A woman with haemolytic anaemia

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CASE REPORT

A 73-year-old woman was admitted to the outpatient clinic of the Department of Internal Medicine for analysis of an anaemia.

For a few months she had been experiencing progressive fatigue and shortness of breath. During this period she also noticed peripheral oedema and had lost her appetite because of upper abdominal distension. Her medical history revealed atrial fibrillation and recurrent epistaxis. The patient's medication consisted of atenolol, digoxin, acenocoumarol and bumetanide. On physical examination her blood pressure was 140/55 mmHg, heart rate 50 beats/min, and the jugular venous pressure was elevated. There was a systolic murmur which was maximal at the third left intercostal space and a thrill was heard at the right upper abdominal quadrant with murmurs across the abdomen. Further examination of the abdomen showed hepatomegaly and she had pitting oedema up to the ankles. Laboratory tests showed a haemoglobin concentration of 6.1 mmol/l, MCV 81, leucocytes 4.6 x 109/l, thrombocytes 118 x 10⁹/l, LDH 627 U/l, haptoglobin 0.1 g/l, red blood cell fragmentation in the blood smear, Coombs direct and indirect negative, ferritin 25 µg/l, no signs of diffuse intravascular coagulation, creatinine 66 μ mol/l and bilirubin 47 μ mol/l. So the diagnosis of mechanical haemolytic anaemia was

made. On chest X-ray cardiomegaly with pronunciation of the hilar and pulmonary vessels was seen.

The echocardiography showed elevated right ventricular pressure and a severe tricuspid valve regurgitation. The vena cava inferior did not collapse with inspiration. The left ventricular function was intermediate. Liver veins were enlarged. Because of the unexpected finding of enlarged liver veins on echocardiography, computed tomography (CT) and subsequently an angiography of the abdomen were performed (*figures 1-3*).

WHAT IS YOUR DIAGNOSIS?

See page 261 for the answer to this photo quiz.

Figure 1. Abdominal CT scan with intravenous contrast, enlarged liver veins filled with contrast



Figure 2. Aneurysm of hepatic artery

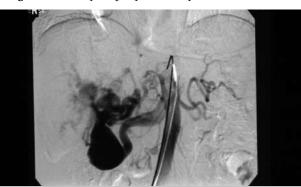
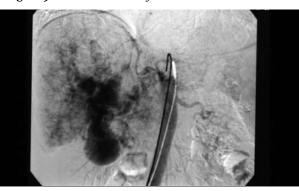


Figure 3. Arteriovenous malformation



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ANSWER TO PHOTO QUIZ (ON PAGE 259) A WOMAN WITH HAEMOLYTIC ANAEMIA

DIAGNOSIS

The CT scan of the abdomen showed distended veins in the portal system (*figure 1*). An angiography of the abdominal arteries showed arteriovenous malformations of the hepatic artery and mesenteric artery. There was a large aneurysm at the shunting hepatic artery (*figure 2* and 3). These vascular malformations are characteristic of Rendu-Osler-Weber disease.

Therefore, the diagnosis of Rendu-Osler-Weber disease with mesenteric arteriovenous fistula leading to severe high-output cardiac failure and secondary pulmonary hypertension was made.

The hypothesis for the cause of the haemolytic anaemia is turbulent blood flow. Turbulent flow is definitely present in arteriovenous malformation and possibly in tricuspid regurgitation. Recently, a case report described mechanical haemolytic anaemia in a haemodialysis patient with carotid-jugular arteriovenous fistula.¹

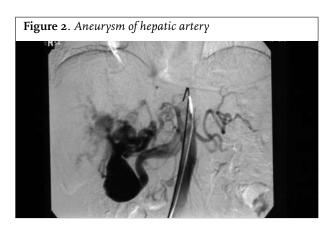
Tricuspid regurgitation has never been described as a factor for haemolytic anaemia, only an artificial tricuspid valve can be a cause for haemolysis.

There are a few therapeutic options to improve arteriovenous malformations. The treatment of choice is embolisation of the malformation. If embolisation fails or can not be carried out, surgery can be performed.

REFERENCE

 Kuo KL, Chou YH, Tarng DC. Mechanic haemolysis in a hemodialysis patient with carotid-jugular arteriovenous fistula. Clin Nephrol 2004;61:74-7. Figure 1. Abdominal CT scan with intravenous contrast, enlarged liver veins filled with contrast







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