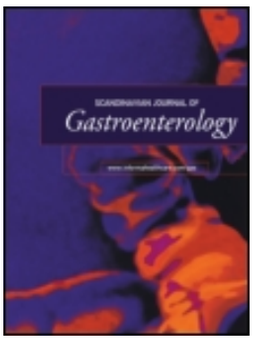

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CASE REPORT

Recurrent cholangitis associated with biliary sludge and Phrygian cap anomaly diagnosed by magnetic resonance imaging and magnetic resonance cholangiopancreatography despite normal ultrasound and computed tomography

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Abstract
A 31-year-old woman presented with a one and half years' history of intermittent right upper quadrant (RUQ) pain, high fever and severely painful, warm and reddish swollen skin lesions on the fingers. Acute attack resolution occurred within 2 weeks after treatment with non-specific antibiotics. Low-grade fever (around 37.5°C) and less painful swellings continued for 6 months after each attack. Abdominal ultrasound and computed tomography (CT) scans did not show any abnormality during the attacks. Biopsy of the skin lesions after the second attack revealed lymphocytic vasculitis. All laboratory studies including rheumatologic serology panel were normal. One month after the complete resolution of the second attack, the patient was observed to have high fever, the same skin lesions on the fingers as at the initial stage, nausea and marked abdominal pain in the RUQ. Routine laboratory studies including complete blood count, liver function tests and serum amylase and lipase levels were normal. An abdominal CT scan revealed a slight thickening of the gallbladder wall (3.9 mm). Two weeks later, abdominal magnetic resonance imaging (MRI) and magnetic resonance cholangiopancreatography (MRCP) were performed because of persistent abdominal pain. They revealed both biliary tract and pancreatic gland alterations consistent with past cholangitis and pancreatitis with coexisting Phrygian cap anomaly and biliary sludge on the neck of the gallbladder.

Key Words: *Biliary sludge, cholangitis, fever, pancreatitis, Phrygian cap anomaly, vasculitis*

Introduction

Recurrent pyogenic cholangitis (RPC), known as oriental cholangitis, is characterized by recurrent inflammation of the bile ducts [1]. RPC typically occurs in young patients from Southeast Asia [2–4]. In this paper, we present a young adult woman with recurrent cholangitis and palmoplantar vasculitis associated with biliary sludge and Phrygian cap anomaly of the gallbladder diagnosed by magnetic resonance imaging (MRI) and magnetic resonance cholangiopancreatography (MRCP). Clinically, this case presented with recurrent high fever, nausea, abdominal pain and painful, swollen skin lesions on the fingers, while serum liver function tests, pancreatic enzymes, abdominal ultrasound and computed tomography (CT) scan were all normal.

Case report

A 30-year-old woman was admitted to our hospital (in February 2003) with high fever (39°C, axillary), fatigue, headache, loss of appetite, sputum with cough, nausea and pain in the right upper quadrant (RUQ) of the abdomen, and painful, reddish, warm, nodular swellings on her fingers for the past 7 days. The patient had no prior history of jaundice, hepatitis, parasitic infections, gallstones, peptic ulcer or abdominal trauma, apart from a history of tuberculosis (when she was 6 years old). Also, she

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believed she had pulmonary tuberculosis when she was 20 years old, with a 12-kg weight loss, abdominal pain, nausea, fever and similar painful swellings on her toes. At that time, she had undergone a 300 mg a day isoniazid regimen for 4 months, as well as treatment with antibiotics for 2–3 weeks. There was no history of alcohol consumption, any form of herbal medicine or smoking. The patient was not on any particular diet or taking oral contraceptives and she was not sexually active. She did not report on any recent travel anywhere outside her home country, or any visits to the dentist, or blood transfusions, or surgery in the past 6 months. There was no family history of tuberculosis, hepatitis, cholangitis, gallstones or any rheumatologic disease, particularly vasculitis.

Physical findings were limited to the skin and gastrointestinal system. The patient was of normal weight with a body mass index of 24. Fever was 37.7°C, axillary. There was slight tenderness in the RUQ. Painful, reddish, warm and swollen skin lesions were seen on her fingers, bilaterally. These very sensitive swellings were located mainly on the palmar face of the fingers, but also on the dorsal sides. Their size varied from 0.1 to 0.5 cm. There was no other rash, or any articular deformity, or any chronic liver disease stigmata. Laboratory evaluations revealed haemoglobin of 12.9 g/dl, white blood cell count of 4000/mm^3, with 56% neutrophils and 44% lymphocytes and platelet count of 170,000/mm^3. The erythrocyte sedimentation rate (ESR: 6 mm/h), C- reactive protein (CRP: 0.5 mg/dl), ASO (114 IU/ml), RF ($<$40 IU/ml), antinuclear antibody tests, ds-DNA, and p-ANCA were all within the normal range. The following laboratory tests were also within the normal range: liver function tests, serum albumin, globulin, α-1 antitrypsin, cholesterol, triglyceride, fasting sugar, sodium, calcium, magnesium and prothrombin time (and INR (international normalized ratio) of prothrombin time). Viral serology screening was negative for hepatitis B surface antigen (HBsAg), hepatitis B core immunoglobulin G (IgG) and M (IgM), anti-hepatitis C virus (anti-HCV) and anti-HIV. Results of autoantibody tests (smooth muscle antibodies, antibodies to liver/kidney microsome type 1 and mitochondrial antibodies), the Wright test for brucellosis and the Venereal Disease Research Laboratory (VDRL) test for syphilis were all negative. An eye examination was performed but revealed no abnormality. PPD was measured with 20 mm induration. Chest X-ray showed calcifications on the left upper lobe caused by the previous tuberculosis. CT scanning of the chest and upper abdomen did not reveal any active lesions, with the exception of signs of past tuberculosis. Nor did abdominal ultrasound show any abnormality of the liver, gallbladder and pancreas. Non-specific respiratory tract infection, gastritis and vasculitis (clinically) were diagnosed. A non-specific regimen of amoxicillin-clavulonic acid and proton-pump inhibitor therapy was prescribed for 14 days. The patient gradually recovered, but her painful swellings and fever persisted. Fever and skin lesions recovered completely after 6 months.

In December 2003, the patient was again admitted with high fever, RUQ pain, nausea, weakness, loss of appetite and the same painful swellings on her fingers. Her physical examination was normal, apart from the same swellings on her fingers. Two weeks later, we saw another two similar lesions on her toe. All laboratory investigations (liver and renal function tests, electrolytes, lipid panel, thyroid function tests, muscle enzymes, fasting cortisol, CRP, ESR, FANA, ds-DNA, p-ANCA, RF, complete blood count and urine analysis) were performed and revealed no abnormality. Iron and copper studies were also normal. Chest X-ray and abdominal ultrasound did not show any recent findings. Biopsies of skin lesions were performed on the 45th day after start of the initial lesions, and revealed marked lymphocytic infiltration around both the venules and small arterioles, and endothelial swelling. Lymphocytic vasculitis was diagnosed, pathologically. The patient was again prescribed some antibiotics. The painful skin lesions and fever continued for 6 months.

She was seen in June 2004 with fever (37.6°C, axillary), weight loss (3 kg within the previous 10 days), nausea and a moderate pain in the RUQ which radiated to the back. Her whole body examination was normal except for finger lesions at the initial stage and marked tenderness in the RUQ of the abdomen. Laboratory examination revealed normal values of serum liver function tests, amylase and lipase. Furthermore, both ESR and CRP were normal. A CT scan of the abdomen revealed a slight thickening of the gallbladder wall (3.9 mm), but otherwise normal. Serum levels of CEA and CA 19-9 were also normal.

Two weeks later, upper abdomen MRI and MRCP were performed because of persistent abdominal pain and these revealed past cholangitis (Figures 1a and b) and pancreatitis features, as well as biliary sludge on the gallbladder neck and a Phrygian cap anomaly of the gallbladder (Figures 2a and b). In order to further investigate the possibility of a peptic disease, sarcoidosis, tuberculosis, amyloidosis and parasites, an upper gastrointestinal endoscopy and tissue biopsies were performed. The specimens were found to be normal,

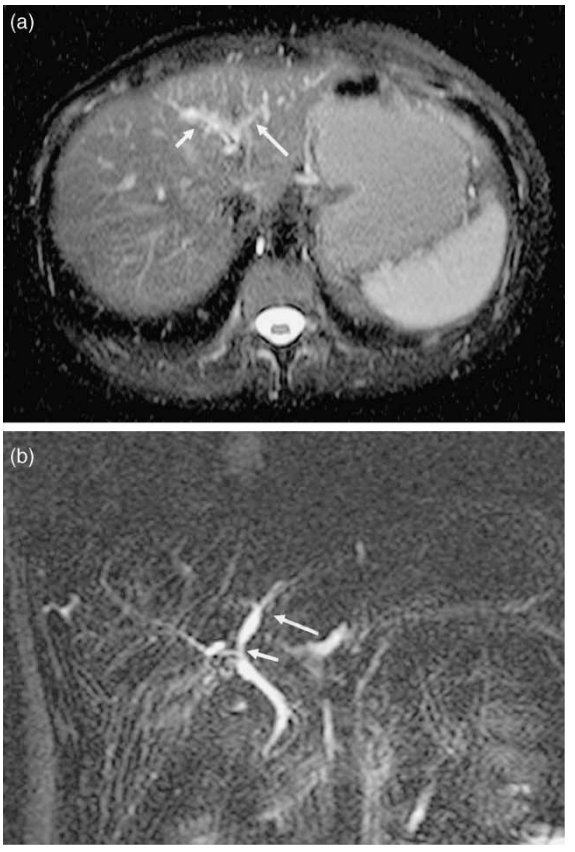

Figure 1. Magnetic resonance imaging (MRI) and magnetic resonance cholangiopancreatography (MRCP) of the liver. T2-weighted fat saturated image (a) reveals strictures and dilatations of the the biliary tree in the left hepatic lobe (*arrows*) secondary to past cholangitis; the MRCP image (b) shows the same findings.

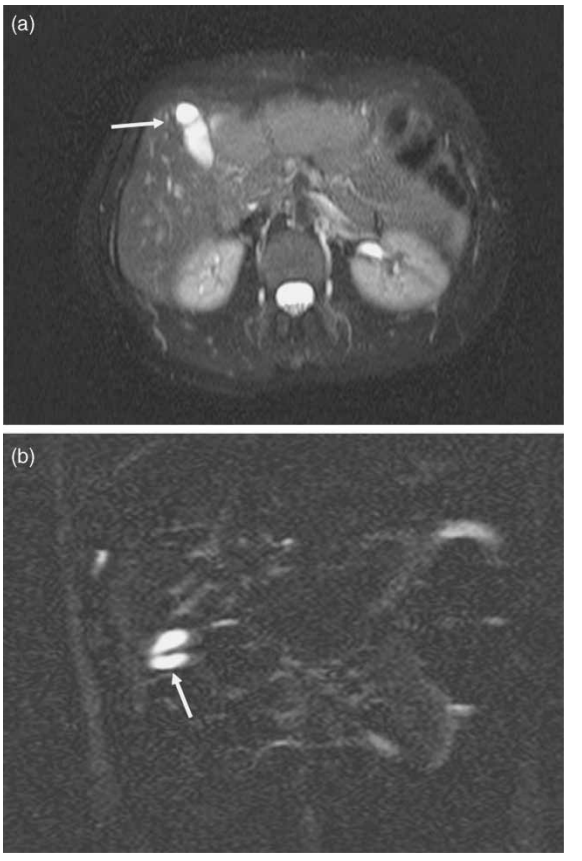

Figure 2. Magnetic resonance cholangiopancreatography (MRCP) of the same patient. On the axial T2 weighted fat saturated MRCP image (a), the fundus of the gallbladder reveals the Phrygian cap abnormality (*arrow*). The coronal MRCP image of the same patient (b) shows the same finding (*arrow*).

except for mild gastritis and *Helicobacter pylori* colonization. Culture and acid-fast staining of the gastric aspiration material were performed but no abnormality was found. An antibiotic therapy regimen comprising a combination of ciprofloxasin and amoxicillin-clavulonic acid was prescribed for 14 days. Her condition improved and the fever was unremarkable. However, marked tenderness in the RUQ and painful skin lesions were noted. Laparoscopic cholecystectomy was performed immediately. A Phrygian cap anomaly and concentrated sludge in the gallbladder and localized fibrosis of the liver capsule with normal liver tissue under the capsule were observed during the operation. Pathologic examination of the gallbladder revealed no abnormality such as tuberculosis, sarcoidosis or parasites, apart from chronic inflammation. Nor were any parasites or bacteria found on microbiological investigation of the sludge. Three months after the operation, the patient no longer complained of abdominal pain and fever. There were no new finger lesions and her condition had improved.

Discussion

RPC (cholangiohepatitis) is chronic inflammation of the bile ducts. Recurrent attacks of fever are the main clinical characteristics of patients with RPC. Nausea and abdominal pain, particularly in the RUQ, usually occur along with fever. The young woman presented in this paper experienced four cholangitis attacks. The first attack occurred when she was 20 years old, but it was misdiagnosed as pulmonary tuberculosis activation. Then, there was a silent term of 10 years. Another three attacks were observed within a short period (about one and half years). Finally, this case was diagnosed by upper abdominal MRI and MRCP, which were ordered because of persistent abdominal pain during the 4th attack. Interestingly, all blood examinations such as ESR, CRP, liver function tests and pancreatic enzymes, and imaging tests such as abdominal ultrasound and CT scanning were found to be normal during the attacks.

In the case of RPC, parasites and recently reported biliary sludge are usually found in the

biliary tracts. In our case, we also demonstrated biliary sludge in the gallbladder as a cause of these cholangitis attacks. Clinical conditions and events associated with the formation of biliary sludge include rapid weight loss, pregnancy, ceftriaxone therapy, octreotide therapy and bone marrow or solid organ transplantation [5–9]. In the case presented here, we found none of the associated conditions mentioned above. However, the patient had Phrygian cap anomaly of the gallbladder. A patient with this anomaly is prone to bile stasis and biliary sludge formation, as in our case [10–14]. This anomaly is the pseudoseptation of the gallbladder fundus due to kinking, or sometimes a true septum, and is found in 2–6% of all gallbladders.

Biliary sludge can appear, disappear and reappear. Its formation is a dynamic, reversible process. A partial blockage to the flow of bile may occur when sludge blocks any part of the biliary ductal system. This allows bacteria to flow back into the common bile duct and creates an ideal condition for growth of bacteria. If the common channel is obstructed by sludge, bile refluxes to both the pancreatic duct (pancreatitis attack) and the common bile duct (cholangitis attack), as in our case [15–18]. The recurrent character of the infection leads to progressive destruction of the biliary ductal system, thickening of the gallbladder wall without stones, hepatic abscesses and eventually biliary cirrhosis, hepatic atrophy and portal hypertension. Furthermore, patients with RPC have an increased risk for cholangiocarcinoma in up to 5% of patients. Thus, if a patient with biliary sludge develops symptoms or complications, as in our case, cholecystectomy should be considered as the definitive therapy. Cholecystectomy also protects against recurrent attacks of pancreatitis and cholangitis.

If a gallbladder or liver abnormality is suspected in practice, abdominal ultrasound is the first imaging modality of choice. Evaluation of the hepatobiliary abnormalities by CT scanning should be considered later. Abdominal CT scanning usually defines the areas of intra- and extrahepatic biliary dilatation more clearly than ultrasonography and provides three-dimensional information. However, both abdominal ultrasound and CT scans failed to show gallbladder and hepatobiliary pathologies in this case until a slight thickening of the gallbladder wall developed owing to the recurrent attacks. Gallbladder wall thickening without a gallstone is a hallmark of acalculous cholecystitis and biliary sludge [15,16]. The availability of MRI and MRCP techniques increased the chance to demonstrate biliary sludge and related complications such as cholangitis and pancreatitis, as in our case. The sensitivity of endoscopic ultrasonography to reveal biliary sludge is approximately 96%. However, this new tool is not available in all centres.

As mentioned above, abdominal ultrasound and CT scans failed to diagnose the Phyrigian cap and RPC in this case, although subsequent MRCP clearly demonstrated the Phyrigian cap and RPC that led to the correct diagnosis and appropriate treatment. A multislice CT scanner would have depicted the Phyrigian cap with the use of thinner slices and with its multiplane reformatting capability. The ultrasound examination failed to show the Phyrigian cap because it is an operator-dependent examination as compared with the objective MRCP examination. Ultrasound also missed the RPC, because this can only evaluate the dilatation of the biliary tree and not the inflammatory response of the biliary tree wall, which is best recognized by contrast enhancement. The delayed phase contrast enhanced imaging on MRI, as done in our imaging protocol, can depict the RPC. Abdominal CT scanning is known to be one of the best imaging modalities for RPC. However, the section thickness in our CT scan examination was not enough to depict the subtle contrast enhancement in the initial examination.

RPC most commonly affecting the left hepatic ducts and predominantly left-sided findings should prompt consideration of this diagnosis in patients from endemic areas. In our case, we also saw features of past cholangitis in the left lobe. However, we did not observe parasites, apart from biliary sludge.

Apart from bacteraemia caused by the recurrent cholangitis, we have no other explanation for the clinical picture defined in this paper. Tuberculosis, sarcoidosis, amyloidosis or parasites could not be a cause because all investigations such as blood analysis, imaging tests and tissue biopsies did not demonstrate any specific cause of that kind. Nor did rheumatologic investigations such as blood tests including rheumatology serology panel and tissue biopsies indicate any specific rheumatologic disease to explain the vasculitic skin changes (palmoplantar vasculitis).

In conclusion, upper abdomen MRI and MRCP should be the next imaging modalities in patients suspected of having hepatobiliary ductal system disease, even if both the abdominal ultrasonography and CT scan findings are normal.

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