Giant aneurysm of the main hepatic artery secondary to hereditary hemorrhagic telangiectasia:
3D contrast-enhanced MR angiography features

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SUMMARY

This case report describes a patient with Hereditary Hemorrhagic Telangiectasia and a giant hepatic artery aneurysm. Aneurysms of the hepatic artery are the second most common form of visceral artery aneurysms. The causes of hepatic artery aneurysms are atherosclerosis in 30% of cases, arteriitides, periarterial inflammation, liver transplantation, and hepatic tumor embolization. To our knowledge no giant hepatic artery aneurysm has been described in relation to Weber-Rendu-Osler disease in the literature. These aneurysms probably develop because of hepatic arterio-venous fistulas and secondary to changes in arterial hemodynamics. The increased use of non invasive imaging techniques such as MRI before liver transplantation in patients with hereditary hemorrhagic telangiectasia reveals these asymptomatic aneurysms and makes it possible to choose the best therapeutic approach.

RÉSUMÉ

Anévrisme géant de l’artère hépatique commune dans le cadre d’une maladie de Weber-Rendu-Osler : aspect en angio-IRM

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Nous présentons un cas d’anévrisme géant de l’artère hépatique compliquant une maladie de Rendu-Osler. Les anévrismes héréditaires constituent la seconde localisation des anévrismes viscéraux. Ils sont d’origine athéroscléreuse dans 30% des cas ou secondaire à une artériite, à une inflammation périvasculaire, à une transplantation hépatique ou à une embolisation tumorale. Aucun cas d’anévrisme géant n’a été décrit dans le contexte de maladie de Rendu-Osler. L’hyperdébit secondaire aux fistules artério-sushépatiques explique la formation de telles lésions. L’utilisation de technique d’exploration vasculaire non invasive comme l’IRM, dans l’exploration des maladies de Rendu-Osler, en particulier avant greffe hépatique, permet de les découvrir et de choisir les meilleures options thérapeutiques.

Introduction

Hereditary hemorrhagic telangiectasia is a rare autosomic dominant disease, with vascular dysplasia as an elementary lesion [1]. Hepatic involvement occurs in 8 to 31%, of patients and includes telangiectases, arteriovenous shunting, hepatic arterial dilatation, focal liver lesions, and ischemic cholangitis. 3D Contrast-Enhanced Magnetic Resonance Angiography (3D CE-MRA) has been shown to be effective in diagnosing vascular diseases [2]. In this case report, we describe a giant aneurysm of the main hepatic artery in a patient with hereditary hemorrhagic telangiectasia, confirmed by 3D CE-MRA, and discuss the role of magnetic resonance imaging (MRI) in determining the best therapeutic option in these cases.

Case report

A 64-year-old woman with hereditary hemorrhagic telangiectasia with a diagnosis based on clinical and familial features according to the Curacao criteria [3] presented with paralysis of the lower left leg, and signs of cardiac failure secondary to atrial fibrillation. Laboratory analysis demonstrated cholestasis with serum bilirubin levels of 25 mmol/L and serum alkaline phosphatase 220 U/L. Follow up ultrasound revealed liver involvement with an enlarged main hepatic artery, telangiectases and intrahepatic fistulas without bile duct dilatation. One Step liver MRI combined MRCP and 3D CE-MRA was performed on a 1.5T clinical unit (Symphony Quantum, Siemens, Erlangen, Germany) with a body phased array coil for further evaluation of the hepatic vasculature, especially the hepatic artery, parenchyma and biliary tract. The examination protocol included the following sequences.

Parenchymal sequences with transverse in-phase and out of phase T1-weighted breath-hold spoiled gradient-echo sequences of the upper abdomen (time repetition time (TR)/ time echo (TE), 167/2.38, 4.76 msec; flip angle, 70; section thickness of 8 mm; 10% intersection gap; matrix size of 111x156), transverse dual echo T2-weighted fat-suppressed turbo spin-echo sequence (TR/TE, 3 820/88-160 msec; section thickness of 8 mm; 20% intersection gap; matrix size of 197x512).

Biliary dedicated sequences with an Half-Fourier Single-shot Turbo spin Echo T2-weighted sequence in the transverse plane (TR/TE, 183/∞ msec, section thickness of 7 mm and a matrix size of 176x512), coronal and coronal oblique Rapid Acquisition Relaxation Enhancement (TR/TE, 2390/∞ msec, section thickness of 20 mm and a matrix size of 242x512).

Contrast Enhanced MR angiography (MRA) was performed with a dual phase protocol to visualize the arterio-venous liver vasculature. A 3D spoiled gradient echo sequence was used with an intermittent fat saturation pulse, performed in the coronal plane. Imaging parameters included: TR/TE 5.2/2 msec; 35° flip angle; 192x512 matrix; 400 mm mean field of view; 100 mm slab thickness; 64 partitions; 1.79 mm slice thickness; and 25 sec acquisition time. Gadopentate dimeglumine (0.2 mmol/kg of body weight) was intravenously administered with a power injector, immediately followed by 20 mL of normal saline solution. The imaging protocol was completed with one contiguous data acquisition (venous phase), with 12 seconds delay between 2 measurements.

MR angiograms were post processed with a series of maximum intensity projection (MIP) images and/or Volume Rendering Texture artefact (VRT) created by using the entire acquired volume (Leonardo, software version 1; Siemens Medical Systems).

A 4 cm in diameter fusiform aneurysm of the main hepatic artery, arising from the celiac trunk (figure 1) was identified in the MRA images, associated with an enlarged segment of the initial portion of the hepatic artery. The Arterial phase demonstrated early enhancement of venous structures suggesting right arteriohepatic vein fistula (figure 2) and right arterioporal fistula (figure 3). Biliary sequences revealed biliary duct strictures and upstream dilatation suggesting ischemic cholangitis.

Due to cardiac failure, biliary involvement and the presence of a giant arterial aneurysm, the patient was treated with liver transplantation. Transplantation was performed with special care because of the high risk of hemorrhage secondary to the aneurysm. The aneurysm was visualized
by MRI so that the surgeon could plan the dissection and exclusion of the main hepatic artery. The common hepatic artery was tied first. Because the common hepatic artery was enlarged (10 mm), arterial anastomosis was performed between the recipient common hepatic artery and a celiac patch of the donor artery. Pathological examination of the explant confirmed MRI findings. There were no complications and the patient is symptom free.

Discussion

Hereditary hemorrhagic telangiectasia is a rare autosomic dominant disease with includes abnormal vascular architecture secondary to genetic mutations. Hepatic involvement occurs in 8 to 31% of cases and involves a vascular, parenchymal and biliary pattern with characteristic lesions such as telangiectases and arterioportal or arteriovenous shunts [1]. Ultrasound [4] and Multi-row Detector Computed Tomography (MDCT) with angiographic reconstruction are used to explore the abdominal vasculature, and screen vascular abnormalities in these cases [5], while conventional angiography is reserved for interventional procedures [6].

Aneurysms of the celiac vasculature have been described in hereditary hemorrhagic telangiectasia by ultrasound and on conventional angiography and occur due to the high liver output secondary to intrahepatic shunts [7-9].

Fig. 1 – a) 3D CE-MRA sequences with MIP reconstruction on the arterial phase in the sagittal plane show the large aneurysm of the main hepatic artery, arising from the celiac trunk (arrow); b) 3D CE-MRA sequences with MIP reconstruction on the arterial phase in the coronal plane show the largest part of the aneurysm of the main hepatic artery (arrow) and the right and left branches of the hepatic artery (arrowhead).

a) Séquence d’ARM 3D à la phase artérielle avec reconstruction MIP dans le plan sagittal montrant un anévrysme géant de l’artère hépatique commune (flèche) ; b) Séquence d’ARM 3D à la phase artérielle avec reconstruction MIP dans le plan coronal montrant la partie la plus large de l’anévrysme étendu à la bifurcation de l’artère hépatique (tête de flèche).

Fig. 2 – 3D CE-MRA sequences with MIP reconstruction on the arterial phase in the coronal plane show early enhancement of the right hepatic vein (arrow) suggesting arterio to hepatic vein fistula. The distal part of the aneurysm is also visible (arrowhead).

Séquence d’ARM 3D à la phase artérielle avec reconstruction MIP dans le plan coronal montrant le rehaussement précoce de la veine sus-hépatique droite (flèche), suggérant une fistule artério-sus-hépatique. La partie distale de l’anévrysme est visible (tête de flèche).

Fig. 3 – 3D CE-MRA sequences with MIP reconstruction on the arterial phase in the coronal plane show early enhancement of the right portal vein (arrow) suggesting arterioportal fistula.

Séquence d’ARM 3D à la phase artérielle avec reconstruction MIP dans le plan coronal montrant le rehaussement précoce de la branche portale droite (flèche) suggérant une fistule artérioportale.
Our case report emphasizes the specific problems encountered in the management of patients with severe involvement in hereditary hemorrhagic telangiectasia. The main difficulty is choosing the best therapeutic option especially if there are life-threatening complications, such as high-output cardiac failure secondary to arterial to hepatic shunts and/or arterial aneurysms. In case of cardiac involvement, the therapeutic options are medical (beta-blockers, digitalis and diuretics) [10], radiological (embolization of the hepatic artery) or surgical (ligation of the hepatic artery) [11, 12]. However, the best treatment for cardiac failure in hereditary hemorrhagic telangiectasia is liver transplantation [13].

To our knowledge a giant aneurysm associated with cardiac involvement has never been presented. The different options were medical treatment of the cardiac involvement and a radiological procedure such as hepatic embolization, or liver transplantation. Embolization was dangerous because of the risk of secondary repermeabilization of the aneurysm [11] and death from spontaneous rupture [14].

One step MRI helped our team choose liver transplantation. MRI provided a perfect image, showing segments of the hepatic artery free from the aneurysm, allowing anastomosis. Moreover, biliary sequences showed signs of ischemic cholangitis, which would have been worsened by embolization. Numerous arterial to hepatic vein and arterioportal shunts were also identified, which have not been treated by any other procedure. MRI showed the degree of liver involvement, guiding surgical treatment, which included a liver graft [13].

An aneurysm of the hepatic artery is a life-threatening complication in case of liver involvement in hereditary hemorrhagic telangiectasia. Because the aneurysm is not isolated and is a secondary complication of intrahepatic fistulas, conventional endovascular or surgical treatment is not effective. MRI with 3D CE-MRA sequences shows the complex pattern of liver involvement of hereditary hemorrhagic telangiectasia and should be performed in these cases to help choose the best therapeutic option.

REFERENCES