggravated by any movement. The patient did not give any history of previous trauma or similar pain. He had been unable to take anything orally for over 24 hours prior to admission. There was nothing relevant in his past medical history, except that five years previously, he had been hospitalized for respiratory infection for five days. Details of this were not available. For the previous six years, the patient had been smoking about thirty cigarettes per day. He had not consumed alcohol for the past two years. There was no history of diabetes, hypertension, hyperlipidemia or recurrent abdominal pains in his family.

On examination, the patient was mildly dehydrated. His temperature was 37.5°C, pulse rate was 115/min, and

blood pressure was 110/85 mm Hg. Systemic examination was unremarkable. The abdomen was not moving adequately with respiration, and there was pain on coughing, especially in the right lower quadrant. The flanks were not full. There was guarding and tenderness all over the abdomen, especially in the right iliac fossa. Bowel sounds were audible. Total leukocyte count was 20,000/ μ L, Hb 16.8 g%, urea 4.3 mmol/L, creatinine 87.9 mmol/L, glucose 4.6 mmol/L, ALT 22 IUL, AST 28 IUL, amylase 104 IUL, cholesterol 3.5 mmol/L and triglycerides 0.34 mmol/L. Chest x-ray was normal. Abdominal x-ray showed nonspecific gas-filled loops of large intestine. The patient was suspected to have acute appendicitis with possible perforation.

The abdomen was opened by right grid-iron incision, revealing unusually pale and white subcutaneous fat. The epimysium over the fibrous sheath aponeurosis of external oblique showed multiple strands with an interwoven network of whitish streaks. Between the strands was a whitish thickening of 1-2 mm. When the peritoneum was opened, milky white, odorless, non-clotting fluid gushed out. About one liter of this fluid was drained, and samples were taken for bacteriological and biochemical examination. On gross examination, the appendix appeared normal. Since no source giving rise to such a fluid collection was found, a formal laparotomy was performed through midline incision. The peritoneal cavity was full of the same milky fluid. The root of mesentery appeared normal. No abnormality could be detected in any of the solid and hollow abdominal viscera. The entire retroperitoneal space also contained the same whitish fluid with an intact overlying peritoneum, so the retroperitoneal space was opened. There were no dilated lymphatics, and no source of leakage of chyle was found. A provisional diagnosis of acute chylous peritonitis of unknown etiology was made, and a prophylactic appendectomy performed. A thorough peritoneal wash with warm normal saline was performed. Both grid-iron and midline incisions were closed. A tube drain (24 F) was kept in the right paracolic gutter, which drained 50 mL of the whitish fluid or chyle for the first 24 hours. The drain was removed after 48 hours. Biochemical analysis of the peritoneal fluid revealed triglycerides 8.60 mmol/L and protein 45.2 g/L. Simultaneous serum triglyceride level was 0.34 mmol/L, and the plasma protein was 67.2 g/L. Serum amylase level

From the Departments of Surgery (Dr. Fazili) and Gastroenterology (Dr. Khawaja), King Fahad Hospital, Medina Munawarah, Saudi Arabia.

Address reprint requests and correspondence to Dr. Fazili: P.O. Box 5147, Medina Munawarah, Saudi Arabia.

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was normal. The fluid was bacteriologically sterile. The patient had a smooth postoperative period and was discharged on the sixth postoperative day with diagnosis of acute chylous peritonitis. He has remained in good general health without any abdominal complaints during nine months of follow-up.

Discussion

Acute chylous peritonitis is defined as an acute abdomen with all the signs of acute peritoneal irritation resulting from free chyle in the peritoneal cavity without any underlying disease. Renner¹ described the first case in 1910. Since then, very few cases have been reported in the literature. The striking appearance and relative rarity of chylous peritoneal effusions have provoked considerable interest among clinicians, but surprisingly, little information is available regarding its etiology, diagnosis and treatment.

Differential Diagnosis of Milky Peritoneal Fluid

It is important to emphasize that all cloudy or milky fluids aspirated from the peritoneal cavity are not chyle. True chyle must be differentiated from pus, chyloform ascitic fluid and pseudo-chyle.² Chyle is essentially lymphatic fluid with a milky appearance similar to peripheral lymph. It is modified by the state of absorption which prevails at the time of collection. It is normally odorless, however, it occasionally has the odor of digested food. The chemical composition of chyle is quite specific. No peritoneal fluid other than chyle has fat content greater than plasma (0.04%-4%). The cholesterol concentration is also high. Albumin and globulin levels are lower than the blood. Chyle separates into three layers on standing: an upper creamy layer, a dividing watery layer, and a lower colorless opaque layer.

True chyle can be identified by adding ether, which floats on top of the extracted fat over the clear bottom layer (Ether test). Chyle has alkaline reaction and stains positive for fat (Sudan III). The white cells average 5000/mL, and are predominantly lymphocytes. It is bacteriostatic, sterile on culture, and resists putrefaction.³ Pseudo-chyle is an opalescent chyle-like fluid aspirated from patients with widespread peritoneal seedling of malignant neoplasm. The opalescence is due to degenerated cells or substances of a lecithin globulin character.⁴ Fluids from the pseudomyxoma peritonei and peritoneal tuberculosis of the wet variety can easily be differentiated from chyle through gross appearance and laboratory tests.⁵ The basal flow rate of lymphatic fluid through the thoracic duct averages about 1.0 mL/kg/hour, with a total volume of 1500-1700 mL per day. These volumes are markedly increased by ingestion of fats, and fluid rates as high as 200 mL/hour have been reported.⁶ Loss of this fluid to outside or a fistulous communication will have profound fluid, electrolyte and nutritional effect upon the patient. Normally, the

lymphatics drain the tissues of any excessive fluid, which remains after filtration, by a re-absorptive process through the capillary walls. Once the pressure in tissue fluid rises, there is dilatation of lymphatic vessels, and hence increase in the formation of lymph. There is weeping of chyle from dilated lacteals on the bowel surface.

Factors producing chylous effusions can be intrinsic or extrinsic mechanical obstruction, or can be due to injury to cisterna chyli. Krizek et al.⁵ classified patients with chylous peritonitis into obstructive, traumatic, idiopathic, and those associated with mesenteric cyst. In the obstructive type, the increased pressure within the thoracic duct or liver results in lymphostasis, edema, and in apparent rupture of small lymphatic vessels, which would lead to chylous effusion.⁷ However, there may be increased thoracic duct pressure without any evidence of chylous effusion.⁸ Various other causes include intra-abdominal, retroperitoneal, thoracic or mediastinal neoplasms, incarcerated hernias, midgut volvulus and enlarged lymph nodes. In traumatic cases, there is free peritoneal leak of chyle due to blunt trauma abdomen, causing laceration or bursting of cisterna chyli. Trauma forms 10%-15% of cases, and the most common site is the base of the mesentery. Trauma does not necessarily have to be violent to produce chylous leakage. Even minor trauma, such as severe coughing or attempts at resuscitation may result in injury to delicate lymph channels.

Other causes include congenital disease of lymphatic system, such as atresia or stenosis of lymphatic system, and inflammations such as syphilis and tuberculosis. In mesenteric cyst, the rupture of chyle containing cysts of mesentery, may cause acute chylous peritonitis. Congenital causes are more common in infants, while inflammatory and neoplastic causes are more common in adults. Embolic occlusion of subclavian vessels, nephritis, filariasis, bilharziasis and thoracic duct obstruction by aneurysm are other conditions that have been reported to be associated with chylous effusions.³ There is a high incidence of patients, such as the case described, in whom etiology is not determined, either at surgical exploration, or at post-mortem examination.^{9,10}

Clinical Features

Chylous peritonitis usually occurs in previously healthy individuals, and frequently follows meals rich in fat.¹¹ There is no predilection for sex or race, and age ranges from one month to 72 years. The symptoms and signs of peritonism are usually maximal in the right iliac fossa, as was the presentation in our case. This is probably due to the pooling of chylous fluid in the pericolic gutter. Anorexia, nausea and vomiting are frequent. Laboratory tests are of value in a negative way, being helpful in excluding diseases such as pancreatitis, hyperlipidemia, liver, renal or hematological abnormality. White blood cell count is usually elevated, as it is in any inflammatory process, with a shift to the left in a mild lymphopenia,

although the amount of chyle lost is usually insufficient to produce a noticeable drop in circulatory lymphocytes. Amylase level is not elevated. Preoperative BUN, electrolyte, protein, and cholesterol are of normal value. Radiological examination of the chest is usually normal, but in a few cases, intrathoracic pathology obstructing the thoracic duct may be seen. Roentgenogram of the abdomen is helpful in excluding other pathology, and may suggest free fluid in the abdominal cavity, but is of low diagnostic yield. The only diagnostic test in this syndrome is an abdominal paracentesis, which is only possible if such a diagnosis is preoperatively suspected, or the paracentesis has been done for assessing the nature of fluid responsible for peritonism.⁵

The optimal management of true chylous peritonitis depends upon the underlying etiology. In patients with symptoms of an acute abdominal process, immediate exploration should be done. The disease process is often established at laparotomy, and in a number of instances proves to be surgically correctable. The source of chylous extravasation is corrected by ligation of leaking lymphatics or removal of offending lesion, such as the release of adhesions, removal of mesenteric cysts, excision of small tumors or repair of mesenteric traumatic tear.¹⁰ In these cases, complete recovery usually follows, and overall results are gratifying. Management of mesenteric cysts is more challenging. If they are well localized, they may be shelled out easily. Sometimes a limited resection of adjacent bowel in conjunction with the cyst has to be performed. A large diffuse cyst or multiple small cysts make limited resection technically difficult, especially when the cysts are located at the base of the mesentery. In such cases, hazardous dissections should be avoided. A limited excision and marsupialization has proved successful with only minimal complications and a low recurrence rate.⁵ Granulomas, particularly those of tuberculous nature, need special attention—closure of the fistula with a plug of omentum and anti-tuberculous therapy can assure reasonable recovery. When lymphomatous process has been discovered, radiation therapy has been effective in controlling chylous ascites and effusions, but the overall results have been poor.³ When a specific etiological factor is not found at the time of operation, retroperitoneal dissection in the search of a leak is not recommended. Usually the site of leakage cannot be identified. A spontaneous closure of the leak and a complete recovery is the rule, as happened in our case. No report of persistence of leak has been reported. A lymph node biopsy may be considered when acute mesenteric or retroperitoneal adenitis is suspected as an etiological factor. Additional incidental or associated surgical procedures in the presence of chylous peritonitis can be carried out safely.

The failure of bacteriological invasion is due to sterile and bacteriostatic quality of chyle. Abdominal drainage is not necessary, as results are the same with or without

drainage.⁵ Other therapeutic approaches include repeated paracentesis, bed rest, diuretics, and salt restriction. Hyperalimentation to give rest to the bowels and a low-fat diet supplemented with medium-chain triglycerides have been tried with success. The condition may subside after single or multiple paracentesis and supportive therapy. Severe malnutrition and often death accompany multiple paracentesis and removal of massive quantities of chyle over short periods of time.¹²

Several surgical techniques have been advocated to provide internal drainage of chyle into subcutaneous tissue, or venous system in order to avoid the potentially dangerous losses of chyle that accompany multiple paracentesis. Venoperitoneal shunts have been credited with affording good internal drainage, but these shunts remain effective for only short periods of time.¹³

In summary, a rare cause of acute chylous peritonitis mimicking acute appendicitis is described where no cause could be identified. Simple peritoneal lavage resulted in complete recovery of the patient.

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