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Case Report

A rare pediatric case of portal vein aneurysm thrombosis ☆,☆☆,★,★★

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ABSTRACT

Portal vein aneurysm (PVA) is rarely encountered, and published papers describing this etiology in adults and children typically include only case reports or small case series. We present a clinical case of PVA in a child associated with severe complications, including diffuse thrombosis of the portal venous system. A 10-year-old boy presented with abdominal pain and vomiting, resulting in an initial diagnosis of pancreatic head tumor based on suspicious images on abdominal grayscale ultrasound. Contrast-enhanced computed tomography confirmed a diagnosis of occlusive PVA thrombosis (36 × 37 × 95 mm). Lacking drastic symptoms, the patient was treated with conservative anticoagulant therapy. On follow-up, the thrombosis appeared to shrink gradually and disappeared at 6 months based on Doppler ultrasound imaging. The PVA was reduced in size, and hepatopetal flow was restored. Surgeons and radiologists should be aware of this rare entity to ensure that a precise diagnosis can be established and to provide suitable treatment.

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Introduction

Portal vein aneurysm (PVA) is a rare disorder, with only 200 cases recorded in the literature, in the form of clinical case reports or case series describing small numbers of patients, most of whom have been adults.[1,2] A portal vein diameter larger than 2 cm on ultrasound or computed tomography imaging is considered the definitive diagnostic criteria for PVA [3], which is often detected accidentally in patients who present without symptoms or serious comorbidities. [1,2] Possible complications of PVA include thrombosis, aneurysm rupture, and portal hypertension, among which thrombosis is the most common complication.[1] Due to its rarity, the treatment of PVA and its complications remain controversial, with no specific guidelines established for this disorder.[4] Our case was a 10-year-old child who suffered from PVA with thrombosis, which was initially misdiagnosed as a pancreatic head tumor. The final diagnosis was accurately made following abdominal contrast-enhanced computed tomography, and the patient was successfully treated with subcutaneous anticoagulant therapy.

Case report

A 10-year-old boy presented with abdominal pain and vomiting 6 hours prior to hospital admission. The patient's individual and familial medical histories were unremarkable. Physical examination revealed mild periumbilical pain without guarding. Biochemical blood tests (Aspartate aminotransferase-AST, alanine transaminase-ALT, blood urea nitrogen-BUN, creatinine, bilirubin, and amylase levels) and complete blood counts were within the normal ranges except for a mild increase in lipase of 260 IU/L (normal range: 7 –60 IU/L). An abdominal ultrasound scan revealed a hypoechoic 35 × 90 mm mass at the pancreatic head, which led to an initial diagnosis of a pancreatic head tumor.

The mass was assessed by intravenous, contrast-enhanced computed tomography, which revealed a 36 × 37 × 95 mm low radiodensity mass that did not collect the contrast agent. The mass was located diffusely in the portal system, including the main portal vein, the 2 branches of the portal vein, the distal segment of the splenic vein, and the superior mesenteric vein. A thin margin with increased density appeared around the mass. Several periumbilical and gastric venous collaterals were also identified. No images displayed evidence of hepatic parenchymal ischemia, splenomegaly, or intra-abdominal tumors (Fig. 1A and B).

A diagnosis of extra-hepatic PVA thrombosis was made, and screening tests were performed for hypercoagulation, including the evaluation of prothrombin time (PT), activated partial thromboplastin time (aPTT), fibrinogen levels, international normalized ratio (INR), protein S levels, and protein C levels revealed values within the normal ranges.

After 3 days of treatment with 36 mg enoxaparin (1 mg/kg), subcutaneously administered twice daily, the patient was free from abdominal pain and vomiting. He was discharged from the hospital with the continuation of anticoagulant treatment at home. A dose of 1mg/kg/12 hours enoxaparin was prescribed and monitored by serum anti Xa level. During the first 3 months, although the patient remained asymptomatic, thrombosis did not vary much in size. Obvious improvement began to be observed after 3 months when the thrombosis began to shrink on each re-examination. At 6 months, the thrombosis disappeared completely on Doppler ultrasound. The PVA reduced in size (to 17 mm), and the hepatopetal flow of the portal vein was restored (Fig 2A andD).

Discussion

Among visceral aneurysms, PVA is the most common type of venous aneurysms with an incidence of 0.06% [2,4-6] and accounting for 3% of all venous aneurysms.[2,3,7,8] The first case was reported in 1956, and only approximately 200 cases have been recorded since then, predominantly identified in adult patients.[4] The most common location for PVA is the confluence of the splenic vein and the superior mesenteric vein.[2,3] Most venous aneurysms are asymptomatic, and aneurysmal rupture or the compression of neighboring organs due to thrombosis are rare occurrences.[2] Thrombosis of the PVA occurs in approximately 50% of clinical cases [2]; however, this complication could induce portal hypertension, with severe clinical manifestations, including gastrointestinal bleeding.[2,9,10] According to Koc et al.[2], the adult patients with clinical symptoms have larger PVAs (33.6 mm ± 9.9 mm, compared with 23.1 ± 3.3 mm, $P < .0001$). Our pediatric patient presented with abdominal pain and vomiting and was diagnosed with a PVA sized 36 × 37 × 95 mm, which we consider to be a large PVA.

The etiology of PVA remains unclear and could be congenital or acquired. Acquired etiologies are often associated with portal hypertension, severe pancreatitis, and malignancy metastasizing into the portal vein. Congenital etiologies include abnormalities of the congenital venous wall or the incomplete degeneration of the right vitelline vein.[11–14] Our patient was a child without any history of trauma or significant comorbidity, suggesting a congenital etiology for this PVA.

PVA might induce many complications, such as thrombosis, rupture, duodenal compression, inferior vena cava syndrome, and jaundice. Thrombosis is the most common complication, with a prevalence that fluctuates between 6% – 13% [12], and is the complication we observed in our clinical case. Aneurysmal rupture is a rare occurrence, with few reported cases to date.[12,14] The diagnosis of PVA primarily depends on abdominal color Doppler ultrasound, computed tomography, or magnetic resonance imaging.[15] In cases without thrombosis, the abdominal color Doppler ultrasound displays a continuous dilated portal vein, including the splenic vein and the mesenteric veins, with intra-luminal, low-velocity flow. In cases with total thromboembolism of the portal vein, the flow image on abdominal color Doppler ultrasound would appear negative, and the thrombosis can generate a pseudotumoral image, which is typically mistaken for a tumor on initial diagnosis.[4] Our patient was diagnosed accurately using abdominal, contrast-enhanced computed tomography. The in-

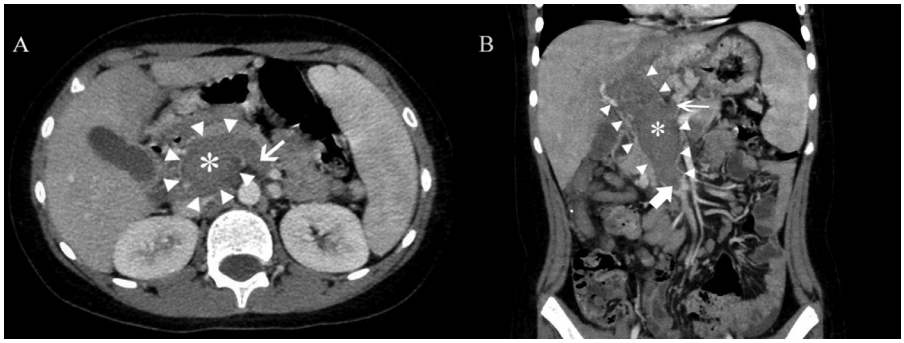

Fig 1 – Portal vein aneurysm thrombosis on abdominal, contrast-enhanced computed tomography. Contrast-enhanced computed tomography images through the axial (A) and coronal (B) planes. The thrombosis (asterisk) appears diffuse throughout the portal vein (arrowheads), the splenic vein (thin arrow), and the superior mesenteric vein (thick arrow)

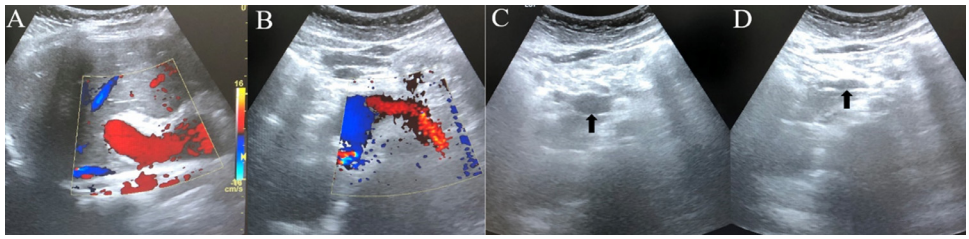

Fig 2 – Doppler ultrasound images of PVA thrombosis at the 6-month follow-up after anticoagulant therapy. (A) Portal vein (17 mm) with positive hepatopetal flow. (B) Positive flow of the splenic vein to the portal vein. (C, D) The superior mesenteric vein (arrow), (C) before and (D) after applying transducer pressure, suggesting no thrombosis

traluminal portal venous thrombosis was characterized by an image of a mass in the lumen of the portal vein that did not collect the contrast agent and appeared continuous with the splenic vein and the mesenteric veins, featuring and a thin margin with increased density.[4] Potential causes of intraluminal portal venous thrombosis, including hepatobiliary cancer, portal vein leiomyosarcoma, intra-abdominal infectious mass, and hypercoagulable states, were excluded in our clinical case.

The current treatment approach for PVA remains controversial due to a lack of scientific evidence.[2,11] According to the current literature, a conservative non-pharmacotherapy approach combined with Doppler ultrasound monitoring is the preferred management strategy for patients with small aneurysms without complications or evidence of cirrhosis or portal hypertension.[2,14,16] 1 case report described a patient with PVA who was monitored for longer than 10 years without any increase in aneurysmal size or the development of complications.[16,17] In cases with thrombosis, pharmacotherapy using anticoagulants results in complete revascularization or partial revascularization in greater than 80% of cases.[14] In cases of anticoagulant pharmacotherapy failure, or if signs of neighboring organ compression are identified, vascular intervention procedures, such as thrombosis aspiration, should be considered.[12] Surgery is typically reserved for large PVAs with severe clinical symptoms or ruptured aneurysms.[18] In cases without portal hypertension, the preferred surgical approach is angioplasty or portal vein replacement.[15,19] A transjugular intrahepatic portosystemic shunt (TIPS) or hepatic transplantation are options available to patients with portal hypertension or other chronic hepatic disorders.[12] In our clinical case, despite the large thrombosis, we opted for a conservative approach using anticoagulant therapy because the child presented with mild symptoms. Improvements in thrombosis size occurred slowly and only became obvious after 6 months, which we consider a long period of time. To our surprise, the thrombosis completely disappeared, indicating treatment success. For PVA complicated by thrombosis, we believe that conservative anticoagulant therapy should be attempted for at least 3 to 6 months prior to considering more invasive therapies.

Conclusion

PVA complicated with thrombosis is a rare pediatric disorder. The pseudotumoral thrombosis images on ultrasound can confuse clinicians, and computed tomography is a valuable diagnostic tool for discerning between tumors and thrombosis. Due to the lack of specific treatment guidelines available for PVA, clinicians must carefully consider several elements, including clinical symptoms, aneurysmal size, complications, and comorbidities, when choosing an appropriate treatment. Conservative therapy using anticoagulants should be selected and continued for at least 3–6 months in patients with no severe clinical symptoms, even in cases with large aneurysmal sizes.

Patient consent

Written informed consent was obtained from the legal guardian of all participating patients.

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