Acta Scientific PAEDIATRICS (ISSN: 2581-883X)

Volume 7 Issue 1 January 2024

Case Report

Tens Ascites as a Presentation of Proteins Deficiency

Mehrnoush GH*

General Pediatrician, Super Specialty Assistant (Fellowship Assistant) of Pediatric Intensive Care, Tehran University of Medical Sciences

***Corresponding Author:** Mehrnoush GH, General Pediatrician, Super Specialty Assistant (Fellowship Assistant) of Pediatric Intensive Care, Tehran University of Medical Sciences.

Received: December 11, 2023

Published: December 22, 2023

Abstract

Introduction: Acites is not a usual finding in prehepatic portal hypertension including portal vein thrombosis, but when portal vein thrombosis is acute and massive, ascites can be a presenting feature. A new condition of interest is protein s deficiency associated with hypercoagulation and recurrent venous thrombosis and ascites. we report the case of a non cirrhotic 9 years old male Whit tense ascites diagnosed with portal and Superior mesentric vein thrombosis caused by protein s deficiency.

Case Report: A 9 years old boy was evaluated for tense ascites which has led to um local hernia since one month before admission. he had not any clinical or laboratory stigmata of paranchymal liver disease. he had history of abdominal pain that led to appendectomy four months ago. Imaging studies showed Superior mesenteric and portal vein thrombosis. In laboratory tests for pre thrombotic states he suffered from significant proteins deficiency.

Discussion: thrombophilic states like as proteins deficiency predispose patient to vascular thrombosis. This vascular thrombosis can be present with signs and symptoms related to their territories. This is essential to prevent advancement of thrombosis or rethrombosis in patient with inherited coagulation disorders in which lifelong anticoagulation therapy is recommended.

Keywords: Protein; Deficiency; Portal Vein Thrombosis; Ascites

Introduction

Ascites is a common complication of portal hypertension in cirrhotic patients in adults [1,2]. Portal hypertension can result from hepatic, post hepatic, prehepatic causes. portal hypertension often develops as result of extra hepatic portal vein obstruction in children [1,2]. In children the most common etiology of portal vein thrombosis is intra-abdominal infectious, history of umbilical vein catheterization in neonatal period and congenital anomalies of portal venous system are other causes. liberated or acquired thrombophilic States can cause vascular thrombosis too [3]. In korea, there are several reports of acute portal vein thrombosis caused by oral contraceptives and pancreatitis but few of them caused by deficiency of anticoagulant factors and reperfused with anticoagulation therapy [4]. Patients with portal vein thrombosis should be tested for an underlying thrombophilic condition [5], such as mutation of the prothrombin, factor 5, deficiency of protein s, c or antithrombin 3 or antiphospholipid syndrome [6,7].

Case Reports

This study presents a case of a 9 years old male with chief complaint of tense as cites led to umbilical hernia from 1 month ago. He had also history of abdominal pain and appendectomy and 4 kg weight loss and evidence of chronic pancreatitis in previous imaging from 4 months ago. We found him ill and cachectic with tense ascites and umbilical hernia in first examination, without any stigmata of chronic liver disease or jaundice or edeme. He did not have organomegaly. His vital signs were in normal range. diagnostic abdominal pancreatitis was done, and chemical ascites (pancreatic

or biliary) were ruled out. and high SAAG (serum albumen ascites gradient) with suggestion of portal hypertension detected. So intra hepatic causes including viral hepatitis, Wilson disease, autoimmune hepatitis was ruled out.

The result of tap of ascites in comparison of serum	Serum	Ascites
Alb	3.1	1.5
Total pr	6.6	2.8
Amylase	1273	457
Lipase	141	129
LDH	536	196

Table a

Imaging studies including mesenteric, portal and inferior vena cava vessels Doppler sonogrphy and CT scan of abdomen and thorax and pelvis with IV and oral contrast were done. Doppler sonography of abdominal vessels showed portal vein thrombosis associated with transformation in portohepatis and mesentric and coronary collateral vein in head of pancreas. chronic pancreatitis and pancreatic du dilation was seen too. (His father had pancreatitis and pseudo cyst surgery 4 years ago). We checked protein c [12.8 (Nl)], protein s [12.2 (low)], homocystein [16.8 (high (upper limit = 12.6)] for him. We asked for surgical consult, said that no surgical intervention is possible because of its chronic process, so that recommended medical therapy with anticoagulants. intra venous heparin and after 2 days, oral warfarin started. At the time of discharge, the maintenance dose of warfarin targeted to PT INR between 2 - 3 was continued. We put him on a low salt diet, diuretic and pancreatic enzymes replacement therapy too. (He showed signs of pancreatic insufficiently in pancreatic function tests.).

Discussion and Conclusion

portal vein thrombosis can led to portal hypertension and reduction in blood supply to the liver [13]. In children the most common etiology of portal vein thrombosis is intra abdominal infections history of umblical vein catheterization in neonatal period and congenital anomalies of portal venous system. Other less common causes include unhurried or acquired thrombophilic states such as mutation of prothrombin or factor 5 or deficiency of protein c or s or antithrombin 3 or antiphospholipid syndrome [3]. In the acute phase the presentation of portal vein obstruction us relatively uncommon and easily missed because the patient may be asymptomatic. Symptoms most often begin in the chronic or sub acute stage. Patiebts can present emergently with sudden onset of right upper quadrant pain, nausea and/or fever. alternatively, the symptoms of primary infection and inflammatory condition that led to portal vein obstruction can be seen eg; right lower quadrant pain in appendecites [13]. Studies have shown that anticoagulation in patients with acute or recent portal vein thrombosis can be recanalized the thrombosed vessel in more than 80% of cases. On patients with inherited coagulation disorders, lifelong anticoagulation therapy is recommend. on complicated cases shunt surgery or TIPS (trans jugular intra hepatic Porto systemic shunt) procedure is used [13]. In Mexico, Majluf-cruz., *et al.* studied 36 patients who had thrombosis related portal hypertension and found an incidence of 30% of protein c deficiency, whereas 9% had protein s deficiency in patients with primary thrombophila [6,9]. Similarly in Mexican patients with non-cirrhotic portal vein thrombosis, 31% had protein c deficiency [8]. however, a French study has found a high number of patients with non-cirrhotic portal vein thrombosis showed protein s deficiency [10,11]. In a study from United Kingdom, protein s deficiency was found in 3-8% of patients with portal vein thrombosis [10,11]. Other reports have also shown protein c or s deficiencies in patients with idiopathic portal hypertension [12]. measurement of protein c and s should be performed in patients with portal thrombosis. In conclusion, our case shows that portal vein thrombosis can be provoked by protein s deficiency. In patients(children) with protein s or c deficiency family member including parents and siblings should be screened.

Bibliography

1. Ginees P., *et al*. "Compensated cirrhosis: natural history and prognostic factors". *The Pathology* 7 (1987): 122-128.

2. Garcia - Tsao G. "Current management of the complications of cirrhosis and portal hypertension: variceal hemorrhage, ascites and spontaneous bacterial peritonitis". *Gastroentrology* 120 (2001): 726-748.

3. Primigani M Martinell., *et al*. "Risk factors for thrombophilia in extrahepatic portal vein obstruction". *The Pathology* 41 (2005): 603-608.

4. Belli L., *et al*. "Portal thrombosis in cirrhoties, A retrospective analysis". *Annals of Surgery* 203 (1986): 286-291.

5. Denninger MM., *et al*. "Causes of portal or hepatic venous thrombosis in adaults: the role of multiple concurrent factors". *The Pathology* 31 (2000): 587-591.

6. Majluf-cruz A., *et al*. "The incidence of protein c deficiency in thrombosis-related portal hypertension". *The American Journal of Gastroenterology* 91 (1996): 976-980.

7. Ruiz Arguelles GJ., *et al*. "Primary thrombophilia in Mexico. V. A comprehensive prospective study indicates that most causes are multifactorial". *American Journal of Hematology* 78 (2005): 21-26.

8. Calls D., *et al*. "Etiology of portal vein thrombosis in adaults. A prospective evaluation of primary myeloprolifrative disorders". *Gastroenterology* 94 (1988): 1063-1069.

9. Orozco H., *et al*. "Deficiency of protein c in patients with portal vein thrombosis". *Hepathology* 8 (1988): 1110-1210.

10. Huang S., *et al*. "Deficiencies in protein c and s in a patient with idiopathic portal hypertension accompanied by portal vein thrombosis". *Korean Journal of Transplantation* 16 (2010): 176-181.

11. Aiach M., *et al*. "Protein c and protein s deficiencies". *Seminars in Hematology* 34 (1977): 205-216.

12. Days SK., *et al*. "Chronic portal vein thrombosis due to combined deficiency of protein c and proteins". *Journal of Indian Medical Association* 109 (2011): 753-754.

13. Deshpande S., *et al*. "Portal vein thrombosis with protein c-s deficiency in non-cirrhotic patient". *International Journal of Medical Sciences* 4 (2016): 5061-5064.