

Infantile Hypertrophic Pyloric Stenosis - A Rare Cause of Hepatoportal Venous Gas

Usman Shakil¹, Talal Waqar and Najmi Usman

¹Department of Radiology, Combined Military Hospital Lahore, Pakistan

²Department of Neonatology and Pediatrics, Combined Military Hospital Lahore, Pakistan

³Department of Pediatrics, Combined Military Hospital Lahore, Pakistan

*Corresponding author: Usman Shakil, Department of Radiology, Combined Military Hospital Lahore, Pakistan, Tel: 00923335145963; E-mail: doc82me@yahoo.com

Rec date: Jul 02, 2014, Acc date: Sep 10, 2014, Pub date: Sep 12, 2014

Copyright: © 2014 Shakil U, et al. This is an open-access article distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.

Abstract

Hepatic portal venous gas (HPVG) is usually associated with necrotizing enterocolitis or bowel ischemia in infants but it's a rare finding with hypertrophic pyloric stenosis (HPS). In such cases, portal venous gas is a benign incidental finding and does not advocate any delay in the surgical treatment. We report this atypical case of 3 weeks old male infant with HPS having portal venous gas that was detected on abdominal ultrasound. Our patient with a history of term delivery was brought to the pediatric department of Combined Military Hospital Lahore on 25th of November, 2013 with complaints of vomiting and constipation for the last 4 days. Physical examination showed that the infant was lethargic and mildly dehydrated with soft and non-tender abdomen. Baseline blood tests revealed metabolic alkalosis with hemoglobin and total leukocyte count within normal limits. Plain xray abdomen showed a distended stomach with no signs of gut obstruction. Abdominal ultrasonography revealed a severely thickened and lengthened pylorus suggestive of hypertrophic pyloric stenosis. Moreover, ultrasound also showed multiple echogenic foci diffusely involving the both lobes of otherwise normal sized liver. Moving air bubbles were also detected in the extra and intra hepatic portal and splenic veins on dynamic scanning confirming the presence of portal venous gas. The infant underwent Ramstedt's pyloromyotomy for HPS. The infant showed a steady recovery and ultrasonography performed at 2nd post op day detected no signs of gas within the hepato portal veins.

Keywords: Pyloric stenosis; X-Rays; Ultrasonography; Portal vein; Laparotomy

Introduction

HPVG is generally considered an alarming radiological sign as it is associated with high mortality in all age groups [1,2]. The most common recognized life threatening conditions leading to portal venous gas include ischemic necrotic bowel, necrotizing enterocolitis, intra-abdominal abscess, inflammatory bowel disease, acute hemorrhagic pancreatitis, gastric ulcer and diabetic ketoacidosis [3-5]. However, studies have also documented few rare and benign causes that do not require aggressive medical or surgical treatment and in appropriate clinical settings can be managed conservatively.

The authors present this case of HPVG in an infant with gastric dilatation due to hypertrophic pyloric stenosis with portal venous gas and report this case due to the fact that portal venous gas is generally thought to be secondary to life threatening conditions carrying high mortality. However when associated with HPS, although rare and transient incidental radiological finding, should not deter the surgical treatment of HPS.

Case Report

A 20 days old male infant was brought to the pediatric department of Combined Military Hospital Lahore on 25th of November, 2013 with complaints of vomiting and constipation for the last 4 days. He was febrile having temperature of 102°F. His mother had a normal term pregnancy with no labor complications. Physical examination

showed that the infant was lethargic and mildly dehydrated. Blood test revealed metabolic alkalosis pH=7.58, bicarbonate 27.6 mmol/L, Na 130 mmol/L, K 3.9 mmol/L, Cl 88 mmol/L, Urea 1127 mmol/L (Normal: ≤ 8), and creatinine 85 U/L (Normal: <100). Hemoglobin and total leukocyte count were normal. The history and laboratory results were consistent with the clinical diagnosis of hypertrophic pyloric stenosis. Olive was not palpable. Abdomen was soft, non-tender with no signs of visceromegaly. Base line imaging studies included plain films abdominal ultrasonography. Plain abdominal x ray showed a distended stomach with paucity of gases distally. There were no signs of gas in the stomach wall or in the portal system. No evidence of intestinal obstruction was seen (Figure 1). Abdominal ultrasonography revealed a severely thickened pylorus, with a diameter of 13 mm, a length of 16.7 mm, and a muscular wall thickness of 4.5 mm. These sonographic features were suggestive of hypertrophic pyloric stenosis (Figure 2)

Ultrasound also showed multiple echogenic foci diffusely involving the both lobes of otherwise normal sized liver. Echogenic foci were also detected in the extra and intra hepatic portal and splenic veins. Moving air bubbles were seen within the hepato portal system on dynamic scanning confirming the presence of portal venous gas (Figure 3). The infant was transferred to the pediatric surgical department. Laparotomy with Ramstedt's pyloromyotomy was performed on the third day post admission. Peroperatively an olive was palpated with apparently normal stomach, duodenum and bowel. The infant showed a steady recovery and ultrasonography performed at 2nd post op day detected no signs of gas within the hepato portal veins.



Figure 1: Abdominal radiograph showing markedly distended stomach without any evidence of small or large gut obstruction.

Discussion

Detection of portal venous gas in infants raises the red flag due to its strong association with necrotizing enterocolitis but benign causes such as acute gastric dilatation, mechanical obstruction, endoscopic procedures, paralytic ileus, blunt trauma and barotrauma should be considered in appropriate clinical settings [3-7]. Infantile Hypertrophic pyloric stenosis (HPS) is the most common cause of non-bilious vomiting in infants and commonest surgical procedure performed on infants [8]. The typical infant presents with nonbilious projectile vomiting and dehydration (with hypochloremic hypokalemic metabolic alkalosis). However, not all children with HPS have electrolyte imbalance. Tutayetal [9] has shown that only 57% of children with pyloric stenosis have a low K. Barium and ultrasound are the radiological modalities used for diagnosis of HPS with accuracy of ultrasound approaching 100% [10]. HPS is one of the rare causes of portal venous gas. A retrospective study was carried by Hussain et al. [11] using Medline publications, covering a period from 1975-2008. These articles reported two hundred and seventy five patients with gas

in the portal venous system, out of which 9% cases were secondary to outlet obstruction and luminal dilatation. Ultrasound can clearly delineate gas in the portal venous system from pneumobilia by detection of echogenic foci in the periphery of the liver, extending within 2cm of the hepatic capsule, whereas gas in the biliary tract appears more central in the liver [12]. Different mechanisms have been proposed for the development of portal venous gas which include disruption of bowel mucosa in cases of ischemia and necrosis, gas forming bacteria in intraabdominal sepsis. In cases of pyloric stenosis and mechanical obstruction the intraluminal dilatation and pressure result in escape of gas within the venolymphatic channels [13]. HPVG secondary to pyloric stenosis is a transient process and generally resolves within days after decompression of stomach by nasogastric tube or following surgery [14]. Knowledge of this rare finding secondary to HPS is essential for the radiologists, clinicians and surgeons so that unnecessary panic is avoided and normal course of treatment of the disease is followed.

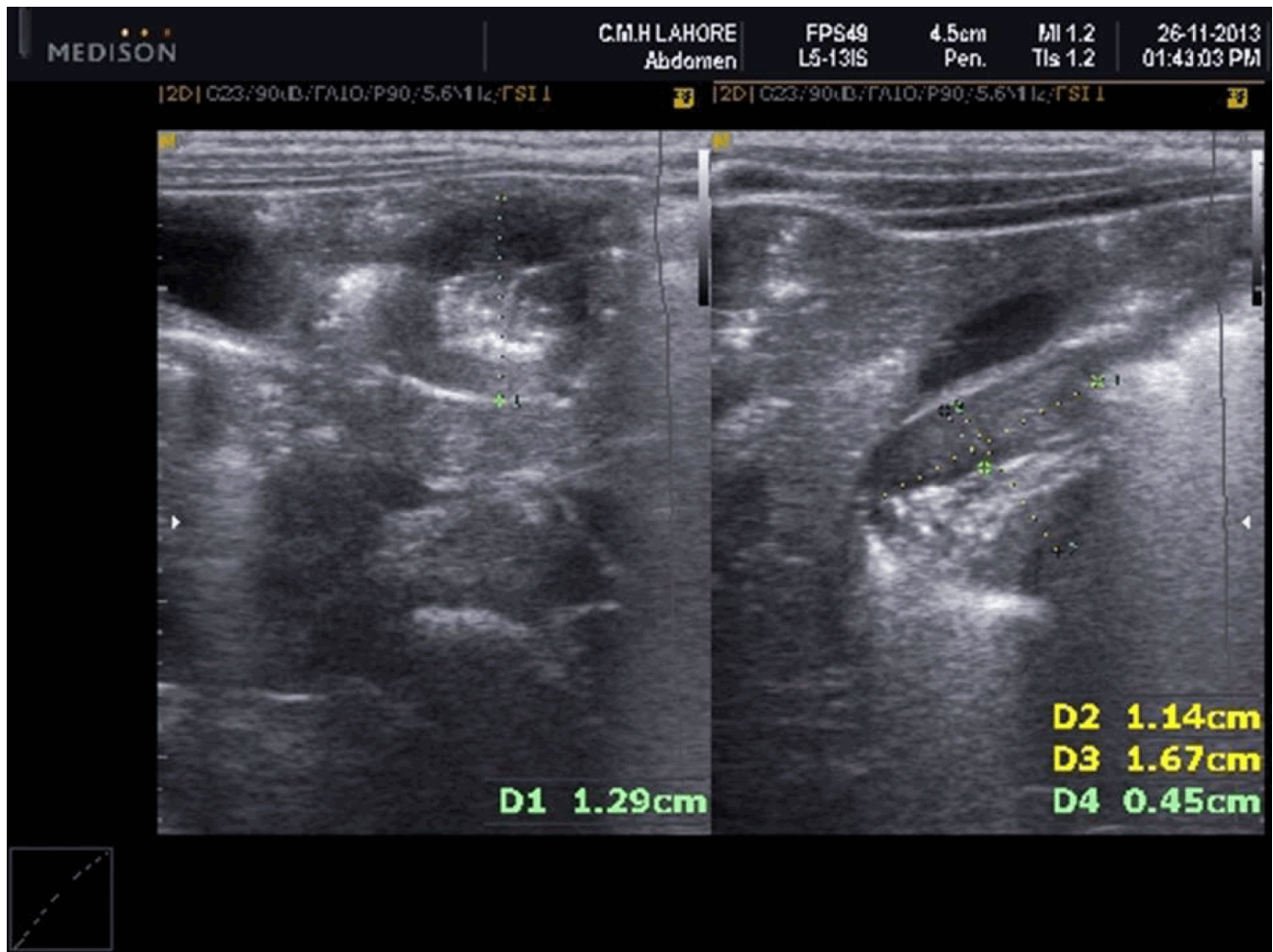


Figure 2: Ultrasound showing thickening and lengthening of pyloric canal suggesting HPS.

Conclusion

Although infantile hypertrophic pyloric stenosis is a relatively common entity but hepato portal venous gas is a rare complication of this disease and no case has been reported in local journals so far. Considering the graveness of the hepatoportal venous gas especially in

neonates which raises the suspicion of NEC or bowel ischemia, our case report clearly demonstrates that presence of portal venous gas even in infants can be due to benign etiologies. Careful clinicolab and imaging correlation is required to accurately diagnose and treat the underlying pathology.

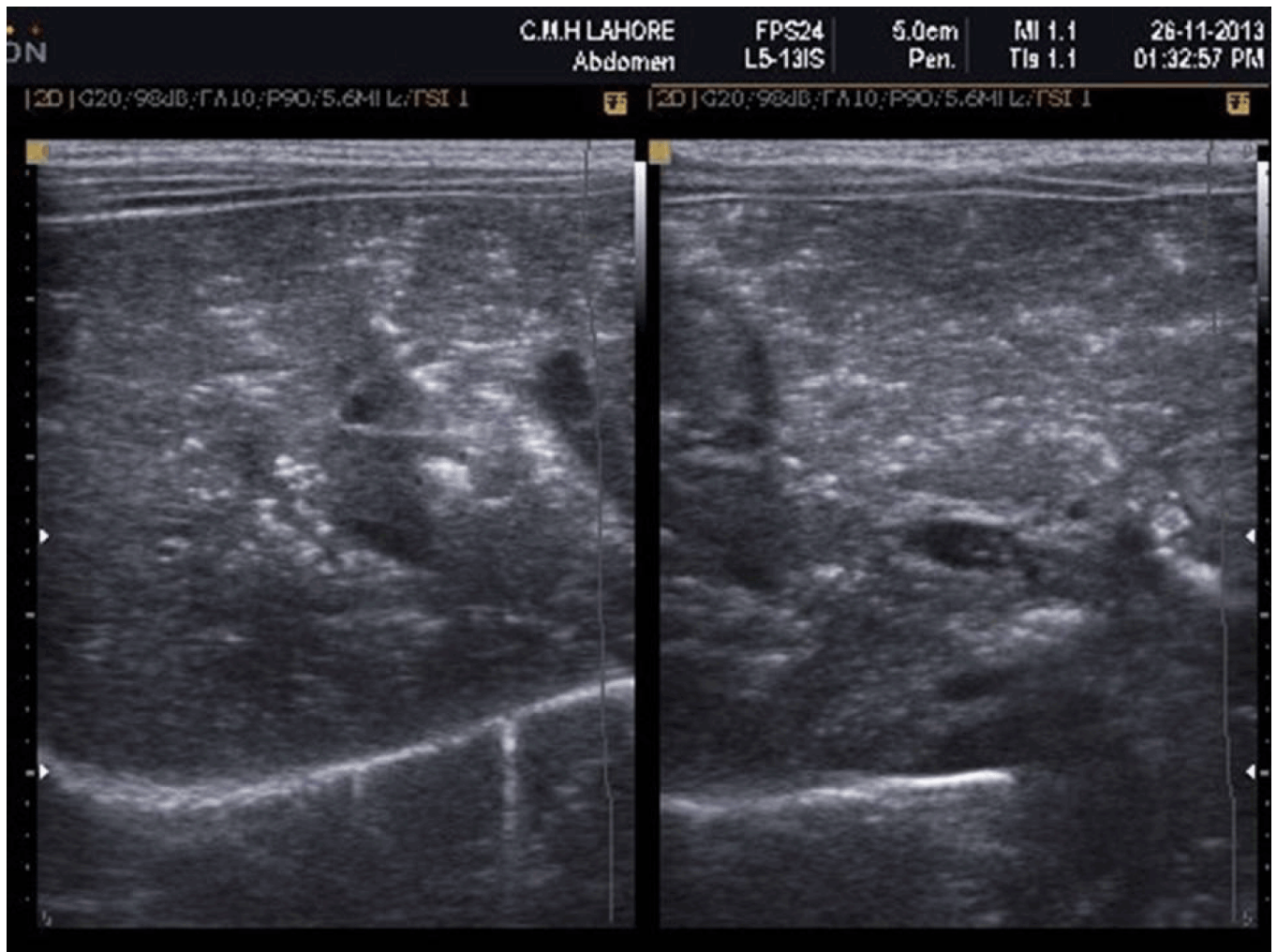


Figure 3: Ultrasound showing portal venous gas as echogenic spots within the liver and portal vein.

References

1. Allaparthi SB, Anand CP (2013) Acute gastric dilatation: a transient cause of hepatic portal venous gas-case report and review of the literature. *Case Rep Gastrointest Med* 2013: 723160.
2. Bhargava P, Parisi M (2009) Gastric pneumatosis and portal venous gas: benign findings in hypertrophic pyloric stenosis. *Pediatr Radiol* 39: 413.
3. Alqahtani S, Coffin CS, Burak K, Chen F, MacGregor J, et al. (2007) Hepatic portal venous gas: a report of two cases and a review of the epidemiology, pathogenesis, diagnosis and approach to management. *Can J Gastroenterol* 21: 309-313.
4. Chirica M, Scatton O (2006) Air in the portal venous system: radiologic differential diagnosis and etiology-specific treatment. *J Chir (Paris)* 143: 141-147.
5. Quirke TE (1995) Hepatic-portal venous gas associated with ileus. *Am Surg* 61: 1084-1086.
6. Radin DR, Rosen RS, Halls JM (1987) Acute gastric dilatation: a rare cause of portal venous gas. *AJR Am J Roentgenol* 148: 279-280.
7. Bani-Hani KE, Heis HA (2008) Iatrogenic gastric dilatation: a rare and transient cause of hepatic-portal venous gas. *Yonsei Med J* 49: 669-671.
8. Cheema HA (2003) Utility of endoscopy in infantile hypertrophic pyloric stenosis. *J Pak Med Assoc* 53: 482-483.
9. Tutay GJ, Capraro G, Spirko B, Garb J, Smithline H (2013) Electrolyte profile of pediatric patients with hypertrophic pyloric stenosis. *Pediatr Emerg Care* 29: 465-468.
10. Sivitz AB, Tejani C, Cohen SG (2013) Evaluation of hypertrophic pyloric stenosis by pediatric emergency physician sonography. *Acad Emerg Med* 20: 646-651.
11. Hussain A, Mahmood H, El-Hasani S (2008) Portal vein gas in emergency surgery. *World J Emerg Surg* 3: 21.
12. Sorantin E, Fotter R, Schimpl G (1995) Reversible portal venous gas in hypertrophic pyloric stenosis: detection by ultrasound. *J Ultrasound Med* 14: 699-701.
13. Sarti J, Kennedy A (2006) Portal venous gas in hypertrophic pyloric stenosis. *J Pediatr Surg* 41: 1935-1936.
14. Klar A, Gross E, Yahya RH, Akerman Y (2007) Portal venous gas and biliary sludge in infantile pyloric stenosis. *World J Pediatr* 3: 305-307.