Unusual association of diseases/symptoms

Membranous Budd–Chiari syndrome in a well-anticoagulated patient

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Summary

We describe an unusual case of a 54-year-old woman who was diagnosed with a subacute Budd–Chiari syndrome due to membranous venous obstruction in the inferior vena cava. The unusual feature of this case was that she had been diagnosed with pulmonary emboli a few years earlier and was on lifelong warfarin with a therapeutic international normalised ratio. She was effectively treated by venoplasty.

BACKGROUND

This is a very interesting presentation of Budd–Chiari syndrome (BCS). In a well-anticoagulated patient you would not expect venous thrombosis to be the cause of the symptoms. The patient was diagnosed a few years earlier with pulmonary embolism, which was proved to be due to antiphospholipid syndrome.

CASE PRESENTATION

A 54-year-old woman with a background history of pulmonary embolism and erythema nodosum was admitted with a 5-day history of pyrexial illness and right hypochondrial pain.

She did not smoke and drank alcohol only in moderation. The only medication she took was warfarin.

On examination, she was pyrexial and was found to be tender in the right hypochondrium. Her initial blood results showed a low platelet count of 99 (normal 140–440) but normal white cell count and haemoglobin. Liver function tests (LFTs) showed a slightly elevated bilirubin of 27 $\mu\text{mol/l}$ (normal 1–20), an increased alkaline phosphatase of 168 μl (normal 30–130), a raised γ -glutamyl transpeptidase (GGT) of 225 μl (normal 1–42) and also a raised alanine transaminase (ALT) of 260 μl (normal 1–41). Her international normalised ratio (INR) was significantly elevated at greater than 10. Her immunoglobulins, autoantibody screen and hepatitis B surface antigen and hepatitis C antibody were negative. Ferritin was significantly elevated at 988 $\mu\text{g/l}$ (normal 12–300) but iron studies were normal.

An ultrasound scan was arranged, which showed a normal liver, gallbladder and biliary tree but there was a trace of free-fluid in the pelvis.

During her hospital stay her pain worsened and radiated to her right shoulder. Her C reactive protein (CRP) and temperature increased so abdominal ultrasound was repeated. This showed mild thickening of the gall bladder wall and a trace of fluid in the gall bladder fossa. There was also thought to be some intrahepatic biliary dilatation and common bile duct dilatation. However, MR cholangiopancreatography (MRCP) examination did not show any duct dilatation.

As her symptoms continued to deteriorate a CT scan of the abdomen was done and this showed ascites and change in the liver architecture. There were multiple areas of low attenuation in the peripheral aspects of both hepatic lobes and appearances were suggestive of an inflammatory or infective process. Due to her continuing abdominal symptoms a laparotomy was performed, which showed a diffuse nodular liver and free-fluid within the abdomen. Liver biopsy was performed at the time of laparotomy and histology revealed widespread necrosis, inflammation and cirrhosis.

Her ascitic fluid serum ascites albumin gradient was more than 11, which indicated ascites due to portal hypertension.

Four months later, she was re-admitted with a 3-day recurrence of epigastric pain. She was found to be pyrexial but her abdomen was soft. Her platelet count was low at 106 (normal 140–440), and her LFTs were abnormal with bilirubin of 25 $\mu\text{mol/l}$ (normal 1–20), an increased alkaline phosphatase of 251 μl (normal 30–130), a raised GGT of 209 μl (normal 1–42) and a raised ALT of 53 μl (normal 1–41). Her INR was 3.3 and CRP was >120. A septic screen was performed and she was started on an intravenous antibiotic (cefuroxime) for a presumed diagnosis of cholangitis. Ultrasound of her abdomen showed only a small amount of free-fluid within the abdomen. CT abdomen showed multiple areas of low attenuation within the liver and appearance suggestive of a web in the inferior vena cava (IVC) (figure 1). BCS was suspected. This was confirmed on venogram and was treated with venoplasty. Thrombophilia screen established antiphospholipid syndrome as the underlying cause. She made remarkable recovery and she is doing well on her regular outpatient follow-up.

INVESTIGATIONS

Ultrasound of the abdomen, MRCP and CT scan of the abdomen. Ascitic tap and laparotomy with liver biopsy.

DIFFERENTIAL DIAGNOSIS

Metastasis, collagen disease, cirrhosis with primary spontaneous peritonitis and peritoneal tumour.

Figure 1 CT scan of the abdomen showing narrowing of the inferior vena cava.

TREATMENT

Venoplasty.

OUTCOME AND FOLLOW-UP

Repeated venoplasty and outpatient follow-up.

DISCUSSION

Membranous venous obstruction of the inferior vena cava is a curable cause of BCS but is very rare in Western Europe.^{1–2} To date, there is only very limited information on membranous BCS in the western world.^{3–4} The clinical presentation depends on the speed and the extent of the occlusion of the veins. In the fulminant form, the patients may die with encephalopathy within a couple of weeks whereas in the more chronic form the patients present with pain due to enlarged liver and ascites.^{5–7} Our patient did have right hypochondrial pain, ascites, as well as hepatomegaly.

We had some difficulty in the initial steps of the diagnostic process since the patient was already on warfarin and the INR was well above the therapeutic range and the ultrasound suggested cholecystitis and dilated common bile duct. This was compounded by the fact that the patient had pyrexia and laparotomy suggested heterogeneous liver

might be due to malignancy. It was not picked up on the CT scan that had been done on first admission. Data from Ciesek *et al* did not find the CT as a good modality in establishing the diagnosis membranous BCS.⁸

BCS can be classified into two types: primary hepatic vein thrombosis and primary IVC obstruction. The latter is commonly called membranous obstruction of the IVC (MOVC), because the thrombus organises into a fibrous and frequently membranous occlusion of the IVC. The hepatic vein orifices are affected to varying degrees resulting in congestive liver damage. The MOVC type appears to be insidious or chronic type. Our patient had an unknown aetiology for developing MOVC similar to other reported types of MOVC. MOVC may be a congenital disease but Okuda *et al* considered it more a consequence of organised thrombosis.⁹

Therapeutic modalities in MOVC include transcatheter membranotomy, venacaval-atrial bypass surgery, various shunt surgeries, balloon angioplasty and expandable stent into the occluded veins. Our patient did have angioplasty and this did improve her symptoms. Wu *et al* reported 97.6% of 41 patients successfully treated with percutaneous balloon angioplasty. They concluded that percutaneous balloon angioplasty is a safe and effective treatment of BCS caused by IVC obstruction.¹⁰

Lee *et al* found also that endovascular treatment provides excellent results and potentially halts parenchymal deterioration as the case in our patient.¹¹

In conclusion, we report subacute BCS in a well-anticoagulated patient, caused by MOVIC, which is a rare disease. BCS due to MOVIC although rare is an important cause of liver cirrhosis. A diagnosis should be suspected when there is right hypochondrial pain, abnormal LFTs, ascites and classical appearance on the CT scan or angiogram. This case illustrates the importance of recognition and treatment of a rare illness that can lead to excellent recovery.

Learning points

- ▶ Budd–Chiari syndrome due to membranous obstruction can occur in patients who are well anticoagulated.
- ▶ Venoplasty is an excellent treatment for this type of Budd–Chiari syndrome.
- ▶ Antiphospholipid syndrome as a cause should be thought of in patients with recurrent venous thromboembolism.
- ▶ Repeated thrombotic events may be suspected, which may lead to a cirrhotic state if there is no evidence of viral hepatitis infection.

Competing interests None.

Patient consent Obtained.

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