# Intrahepatic Portal Vein Aneurysm: A Case Report of an Uncanny Late Complication of Liver Transplantation

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## ABSTRACT

**Background.** Whereas chronic rejection, opportunistic infections, post-transplant lymphoproliferative disorder, hemophagocytic lymphohistiocytosis, biliary tract issues, and cardiovascular side effects of immunosuppressive drugs are frequently reported as late complications in liver transplanted patients, intrahepatic portal venous aneurysm following liver transplantation remains exceptional and unusual.

**Case report.** We report the case of a 25-year-old man who underwent a liver transplantation in 1997 because he had glycogen storage disease type 4. The patient developed a late postoperative complication, an intrahepatic portal aneurysm, and 2 episodes of acute cholangitis.

**Discussion.** By reviewing and scoping the literature, we tried to spotlight the best therapeutic attitude concerning the management of this rare complication.

PORTAL venous aneurysms represent 3% of all venous aneurysms. Intrahepatic portal aneurysms are defined by a vein diameter larger than 9 mm [1]. According to López-Machado et al [2], over a cohort of 13 cases of portal vein aneurysms, 10 were extrahepatic and 3 were intrahepatic (23% of all portal vein aneurysms).

Concerning the etiology of intrahepatic portal venous aneurysms, they can be either congenital or acquired. Among the acquired causes, we can mention portal hypertension, surgery, infections (such as schistosomiasis), hematological conditions (such as thalassemia), and pancreatitis. Among the congenital etiologies, we can cite the abnormal development of the portal vein during the embryonic life. Furthermore, aneurysms can have different configurations: they can be either saccular or fusiform [1,3,4].

The clinical presentation depends on the size of the aneurysm. Small aneurysms can be asymptomatic and can be discovered incidentally, whereas bigger aneurysms can be symptomatic, causing abdominal pain, vomiting, jaundice [3], biliary tract obstruction (and infections); a compression of adjacent anatomic structures could also lead to portal vein thrombosis, and even to portal hypertension in that condition.

## CASE REPORT

We present the case of a 25-year-old patient who had been hospitalized for a medical checkup in because of repetitive cholangitis episodes.

In 1997, after 4 months of waiting on the list, the patient underwent a whole liver transplantation from a 21-month-old male brain-dead donor, with the piggyback technique and with a 50-cm-long Roux-en-Y operation, because of a glycogen storage disease type 4 that has led to early cirrhosis and weight loss (BMI 16.2). The early post-transplant follow-up has been marked by a rejection, with a favorable evolution, and a lymphoproliferative disorder due to an Epstein Barr virus primo-infection with spontaneous recovery after immunosuppression reduction.

In 1998, 1 year after transplantation, elevated gamma glutamyl transferase (3 times the upper limit normal [ULN]) were observed, left hepatic vein thrombosis was detected on an ultrasound, associated to a left biliary duct stenosis, justifying left hepatic lobectomy. The patient subsequently presented median hepatic vein thrombosis requiring heparin therapy followed by a 3-month treatment with low molecular weight heparin with good evolution.

The patient presented repetitive episodes of cholangitis, supported by a long-term antibiotic therapy with cefuroxime that was eventually stopped in 2015.

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0041-1345/20
https://doi.org/10.1016/j.transproceed.2022.05.013

**Table 1. Measurements Profile of Portal Vein Ectasia in Recent Years on Doppler Ultrasound Imaging**

| Year | Diameter of the Ectasia (mm) |
|---|---|
| 2005 | 30 |
| 2006 | 25 |
| 2011 | 25 |
| 2012 | 30 |
| 2013 | 35 |
| 2014 | 35 |
| 2015 | 33 |
| 2016 | 38 |
| 2017 | 41 |
| 2019 | 35 |
| 2021 | 35 |

As part of the transplant follow-up, many abdominal ultrasound examinations were carried out each year.

In 2001, portal vein ectasia was detected on ultrasound for the first time 3 mm above the anastomosis and measuring 15 to 20 mm, without any consequences regarding the segmental vascularization (Table 1).

We noticed that there was a moderate increase in the diameter of the ectasia from 2001 to 2012, before a stabilization of the size of the aneurysm between 2013 to 2021. It should be noted that portal flow and hemodynamic circulation beyond the aneurysm was not affected (Table 1).

However, in 2016, we reported a dilation of 6 mm of the biliary duct in segment IV, possibly due to the ectasia.

An abdominal cholangio-MRI and an entero-endoscopic retrograde cholangiography were performed to better understand the reason of the intrahepatic biliary duct dilation. The examinations have shown a stenosis of the hepaticojejunal anastomosis (Fig 1), associated with a dilation of the intrahepatic biliary duct (Fig 2). The cholangiogram showed a pinpoint stricture of the hepaticojejunal anastomosis that was treated with dilation and a stenting (Fig 3). The aneurysmatic dilation of the portal vein could therefore have a mass effect on the extra and intrahepatic biliary tree. From a biological point of view, the patient showed elevated gamma glutamyl transferase (3-4 times the ULN) and alkaline phosphatases (2 times the ULN) with resolution after dilation and stenting of the bile duct.

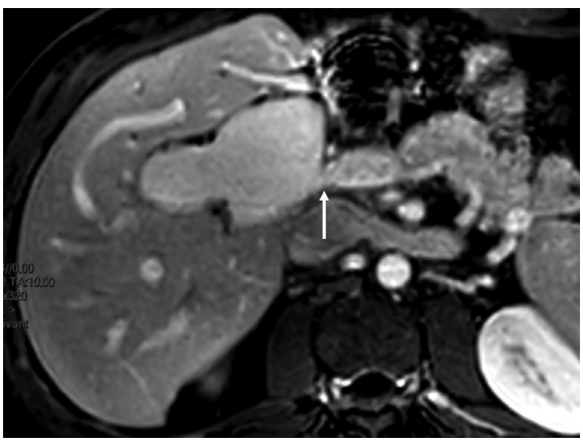

**Fig 1.** T1 magnetic resonance imaging sequence after gadolinium injection, showing the aneurysmal dilation of the upper-anastomotic portal vein. Notice the presence of local stenosis of the portal anastomosis (arrow).

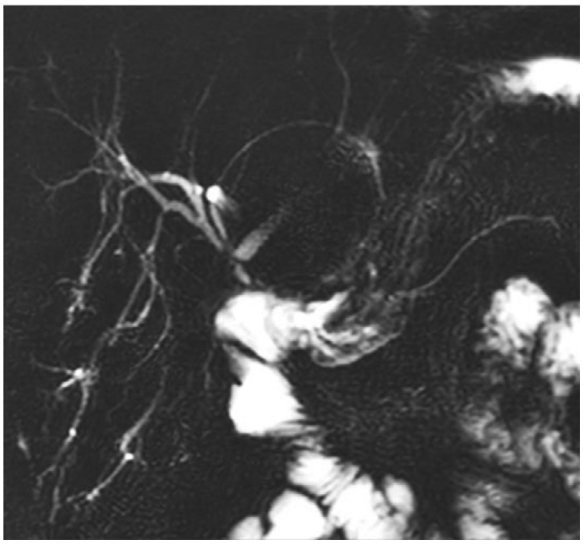

**Fig 2.** Magnetic resonance imaging cholangiography showing a dilatation of the right biliary.

Given that this portal vein dilation was stable with no vascular side effects, we decided to continue a standard follow-up.

## DISCUSSION

Portal vein aneurysms remain a rare entity, and only few publications provide information concerning the therapeutic management.

In our patients, the portal vein aneurysm is probably due to the surgery 1 year post-transplantation and the thrombotic complication leading to focal portal hypertension.

According to Sfyroeras et al [5], most of the portal vein aneurysms require no specific treatment, but a frequent follow-up (by ultrasound imaging), and supportive care for the possible complications (thrombosis, biliary ducts compressions, etc.). In fact, 88% of those patients showed no progression of the aneurysm over the years. However, other data showed that surgery (aneurysmorraphy/aneurysmectomy) could be an option for evolutive aneurysms. Percutaneous embolization has also been performed for evolutive intrahepatic portal vein aneurysms (growing from 1.7 to 3 cm within 3 months in a reported case) [6,7].

Therefore, the therapeutic strategy was part of the follow-up and the management of the possible complications.

In our patient's case, considering that the aneurysm is intrahepatic and saccular, surgical management is not possible except for liver re-transplantation. In this context, following the

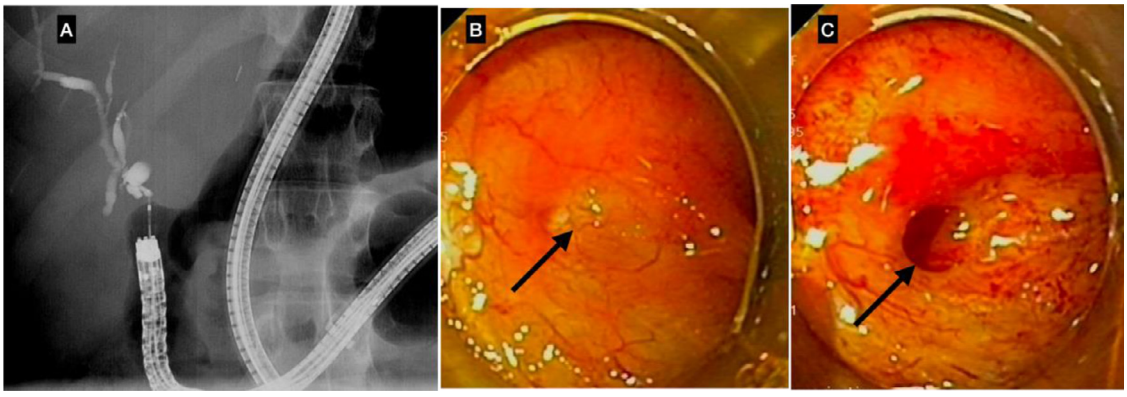


**Fig 3.** Entero-endoscopic retrograde cholangiography showing anastomic stricture, **(A)** pintpoint hepatico-jejunal anastomosis **(B)** before and **(C)** after balloon dilation.

overall stability of the size, wait-and-see management seems reasonable.

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