

Management and treatment of decompensated hepatic fibrosis and severe refractory *Schistosoma mansoni* ascites with transjugular intrahepatic portosystemic shunt

Maria Cristina Carvalho do Espírito Santo¹, Ronaldo Cesar Borges Gryschek^{1,2,3}, Alberto Queiroz Farias⁴, Wellington Andraus⁴, Noêmia Barbosa Carvalho¹, Olavo Henrique Munhoz Leite¹, Felipe Corrêa Castro¹, Giovanni Guido Cerri⁵, Gustavo Henrique Hypóliti⁵, Francisco César Carnevale⁵, André Moreira de Assis⁵

ABSTRACT

This study aimed to report the first case of a patient with hepatosplenic schistosomiasis mansoni, refractory ascites and portal vein thrombosis treated with a transjugular intrahepatic portosystemic shunt (TIPS), at the Instituto de Radiologia, Hospital das Clínicas, Faculdade de Medicina, Universidade de São Paulo, Brazil. After the procedure, the patient recovered favorably and progressed with portal pressure reduction and no deterioration of the liver function. Endovascular shunt modification is a conservative medical approach that often helps in reducing symptoms significantly, making it a less invasive and a safer alternative to liver transplantation for the treatment of schistosomiasis with portal hypertension.

KEYWORDS: *Schistosoma mansoni*. Refractory ascites. Transjugular intrahepatic portosystemic shunt.

INTRODUCTION

Schistosomiasis is a water-borne parasitic infection caused by a trematode of the genus *Schistosoma*. The only species of medical and epidemiological importance in Brazil is *Schistosoma mansoni*. The Brazilian Ministry of Health estimates an average prevalence of seven million infected individuals, with most cases concentrated in Minas Gerais State (14°13'58" e 22°54'00" from South latitude) and the Northeastern States (18°20'07" from South latitude)¹.

Patients with the hepatosplenic form of MS develop non-cirrhotic portal hypertension, which, in advanced stages, may decompensate leading to gastrointestinal bleeding and ascites, thereby compromising the liver function, survival and the carriers' quality of life².

This article aimed to present a case of hepatosplenic schistosomiasis complicated by portal thrombosis and refractory ascites, which was successfully treated by transjugular intrahepatic portosystemic shunt (TIPS) performed at the Instituto de Radiologia, Hospital das Clínicas, Faculdade de Medicina, Universidade de São Paulo, Brazil.

Clinical case description

The patient was a 41-year-old man with schizoaffective disorder, type 2 diabetes

¹Universidade de São Paulo, Faculdade de Medicina, Departamento de Moléstias Infecciosas e Parasitárias, São Paulo, São Paulo, Brazil

²Universidade de São Paulo, Faculdade de Medicina, Hospital das Clínicas, Laboratório de Pesquisas Clínicas da Imunopatologia da Esquistossomose (LIM-06), São Paulo, São Paulo, Brazil

³Universidade de São Paulo, Faculdade de Medicina, Instituto de Medicina Tropical de São Paulo, São Paulo, São Paulo, Brazil

⁴Universidade de São Paulo, Faculdade de Medicina, Departamento de Gastroenterologia, São Paulo, São Paulo, Brazil

⁵Universidade de São Paulo, Faculdade de Medicina, Hospital das Clínicas, Instituto de Radiologia, São Paulo, São Paulo, Brazil

Correspondence to: Maria Cristina Carvalho do Espírito Santo
Universidade de São Paulo, Faculdade de Medicina, Instituto de Medicina Tropical de São Paulo, Av. Dr. Enéas de Carvalho Aguiar, 470, Cerqueira César, CEP 05403-907, São Paulo, SP, Brazil
Tel: +55 11 30618220, +55 11 30618220

E-mail: cristinasanto@usp.br

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mellitus, dyslipidemia, and grade I obesity [body mass index (BMI): 30.9 kg/m²]. He was a smoker (40 pack-year) who was born and raised in the city of Sao Paulo, Sao Paulo State, Brazil. At the age of three, he had traveled to the small town of Antonio Goncalves, located in the North-central region of Bahia State, in the Northeastern region of Brazil, where he stayed for 30 days and had contact with water reservoirs possibly contaminated with *S. mansoni*. The patient was referred with positive detection of *S. mansoni* by parasitological methods, he was treated with praziquantel and progressed with the diagnosis of hepatosplenic schistosomiasis during an irregular follow-up. A history of a previous episode of melena was recorded in October 2010. On examination, hepatosplenomegaly was detected with a palpable spleen, 10 cm from the left costal margin and evidence of collateral circulation. Serological tests for viral hepatitis and HIV were negative, and the Kato-Katz stool test was negative for *S. mansoni* eggs. Although the parasitological tests were negative, the abdominal ultrasound imaging showed a F-pattern fibrosis compatible with schistosomal fibrosis and a significantly advanced central and peripheral periportal thickening³ (Figure 1). The treatment was initiated with propranolol and omeprazole and serial upper digestive endoscopies were performed.

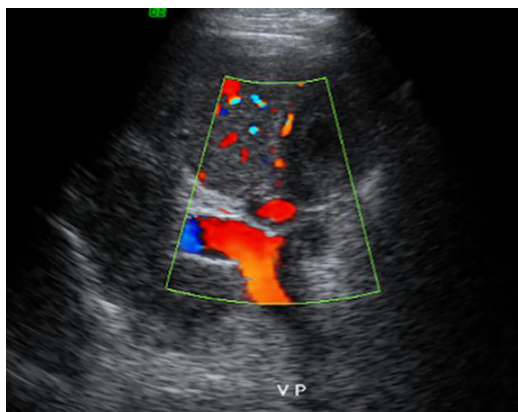


Figure 1 - Abdominal ultrasound showing an enlargement of the portal vein trunk and parietal irregularities due to partial thrombosis (yellow arrow) and periportal hyper echogenicity compatible with periportal fibrosis (blue arrow)

The patient sought medical care in July 2020 presenting with ascites and weight gain of approximately 35 kg. He was hospitalized, given diuretic therapy and underwent two paracenteses with removal of a total of 18.5 liters of fluid, along with adequate albumin replacement.

The patient was discharged after 14 days; the doppler ultrasound of the portal system performed in August 2020 revealed chronic thrombosis with partial recanalization of the portal vein. During an outpatient visit in September 2020, despite the treatment with 160 mg/day of furosemide

and spironolactone 200 mg/day, the patient progressed with significant ascites and gained 11 kg. The patient was hospitalized again in October 2020. On October 14, 2020, the patient underwent TIPS of the middle hepatic vein and also of the left branch of the portal vein. He exhibited thrombosis of the right branch of the portal vein, with signs of cavernous transformation. A Viatorr 10 × 80 mm was used, open to 9 mm (the initial portosystemic gradient was 19 mm Hg and the final portosystemic gradient was 8 mm Hg).

MATERIALS AND METHODS

Technical description of the TIPS procedure

The initial investigation of the hepatic vascular system using doppler ultrasonography of the abdomen showed signs of chronic liver disease and periportal fibrosis along with an increased caliber of the portal vein trunk, partial thrombosis and voluminous splenomegaly (26 cm in the longitudinal axis; average reference value < 13 cm), with no other significant findings.

The patient scored seven points on the Child-Pugh scale and 11 points on the Model for End-Stage Liver Disease with added sodium (MELD-Na) scale. He has also exhibited thrombocytopenia with a platelet count of 51,000/mm³. Although the renal function was preserved, diuretics did not reduce the ascites.

The procedure started with an invasive measurement of the pulmonary pressure under general anesthesia, which showed normal pulmonary artery pressure (PAP, 24 mmHg). After a relief paracentesis and albumin replacement, a guided puncture of the right internal jugular vein was performed using a RUPS 100kit (Cook Medical, Bloomington, Indiana, USA). Indirect portography performed via the right femoral artery revealed a patent portal vein trunk and chronic thrombosis of the right portal branch with intrahepatic portal collateralization. Initial pressure measurements found a severe portal hypertension with a portal pressure of 27 mmHg and a portosystemic gradient of 19 mmHg.

The catheterization of the middle hepatic vein was chosen because of the more favorable angulation to reach the left branch of the portal vein so that a communication between both veins was created. After a successful catheterization of the portal vein trunk, dilation of the parenchymal pathway was performed using an angioplasty balloon, and a 10 × 80 mm Viatorr stent-graft (Gore Medical, Flagstaff, Arizona, USA) was positioned. After calibration of the shunt with an angioplasty balloon (9 mm diameter), a final portosystemic gradient of 8 mmHg was

observed. The last direct portography showed adequate stent positioning and the reestablishment of normal hepatopetal blood flow through the portal system (Figure 2).

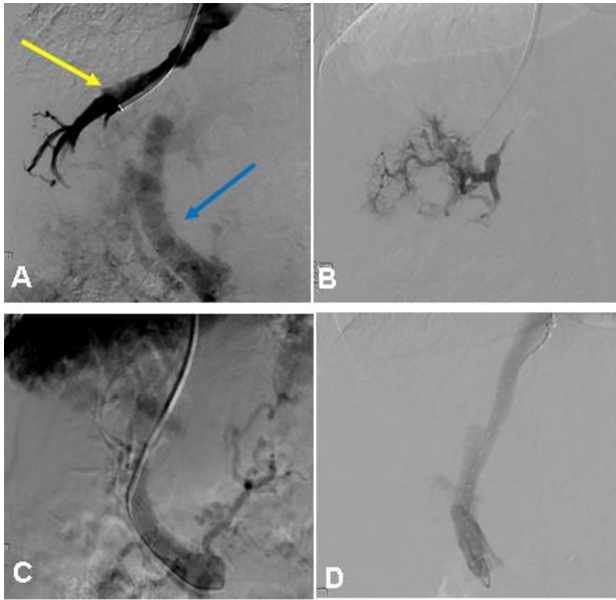


Figure 2 - A) Simultaneous angiographies of the right hepatic vein and superior mesenteric artery showing the patency of the portal vein trunk (blue arrow) and its relationship with the right hepatic vein (yellow arrow); B) Periportal varices after puncture of the right branch of the portal vein; C) Catheterization of the left branch of the portal vein through the left hepatic vein; D) Final appearance of TIPS.

RESULTS

The patient was hospitalized for seven days. He spent the first 24 h in intensive care for monitoring, without deterioration of liver or kidney functions or signs of hepatic encephalopathy. An abdominal doppler ultrasonography was performed before discharge, which showed patency of the TIPS and normalization of the hepatopetal portal flow.

The final results of the transjugular intrahepatic portosystemic shunt procedure performed in the patient are shown in the sequence of images in Figure 2.

DISCUSSION

To the best of our knowledge, this is the first case of TIPS performed in a patient with decompensated hepatosplenic schistosomiasis with refractory ascites and portal vein thrombosis. The patient remained compensated and stable for nine years on medications and endoscopic measures. Even when he developed refractory ascites, he did not have any episodes of gastrointestinal bleeding.

Some investigators have argued that TIPS should not be used in schistosomiasis because the hepatic function is

normal. Procedures that reduce the pressure in the portal system can reduce the liver perfusion and cause liver failure⁴.

However, in schistosomiasis, the portal hyperflow related to hypersplenism contributes to the portal hypertension to a greater extent than in cirrhosis, so the reduction in liver perfusion can be expected to be less relevant⁵. Hepatic encephalopathy should therefore not be expected more frequently in schistosomiasis patients than in those with cirrhosis attributed to other diseases, a finding that has also been confirmed in other case reports⁶. Thus, the risk of encephalopathy after the shunt surgery is exceptionally high in patients with schistosomiasis⁷.

Liu *et al.*⁷, in a retrospective investigation of a cohort of 82 patients with portal hypertension due to schistosomiasis (n = 20) and hepatitis B virus infection (n = 62) reported that the incidence of hepatic encephalopathy following TIPS was similar in both groups (schistosomiasis vs hepatitis B, 25.0% vs. 22.6%, P = 0.68). Other relevant outcomes were also comparable: P = 0.84 for variceal bleeding and P = 0.43 for survival. These results support the indication of TIPS in this case.

In hepatosplenic schistosomiasis, portal hypertension is multifactorial and is partly due to the splenic hyperflow. However, there is a tendency to underestimate this splenic hyperflow in schistosomiasis with portal hypertension⁵, reinforcing the justification of using TIPS, even in schistosomiasis.

The reduction in portal pressure promoted by TIPS also improves the systemic cardiovascular function. However, it increases the renal blood flow and the glomerular filtration rate, resulting in enhanced refractory ascites⁶.

Furthermore, unlike most surgical procedures, the immune function of the spleen is preserved^{8,9}. In fact, patients who undergo splenectomy to treat portal hypertension seem to present with a much higher risk of portal vein thrombosis than patients with TIPS (HR = 19, P < 0.001)⁷.

As liver function is preserved even in advanced schistosomiasis with fibrosis, liver transplantation is rarely indicated in schistosomiasis unless complications such as advanced liver cirrhosis or hepatocellular carcinoma of another etiology, such as chronic active viral hepatitis are present⁹.

Due to the shortage of organs for transplantation in Brazil, there is an initial trend towards an increased application of TIPS in patients with portal hypertension¹⁰. Thus, TIPS is an alternative to surgery in patients with refractory schistosomiasis with liver fibrosis and should be further evaluated in centers experienced in this technique¹¹.

This case presents a reversible and much less invasive alternative offered during the pandemic for a patient

who had already had a partially re-canalized portal vein thrombosis. As the right branch of the portal vein was entirely occluded, the left branch was used for the TIPS creation.

Grade I encephalopathy was controlled using lactulose (667 mg/mL, 20 mL, three times a day) combined with dietary measures such as the replacement of meat protein (commonly consumed by the patient) with vegetable protein present in a vegetarian diet¹². The impact of this management was three daily evacuations. The patient progressed with total regression of the edema and maintained hepatic and cardiovascular functions for four months, with only one episode of sleep-wakefulness alteration observed.

The reduction in the number of visits to perform paracentesis in the outpatient clinic and home isolation decreased the possibility of infection by SARS-CoV-2 or other microorganisms that could have impacted the excellent prognosis, self-esteem and current quality of life of the patient.

An endovascular shunt modification with a conservative medical approach often helps to achieve a significant improvement of symptoms. Six months after the procedure, the abdominal doppler ultrasound showed the TIPS permeability and the absence of ascites. Reflections on this clinical case during the COVID-19 pandemic may be helpful in designing future protocols for the management of schistosomiasis with portal hypertension.

CONCLUSION

The case presented here suggests that TIPS can be used in patients with the hepatosplenic form of schistosomiasis, being beneficial for the patient, making it a less invasive and a safer alternative to liver transplantation for the treatment of schistosomiasis with portal hypertension.

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CONFLICT OF INTERESTS

The authors have declared that there is no conflict of interests.

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