ISSN: 2634 - 680X

Journal of Clinical Case Studies Reviews & Reports

SCIENTIFIC Research and Community

Case Report

Open Access

Rare Case of Portal Vein Aneurysm after Liver Transplantation

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ABSTRACT

Objective: diagnosis and treatment experience of portal vein aneurysm after liver transplantation.

Methods: One recipients with portal vein aneurysm after liver transplantation were analyzed. Clinical features, diagnosis, treatment and prognosis were summarized based on literature review.

Results: One case received high-dose glucocorticoid therapy whereas liver function was not improved, and then patient was recovered successfully after secondary liver transplantaion.

Conclusions: Long-term complication of portal vein aneurysm after liver transplantain probably indicates poor prognosis. Intimate follow-up and active treatment should be conduct Liver transplantation may be a potential treatment regimen.

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Received: April 10, 2024; **Accepted:** April 22, 2024; **Published:** April 27, 2024

Introduction

Portal vein aneurysm (PVA) is a rare condition characterized by the abnormal enlargement of the portal vein, first identified in 1956 by Barzilai and Kleckner [1]. Since then, only a few cases have been documented in medical literature, making it a relatively uncommon condition. In PVA, the portal vein outside the liver is partially dilated, taking on a sac-like or spindle-like shape, which can lead to complications such as rupture, thrombus formation, or further enlargement that may necessitate surgical intervention. Long-term complications following liver transplantation, although rare, can include chronic rejection, opportunistic infections, post-transplant lymphoproliferative disease, and biliary complications [2]. Despite the advancements in multimodal imaging techniques in recent years that have improved the diagnostic rate of PVA, there is still a lack of consensus regarding the underlying causes and optimal treatment approaches for this condition [3]. This cases report of long-term PVA post-liver transplantation, highlighting the potential association between PVA and poor patient prognosis by combining these cases with a comprehensive literature review, the aim is to enhance the understanding among clinical physicians regarding the characteristics of PVA and the most effective treatment strategies for PVA following liver transplantation. This research underscores the importance of continued study and awareness of this rare condition to improve patient outcomes and quality of care after retransplantation of liver [4,5].

Case Report

52 years old Female in 2023 was admitted to our hospital due to abnormal liver function for 3 days after 10 years of liver transplantation. In 2013, the patient underwent classic orthotopic liver transplantation at the General Hospital due to autoimmune hepatitis and post-hepatitis cirrhosis. The donor-recipient portal vein diameter matched well and end-to-end anastomosis was performed. In 2014, it was discovered that portal vein anastomotic stenosis was treated with portal vein stent placement. Afterward, regular follow-up examinations were conducted, and treatment with tacrolimus, sodium mycophenolate, and methylprednisolone for anti-rejection reactions, as well as aspirin for antiplatelet therapy, showed no significant abnormalities in the transplanted liver function and unobstructed portal vein and stent blood flow. In 2022, re-examination revealed portal vein stent obstruction and stent recanalization treatment, followed by anticoagulation with rivaroxaban. In 2023, the patient underwent a follow-up examination at a local hospital due to fatigue and yellow urine. After discovering abnormal liver function, the patient was admitted to our hospital for treatment. Based on medical history, the possible causes of liver dysfunction include vascular complications, rejection reactions, and recurrence of autoimmune hepatitis. The admission test results indicate: positive for anti-nuclear antibodies and positive for anti-mitochondrial type 2 antibodies. Abdominal vascular contrast-enhanced CT shows hepatic portal vein stenosis, PVA-like dilation in the liver, patency of the stent after placement, and portal hypertension. Ultrasound indicates cystic dilation of the main hepatic portal vein and left and right branches of the portal vein, placement of a portal vein stent, and good blood flow signal Magnetic resonance, The clinical type of the patient, and at the same time, after discharge, the patient will receive targeted treatment during the illness, Pancreatic

cholangiography shows local stenosis of the common bile duct and intrahepatic bile duct. The pathological results of liver biopsy indicate scattered focal lesions and fusion necrosis of liver cells, extensive infiltration of lymphocytes and plasma cells in the portal area, and consideration of acute rejection moderate, accompanied by recurrence of autoimmune hepatitis. . After receiving high-dose corticosteroid shock therapy, the patient's liver function did not improve and gradually progressed to liver failure. The patient underwent a second liver transplant and recovered smoothly after the transplant surgery.

Discussion

Portal vein aneurysm (PVA) is a condition that can be divided into two categories: congenital and acquired. The exact cause of PVA is unknown, but it is believed that congenital PVA may be related to the formation of diverticula or venous wall defects in the distal end of the right primordial yolk vein during embryonic development [6]. In contrast, acquired PVA is mainly caused by portal hypertension, which leads to hemodynamic changes accompanied by increased shear stress. In this causes compensatory thickening of the vascular intima, which is replaced by fibrous tissue, ultimately weakening the tensile strength of the venous wall and leading to the formation of tumor-like dilation, Other factors that may contribute to acquired PVA include necrotizing pancreatitis, abdominal trauma, malignant tumors, or surgery. PVA can also occur postoperatively after liver transplantation. Based on previous studies and the characteristics of one cases discussed in this article, risk factors for the long-term occurrence of PVA after liver transplantation include mismatched portal vein diameter between the donor and recipient, stenosis of the portal vein anastomosis, postoperative hypercoagulable state, and recipient portal vein thrombosis or thrombectomy [7]. In the article discuss one case with a patient developed PVA after liver transplantation due to significant portal vein stenosis and thrombosis that was discovered early after surgery. Although a stent was inserted to ensure smooth portal vein blood flow, PVA-like dilation still occurred in the long postoperative period. When there is stenosis in the reconstructed portal vein after liver transplantation, the blood flow into the hepatic vein is prone to turbulence after passing through the stenosis, which continuously impacts the venous wall, leading to endometrial damage and ultimately forming tumor-like dilation. The formation of blood clots may further increase the shear stress of turbulent blood flow, accelerating the dilation of veins [8]. Most patients with concurrent PVA have no obvious symptoms and are accidentally discovered. Some patients experience non-specific abdominal pain, and less than 10% of patients may experience gastrointestinal bleeding, portal hypertension, or adjacent organ compression symptoms such as bloating, and jaundice Thrombosis is a common complication, with an incidence rate of up to 20%. In severe cases, it can lead to intestinal ischemia and necrosis, endangering patient life. Among the 9 cases of PVA after liver transplantation reported in previous literature, 1 case died of transplant liver failure in a few years after the discovery of PVA, 3 case was suspected to be related to PVA compression due to intrahepatic bile duct dilation, and the remaining 5 cases did not show PVA related symptoms. The 9 patients reported in this article, 1 case developed PVA after liver transplantation due to portal vein stenosis, leading to complications such as portal vein stenosis and liver failure which lead to organ failure cause death, there was 3 case with no significant clinical manifestation in the early stage of PVA, but as the condition progressed, the patient developed nodules. Related to PVA compression, the remaining 5 cases did not show PVA related symptoms. The one patients reported in this article there were no obvious clinical manifestations in the early stage of PVA, but as the condition progressed, the patient developed portal hypertension and jaundice, Based on the cause of jaundice, combined with biochemical, imaging, pathological and other examinations, the possibility of portal venous cholangiopathy was considered. The patient ultimately received a second transplant due to liver transplant failure [9].

PVA can be caused by a variety of factors, including chronic liver disease, portal hypertension, pancreatitis, trauma, and surgery. According to Koc et al., portal vein aneurysms were correlated with cirrhosis in 12% of patients, while portal hypertension was identified as the most frequent cause in 32% of cases. Portal vein aneurysm (PVA) can be classified into three types: intrahepatic, extrahepatic, and mixed, based on its location [10]. The extrahepatic type is more common, with the main portal vein being the most frequent location, accounting for about 38.4%, followed by the confluence of the splenic vein and superior mesenteric vein, accounting for 23.6%. Previous cases of PVA after liver transplantation showed four cases of intrahepatic type and five cases of extrahepatic type with no obvious PVA-related symptoms. However, this article presents four cases of intrahepatic PVA after liver transplantation that deserve attention and protocal, It is crucial to mention that among the previous cases, five patients with extrahepatic PVA after liver transplantation, and they did not show any obvious PVA-related symptoms. On the other hand, three out of four cases of intrahepatic PVA after transplantation experienced transplant liver failure, indicating that the location of PVA in different parts of the liver after transplantation is closely related to prognosis [11].

Intrahepatic PVA after liver transplantation indicate that the four patients mentioned above with severe complication and liver failure as compare to extrahepatic PVA after liver transplantation to without obvious PVA related symptoms; Among the previous 2 cases of intrahepatic type, as well as the 2 cases of PVA after intrahepatic liver transplantation in this article, 3/4 of them experienced transplant liver failure, indicating that PVA in different parts of the liver after transplantation may be closely related to prognosis. Among the 9 previously reported cases of postoperative PVA after liver transplantation, 5 were first discovered during routine medical follow-up, and 3 was unexpectedly discovered during CT examination due to other diseases upon admission and other 1 patients prestented with unstable after liver transplant due to sever complication. Regular ultrasound examination after liver transplantation is an effective means to timely detect PVA after liver transplantation, while CT and MRI can provide more accurate information for clinical practice and guide subsequent treatment in terms of precise location, size, relationship with adjacent organs, and the presence of complications.Diagnostic tests such as color Doppler ultrasound, CT, and MRI are valuable tools in identifying and monitoring PVA. Ultrasound results indicate that PVA is a cystic lesion without echoes that is connected to the portal vein system. Color Doppler ultrasound allows us to view the internal venous blood flow and to detect the venous spectrum with pulse Doppler. However, if there is a thrombus formation, blood flow signals and venous spectra are not visible at the site of the thrombus. CT and MRI images reveal that PVA appears as a cystic/spindle-like dilation with equal density (CT) or long T1 and long T2 signals (MRI). During enhanced scanning, significant enhancement can be seen in the venous phase. When acute thrombosis accompanies it, an unenhanced filling defect can be observed in the vascular lumen. In the case of subacute or chronic thrombosis, it can appear as cavernous transformation, which is visible along the dilated collateral venous network produced by chronic thrombosis [12,13].

There are currently no specific guidelines or standards for the treatment of PVA due to a lack of scientific evidence. For most patients, especially those with small PVA diameter, no obvious symptoms, and no concurrent cirrhosis or portal hypertension, regular ultrasound follow-up is considered the best option. Most PVAs are stable, with a low risk of complications, and 88% of patients do not experience tumor size or progression of complications during follow-up. However, when acute portal vein thrombosis occurs, timely anticoagulation treatment is recommended, and about 80% of patients can achieve complete or partial reperfusion. However, when anticoagulation treatment fails and the condition progresses, widespread thrombosis of the splenic vein and superior mesenteric vein occurs, and corresponding symptoms occur due to venous obstruction, intravascular intervention technology can be adopted, such as thrombolysis or thrombosis under intervention. Clear to restore venous patency of surgery in PVA treatment is still controversial. Overall, the treatment of PVA should be determined on a case-by-case basis based on symptoms, the presence of cirrhosis, the size of PVA, and anatomical conditions. For complex PVA cases, such as obvious symptoms, progressive expansion of the tumor, significant compression on adjacent structures, thrombosis, or rupture of the tumor, surgical intervention can have a therapeutic effect The surgical plan varies depending on whether portal hypertension is present. In patients without portal hypertension [14-16].

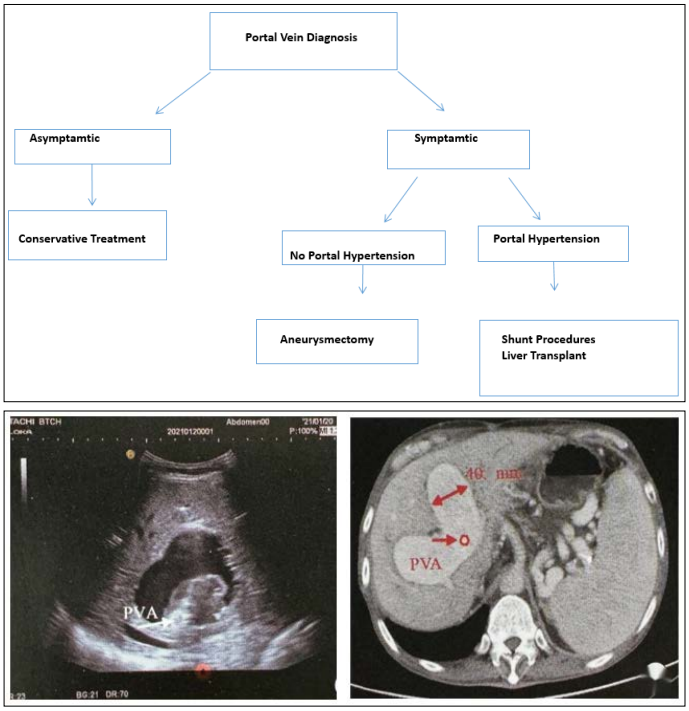
The prevalence of Portal Vein Aneurysm (PVA) in the general population ranges from 0.06% to 0.43%, which is relatively lower in the liver transplant population. The clinical data of PVA patients after liver transplantation is shown in Table 1. There are successful reports of PVA repair or venous tumor resection for restoring the normal diameter of the portal vein. In cases of concomitant portal hypertension, venous shunting or liver transplantation can be alternative surgical treatments. Mesenterico-Rex bypass has shown good clinical efficacy in three pediatric patients with intrahepatic PVA. However, surgical treatment involves certain risks. Among the 190 patients reported in the literature, 21.1% (40/190) underwent surgical treatment, and their postoperative mortality rate was 17.5%, For postoperative PVA after liver transplantation, all five cases were found and followed up for a long time. Four out of five patients did not show any PVA-related symptoms, and only one patient developed bile duct stricture, which was suspected to be related to the mass effect of PVA. However, the patient's prognosis was good after treatment with bile duct dilation, However, after the first discovery of PVA after liver transplantation in the two patients in this article, dynamic observation, thrombolytic therapy, and interventional shunt treatment were performed, and the transplant liver failure still occurred, reaching the indication for secondary transplantation. The most frequently performed procedures for portal vein aneurysms are surgical thrombectomy and aneurysmorrhaphy. However, there is no conclusive evidence regarding the long-term use of anticoagulation therapy for thrombosed portal vein aneurysms. A previously reported case of a patient receiving postoperative anticoagulation therapy with warfarin resulting in a hemorrhagic complication requiring reoperation raises concerns about the efficacy of anticoagulation therapy in such cases. Hence, cautious consideration is recommended while contemplating the use of anticoagulation therapy for the treatment of thrombosed portal vein aneurysms [17-22].

Table 1: Clinical Data of 9 Patients with PVA after Liver Transplant Reported in Litrature Review

Author	Patient	Age	Gender	Year	Etiology	Managment	Cause	PVA position	Outcome
Ferraz-Neto et al [12]	1	59	M	2004	HCV cirrhosis	Liver Transplant	Acquired	Intrahepatic	Died
Cho et al [13]	1	62	F	2007	HCV cirrhosis	Liver Transplant	Acquired	Extrahepatic	Survive
Miyazaki et al [14]	1	57	F	2008	HCV cirrhosis	Liver Transplant	Acquired	Extrahepatic	Survive
Francesco et al [15]	1	60	M	2010	HCV cirrhosis	Liver Transplant	Acquired	Extrahepatic	Survive
Schwope et al [16]	1	23	F	2010	HCV cirrhosis	Liver Transplant	Acquired	Intrahepatic	
Quan et al [17]	1	46	M	2012	HBV cirrhosis	Liver Tansplant	Acquired	Extrahepatic	Survive
Molinares et al [18]	1	6	F	2013	HAV	Liver Transplant	Acquired	Extrahepatic	Survive
Majeed et al [19]	1	76	F	2022	HCV cirrhosis	Liver Transplant	Acquired	Intrahepatic	Survive
Nassafi et al [20]	1	25	M	2023	Glycogen storage disease type 4	Liver Transplant	Acquired	Intrahepatic	Survive

Conclusion

In summary patients with long-term PVA after liver transplantation in this article experienced transplant liver failure, indicating that the occurrence of PVA after liver transplantation may be related to poor prognosis. A correct understanding, close follow-up, and active management may be the scientific treatment strategy for PVA in liver transplant recipients in the long term, and timely re liver transplantation is an optional treatment plan.



Note

Figure show an irregular shape in the liver with portal venous system on ultrasound doppler, Blood flow filling defects can be seen inside the aneurysm, and strong echogenicity can also be seen; The image shows that the lesion has turbulent blood flow signals originating from the venous system, It is diagnosed as a left and right hepatic portal vein aneurysm. Intratumoral blood flow filling defect without blood flow signal, diagnosed as portal vein aneurysms. Figure shows tumor like dilation of the left and right branches of the portal vein on CT plain scan, with a maximum diameter of 40.mm and visible TIPS stent shadow (arrow).

Conflict of Interest

The authors have no conflicts of interest to declare.

Funding Source

This study was not supported by any sponsor or funder.

References

1. Barzilai R, Kleckner Ms Jr (1956) Hemocholecyst following ruptured aneurysm of portal vein; report of a case. AMA Arch Surg 72: 725-727.
2. Brock PA, Jordan PH Jr, Barth MH, Rose AG (1997) Portal vein aneurysm: a rare but important vascular condition. Surgery 121: 105-108.
3. Ozbek SS, Killi MR, Pourbagher MA, Parildar M, Katranci N, et al. (1999) Portal venous system aneurysms: report of five cases. J Ultrasound Med 18: 417-422.
4. Calligaro KD, Ahmad S, Dandora R, Dougherty MJ, Savarese RP, et al. (1995) Venous aneurysms: surgical indications and review of the literature. Surgery 117: 1-6.
5. López-Machado E, Mallorquín-Jiménez F, Medina-Benítez A, Ruiz-Carazo E, Cubero-García M (1998) Aneurysms of the portal venous system: ultrasonography and CT findings. Eur J Radiol 26: 210-214.
6. Admir Kurtcehajic, Enver Zerem, Ervin Alibegovic, Suad Kunosic, Ahmed Hujdurovic, et al. (2023) Portal vein aneurysm—etiology, multimodal imaging and current management. World J Clin Cases 11: 725-737.
7. Hidaka H, Kokubu S, Sato T, Katsushima S, Izumi N, et al. (2018) Antithrombin III for portal vein thrombosis in patients with liver disease: a randomized, double-blind, controlled trial. Hepatol Res 48: E107-E116
8. Vaidya S, Dighe M, Kolokythas O, Dubinsky T (2007) Liver transplantation: vascular complications. Ultrasound Q 23: 239-253.
9. Gumus M, Yilmaz O, Ersoy O, Demirezen AB, Akin E, et al. (2010) Intrahepatic portal vein aneurysm in an asymptomatic patient. Ann Hepatol 9: 213-214.
10. Koc Z, Oguzkurt L, Ulusan S (2007) Portal venous system aneurysms: imaging, clinical findings, and a possible new etiologic factor. AJR Am J Roentgenol 189: 1023-1030.
11. Alamo JM, Gómez MA, Tamayo MJ, Socas M, Valera Z, et al. (2005) Mycotic pseudoaneurysms after liver transplantation. Transplant Proc 37: 1512-1514.
12. Ferraz-Neto BH, Sakabe D, Buttros DA, Resende MB, Afonso RC (2004) Portal vein aneurysm as late complication of liver transplantation: a case report. Transplant Proc 36: 970-971.
13. Cho SW, Marsh JW, Fontes PA, Daily MF, Nalesnik M, et al. (2008) Extrahepatic portal vein aneurysm--report of six patients and review of the literature. J Gastrointest Surg 12: 145-152.
14. Miyazaki K, Takatsuki M, Eguchi S, Hidaka M, Tokai H, et al. (2008) Living donor liver transplantation for hepatitis C virus cirrhosis with a huge portal vein aneurysm. Liver Transpl 14: 1221-1222.
15. Francesco Fd, Gruttadauria S, Caruso S, Gridelli B (2010) Huge extrahepatic portal vein aneurysm as a late complication of liver transplantation. World J Hepatol 27: 201-212.
16. Schwope RB, Margolis DJ, Raman SS, Kadell BM (2010) Portal vein aneurysms: a case series with literature review. J Radiol Case Rep 4: 28-38.
17. Quan JR, Lu Q, Ling WW, Luo Y (2012) Extrahepatic portal vein aneurysm after orthotopic liver transplantation: a case with 9-year ultrasound observations. J Med Ultrason (2001) 39: 271-274.
18. Molinares B, Alvarez S, García V, Sepúlveda ME, Yepes NL, et al. (2013) Extrahepatic portal vein aneurysm after liver transplantation in a child: case report. Pediatr Transplant 17: E33-E36.
19. Majeed N, Bannan B, Ahmad N, Zia Z, Ashour M (2022) Portal vein aneurysm, a pathological entity or an innocent bystander? A review of six cases. BJR Case Rep 1: 20220073.
20. Nassafi EM, Annet L, Ciccarelli O, Dahlqvist G (2022) Intrahepatic Portal Vein Aneurysm: A Case Report of an Uncanny Late Complication of Liver Transplantation. Transplant Proc 54: 1661-1663.
21. Admir Kurtcehajic, Enver Zerem, Ervin Alibegovic, Suad Kunosic, Ahmed Hujdurovic, et al. (2023) Portal vein aneurysm-etiology, multimodal imaging and current management. World J Clin Cases 11: 725-737.
22. Hidaka H, Kokubu S, Sato T, Katsushima S, Izumi N, et al. (2018) Antithrombin III for portal vein thrombosis in patients with liver disease: A randomized, double-blind, controlled trial. Hepatol Res 48: E107-E116.