CASE REPORT

# Portal Vein Thrombosis - A Rare And Important Cause of Abdominal Lump in Neonates

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**INTRODUCTION :** Neonatal portal vein thrombosis (PVT) previously described by Thompson and Sherlock as exceedingly rare[1] is being recognized more frequently nowadays. Incidence varies from 1 in 100,000 live births[2] to 36 per 1000 NICU admissions[3]. Despite the reported rarity in the neonatal setting, PVT is the major cause of extrahepatic portal hypertension and gastrointestinal bleeding in children[4]. Portal vein occlusion associated with small periportal collaterals is termed as cavernous transformation of the portal vein (CTPV)[5]. The etiology of neonatal PVT differs from that in children and adults. In adults PVT is most commonly secondary to cirrhosis[6]. PVT is a result of liver transplantation, intraabdominal sepsis, splenectomy, sickle cell anemia, and antiphospholipid antibodies in older children . In about 50% of children with PVT, etiology remains unknown[7]. Mechanicaland chemical damage to a vessel wall by a catheter is believedto initiate the thrombotic process, making UVC placementthe major risk factor for PVT[8]. There have been reports of thrombophilia associated with PVT in neonates and children. Case series have reported factor V Leiden, prothrombin gene mutation, protein C deficiency, protein S deficiency, antithrombin deficiency, increased factor VIII, methylene tetrahydrofolate reductase (MTHFR) variants with elevated homocysteine, and elevated lipoprotein[9].

**CASE REPORT :** A female neonate; normally delivered at term with birth wt 3.2 kg with no significant antenatal complaints and no history of resuscitation or admission at birth and normal postnatal course with history of passing stool & urine within 24 hrs of birth and breast fed within 1 hr, was brought at live day (LD) 28 with chief complaint of abdominal distension since 3 days which was non progressive, not associated with respiratory distress or bowel & bladder problems and no feeding problems.

Fever with pustules all over the body was noticed in the baby at LD2 which got relieved by oral OPD treatment within 4 days. Umblical cord was normal at birth & fell off at LD12. There is history of application of ghee for 10-15 days on umblical stump and ghutti (herbal preparation) was given to the baby since 5 days before abdominal distension for a total duration of 7-8 days. The child has no history of umblical catheterization or umblical sepsis, yellowish /bluish discoloration of skin, respiratory distress, bleeding from any sites, rashes, ecchymosis.No vaccination done and no documentation of inj vitamin K given at birth.

Primimother of 19 yrs of age has no history of drug intake, radiation or any other significant illness except that she had fever on 2nd day of delivery & was also given 1 unit of PRBC transfusion.

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On General examination crying sucking activity and neonatal reflexes were normal, craniofacial configuration was normal with normal genitalia. The child was afebrile and heart rate was 148/min regular with all peripheral pulses equally palpable, respiratory rate was 54/min with no retractions, Capillary refill time <3 sec on sternum and Spo2 98 % in right upper & lower limb. Weight on admission was 3.5 kg. Pallor was present. No cyanosis, icterus, lymphadenopathy or odema seen.

Abdominal distension was noted with no flank fullness. No dilated veins over abdomen seen.umblicus centrally placed. The Lump was incidentally noticed, firm, about lemon size and non progressive with no skin redness or crying while touching over the mass.

Abdominal lump was about 5×4 cm firm in consistency, oblongated, felt in periumbilical region with no delineated superior margins but inferior margin well defined, with smooth surface & non mobile. No signs of redness/ abdominal tenderness seen. Inferior margins of left lobe of liver could not be delineated. Tip of spleen was just palpable.Normal bowel sounds present. No bruit was audible over the mass. Other system examinations were normal.

Complete blood count on LD 28 depictednormocytic normochromic anaemia (8.7 gm %), TLC 21000, APC 2.39 lakhs, LDH 357 RFT, LFT, SE, Serum total protein, PT/PTTK with INR and total lipid profile were normal. we repeated the investigations 4 times over a period of 21 days hospital stay and all above test results were normal . PBF depicted microcytic hypochromic anemia and reticulocyte count was 1.2%. CRP was negativewith blood andnormal urine microscopy and urineculture and sensitivitysterile. Protien C was 19%, Protien S was 64% and homocystiene was > 50 micromoles.

USG abdomen on LD 30 revealed portal vein thrombosis extending from terminal SMV, confluence to porta hepatis with minimal ascites.Repeat ultrasound after 15 days suggested splenomegaly with multiple collaterals noted at splenic hilum, periportal & peripancreatic region with Increase in no. of collaterals Portal vein is completely replaced by these periportal collaterals suggestive of cavernoma. Mild ascites present.

MRI abdomen revealed peri portal thickening with non visualization of rightt &left portal vein, main portal vein,splenic vein & superior mesenteric vein-suggestive of thrombus. CECT abdomen revealed main portal vein, right & left of portal vein, splenic vein & terminal part of superior mesenteric vein not opacified by contrast ? thrombus .

Inj Cefotaxime, vancomycin and metrogyl given on basis of high TLC and clinical suspicion for 10 days. Inj Vitamin K given.Abdominal lump gradually disapperared in 10 days. Repeat USG was done. Cavernoma with thrombous seen. Patient was started LMWH enoxaparin (1.5mg/kg/dose) bd SC till LD 45for 10 days during pendency of reports. No worsening/new symptoms were noticed. PT-INR &serial USGs were done which didn't show any adverse events of enoxaparin. Multiple strategies have been reported in the literature from observation, anticoagulation, and thrombolysis. The indications for anticoagulation treatment included the presence of a second, occlusive thrombus with liver parenchymal changes or involving two branches of the portal vein[3].

**DISCUSSION :** The long term follow up and clinical importance of thrombosis detected on ultrasound in asymptomatic neonates are not fully elucidated . The spontaneous regression of catheter-related thrombi detected on ultrasound has been reported[15]. Early spontaneous resolution has been postulated as a reason why PVT was diagnosed relatively rarely in the clinical setting of the neonatal intensive care unit[4,14]. If the PVT does not resolve, It may transform into a cavernoma, with dilated pancreaticoduodenal and prebiliary veins, secondary to portal hypertension[8]. There may be an absence of clinical and laboratory signs with PVT in the neonate.

Thrombocytopenia may be seen at the time of diagnosis, but is not specific for PVT[3]. Consumption from the thrombosis or the concomitant clinical risk factors for thrombosis, such as sepsis, may explain the low platelet count in the acute phase. In the late chronic stage, a low platelet count may be secondary to hypersplenism with portal hypertension[10]. In contrast to adults, liver function is usually normal in children presenting with PVT. There can be mild liver biochemical abnormalities in children with PVT[8]. The extent to which similar abnormalities occur in neonates is less clear. Nine out of 133 (7%) neonates had abnormal liver enzymes as the indication for ultrasound which identified the PVT[3]. A grading system for PVT based on ultrasound findings has been suggested. Grade 1 PVT was defined as non-occlusive PVT with normal liver parenchyma; grade 2 as occlusive PVT with normal liver parenchyma; and grade 3 as occlusive PVT with ultrasonographic abnormalities of the liver parenchyma[3].

Investigational radiographic studies in the diagnosis of PVT include abdominal ultrasound, computed tomography (CT), angiography, and magnetic resonance imaging (MRI). Accurate interpretation of the findings identified by each of these modalities can be difficult, especially in the neonates. Ultrasonography has been supplanted by Doppler ultrasound, which improves assessment by providing information on portal vein patency and blood flow characteristics. Doppler ultrasound and especially color flow Doppler is useful for confirming the changes in flow patterns around the thrombus and the resumption of normal flow pattern in follow-up imaging as the thrombosis resolves[11].

A CT scan accurately identifies PVT and the presence of collateral vessels but requires exposure to radiation and the use of intravenous contrast materials. Similarly, angiogram requires exposure to radiation and intravenous contrast material. MRI performs as least as well or better than CT in diagnosing PVT, without the issues of ionizing radiation exposure and intravenous contrast[11]. In comparing the efficacy of imaging techniques in identifying portal vein patency, Weinreb et al. found MRI superior to CT in visualizing hepatic architecture and vascular anatomy and patency in 27 children undergoing evaluation of suspected liver disease[11].

The role of anticoagulation in PVT management is unclear. There is an absence of prospective data on anticoagulation in the literature. Multiple strategies have been reported in the literature from observation, anticoagulation, and thrombolysis. The indications for anticoagulation treatment included the presence of a second, occlusive thrombus with liver parenchymal changes or involving two branches of the portal vein, post cardiac surgery. Dosages and lengths of treatment varied significantly[3]. Treatment of symptomatic acute PVT, extending to the main portal vein, by means of regional streptokinase infusion has been reported[12]. After initial treatment with UFH or LMWH, vitamin K antagonists (VKA) with a target INR of 2-3 could theoretically be used to continue anticoagulation therapy. However, the use of warfarin in the neonatal period is problematic. Formula-fed infants will receive large amounts of vitamin K, and will be warfarin resistant. Dosing becomes difficult as there is no commercially available liquid formulation of warfarin[13].

Given the anticipated difficulties, anticoagulation with warfarin would not be recommended in the neonatal period. LMWH may be preferable to UFH if anticoagulation therapy is used to treat neonatal PVT, given the predictable pharmacokinetics, reduced monitoring requirements, and possibly decreased rate of major bleeding. In a randomized, controlled trial of therapeutic anticoagulation in children, LMWH therapy was compared with UFH and VKA for the treatment of venous thromboembolic events in children. There was a major bleeding rate of 12.5% in the UFH/ VKA versus 5.6% in the LMWH arm[14]. A major bleeding rate of 0.7% of children treated with the low molecular weight heparin, enoxaparin, has been reported[15].

**Summary :** Neonatal PVT is being increasingly recognized due to sophisticated and advanced imaging technologies. There may be no symptoms or laboratory abnormalities. Some patients may have accompanying thrombocytopenia or liver serum biochemical abnormalities. As a result of the lack of symptoms, the diagnosis may not be suspected. Umbilical catheterization and sepsis are risk factors for neonatal PVT. Thrombophilia may be a contributing risk factor. However, neonatal PVT may still occur in the absence of risk factors. From the available literature, there appears to be a good outcome in the majority of cases followed up to 8 years of age. Non-occlusive thrombosis is more likely to resolve than occlusive thrombosis. Neonates should be followed for at least 5 years after PVT to monitor for the development of portal hypertension in an attempt to avoid presentation with gastrointestinal hemorrhage in childhood. The role of anticoagulation in the management of neonatal PVT is unclear. The role of thrombophilia in neonatal PVT is unclear.

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## References

1. Thompson EN, Sherlock S. The aetiology of portal vein thrombosis with particular reference to the role of infection and exchange transfusion. Q J Med 1964;33:465-80.
2. Heller C, Schobess R, Kurnik K, et al. Abdominal venous thrombosis in neonates and infants: role of prothrombotic risk factors e a multicentre case-control study. For the Childhood Thrombophilia Study Group. Br J Haematol 2000;111:534-9.
3. Morag I, Epelman M, Daneman A, et al. Portal vein thrombosis in the neonate: risk factors, course, and outcome. J Pediatr 2006;148:735-9.
4. Alvarez F, Bernard O, Brunelle F, Hadchouel P, Odievre M, Alagille D. Portal obstruction in children. I. Clinical investigation and hemorrhage risk. J Pediatr 1983;103:696-702.
5. Fisher MR, Wall SD, Hricak H, McCarthy S, Kerlan RK. Hepatic vascular anatomy on magnetic resonance imaging. Am J Roentgenol 1985;144:739-46.
6. Wilson KW, Robinson DC, Hacking PM. Portal hypertension in childhood. Br J Surg 1969;56:13-22.
7. Brady L, Magilavy D, Black DD. Portal vein thrombosis associated with antiphospholipid antibodies in a child. J Pediatr Gastroenterol Nutr 1996;23:470-3.
8. Kim JH, Lee YS, Kim SH, Lee SK, Lim MK, Kim HS. Does umbilical vein catheterization lead to portal venous thrombosis? Prospective US evaluation in 100 neonates. Radiology 2001;219:645-50.
9. Uttenreuther-Fischer MM, Vetter B, Hellmann C, et al. Paediatric thromboembolism: the influence of non-genetic factors and the role of activated protein C resistance and protein C deficiency. Eur J Pediatr 1997;156:277e81.
10. Stringer DA, Krysl J, Manson D, Babiak C, Daneman A, Liu P. The value of Doppler sonography in the detection of major vessel thrombosis in the neonatal abdomen. Pediatr Radiol 1990;21:30-3
11. Morag I, Shah PS, Epelman M, Daneman A, Strauss T, Moore AM. Childhood outcomes of neonates diagnosed with portal vein thrombosis. J Paediatr Child Health; 2011.
12. Rehan VK, Cronin CM, Bowman JM. Neonatal portal vein thrombosis successfully treated by regional streptokinase infusion. Eur J Pediatr 1994;153:456-9.
13. Veldman A, Nold MF, Michel-Behnke I. Thrombosis in the critically ill neonate: incidence, diagnosis, and management. Vasc Health Risk Manag 2008;4:1337-48.
14. Ignjatovic V, Najid S, Newall F, Summerhayes R, Monagle P. Dosing and monitoring of enoxaparin (low molecular weight heparin) therapy in children. Br J Haematol 2010;
15. Neonatal portal vein thrombosis: Diagnosis and management S. Williams, A.K.C. Chan / Seminars in Fetal & Neonatal Medicine 16 (2011) 329-339