etiology cannot be found in most cases, but prior bowel surgery may be associated.[3] Radiological investigations most commonly show a hypoechoic mass in the liver on ultrasound with cystic areas. A CT scan may show heterogenous mass in the liver, which is solitary in about 2/3rd of cases. The mass commonly shows peripheral enhancement with central hypodense areas suggestive of necrosis.[2-4] The mass is poorly demarcated from the surrounding liver parenchyma indicating local invasion mimicking a malignancy. Actinomycosis of the gall bladder is an extremely rare condition and to our knowledge, there are less than 30 reported cases in the literature. It usually occurs after the age of 50 years with no obvious gender predilection.[6] The most common symptom is pain in right upper quadrant of abdomen. Some cases may also present as acutecholecystitiswith pain, fever and tenderness. Ultrasound findings in most cases include a thickened gallbladder with or without features ofcholecystitis. Treatment with antibiotics is as important as surgery since actinomycosis is prone to recurrences. Penicillin G is the drug of choice and has to be given over a prolonged period of 2 to 6 weeks followed by oral amoxicillin or penicillin to be continued for 6 months to 1 year. Doxycycline, minocycline, tetracycline, clindamycin, erythromycin, and cephalosporinshave also been shown to be effective.[6]

CHINTAMANI B GODBOLE[1]
DHAVAL O MANGUKIYA[1]
RITU KAKKAR-KASHIKAR[2]
NILESH H DOCTOR[1]

Correspondence: DrNileshH Doctor
Departments of Surgical Gastroenterology[1] and Radiology,[2]
Jaslok Hospital and Research Centre,
GD Deshmukh Marg, Mumbai - 400026
Email: drnileshbela@gmail.com

References

1. Kyle Anderson, MD, Joseph M. Scherger, MD.Actinomycosis. First Consult. Elsevier.Jan–2012.
2. Wayne MG, Narang R, Chauhdry A, Steele J. Hepatic actinomycosis mimicking an isolated tumor recurrence. *World J Surg Oncol*. 2011;**9**:70.
3. Hayashi M, Asakuma M, Tsunemi S, Inoue Y, Shimizu T, Komeda K, et al. Surgical treatment for abdominal actinomycosis: A report of two cases. *World J Gastrointest Surg*. 2010;**2**:405–8.
4. Lai AT, Lam CM, Ng KK, Yeung C, Ho WL, Poon LT, et al. Hepatic actinomycosis presenting as a liver tumour: case report and literature review. *Asian J Surg*. 2004;**27**:345–7.
5. Kanellopoulou T, Alexopoulou A, Tanouli MI, Tiniakos D, Giannopoulos D, Koskinas J, et al. Primary hepatic actinomycosis. *Am J Med Sci*. 2010;**339**:362–5.
6. Acevedo F, Baudrand R, Letelier LM, Gaete P. Actinomycosis: a great pretender. Case reports of unusual presentations and a review of the literature. *Int J Infect Dis*. 2008;**12**:358–62.

Hirschsprung's disease and Budd-Chiari syndrome: a rare association

Introduction

Hirschsprung's disease (HD) is a congenital aganglionosis of the submucosal and myenteric neural plexuses, principally affecting the rectosigmoid area or varying length of the colon starting from the rectum.[1] Most cases manifest during the neonatal period but in rare instances, the disease is first diagnosed in adulthood.[2] On the other hand Budd-Chiari syndrome (BCS) or hepatic venous outflow obstruction (HVOO) is often an acquired disorder resulting from obstruction to the hepatic venous outflow at the level of hepatic veins or inferior vena cava.[3,4]We present here a 15-year-old girl who had both HD and BCS.To the best of our knowledge such an association has never been reported in literature before. We postulate that repeated enterocolitis due to HD might have produced a hypercoagulable state leading to BCS in this child.

Case report

A 15-year-old girl presented with constipation and abdominal distension since birth requiring enema every 2 to 3 weeks with a history of delayed passage of meconium for 3 days. Her abdominal distension used to get relieved completely after per rectal enema therapy, which she has been receiving since infancy. But for last 3 years the distension has persisted despite per-rectal enemas. She was diagnosed to have ascites 3 years back and treated withlarge volume paracentesis (LVP) and diuretics (spironolactone) which decreased her abdominal distension. There was no history of jaundice, encephalopathy or hematemesis. She received antitubercular therapy for 9 months along with indigenous alternative medicines off and on for the last 10 years for abdominal distension which did not make any difference. Her elder sibling had similar problem of

constipation since birth with delayed passage of meconium and required regular per rectal enema since birth. He died at 18 months of age from suspected enterocolitis. On examination the girl was emaciated with a weight of 20 kg after removing 4 litres of ascites (<5th percentile), height of 131 cm (<5thcentile), and BMI of 11.6 kg/m^2. She had mild pallor, bilateral pedal oedema but no icterus, clubbing, lymphadenopathy or raised JVP. Her abdomen was grossly distended with dilated, tortuous anterior abdominal wall veins, with flow from below upwards. However, there were no back veins. Her abdominal examination after large volume paracentesis revealed dilated bowel loops with visible peristalsis. The liver was just palpable below the right costal margin with a prominent left lobe, and a liver span of 8cm.The spleen was not palpable. She had free fluid in her abdomen and per rectal examination revealed an empty rectum. Investigations revealed a hemoglobin of 8g/dL, WBC 6600/mm^3, platelets 1.3 lakh/mm^3, serum creatinine 0.4mg/dL, serum bilirubin 0.5mg/dL, ALT 27 U/L, AST 40 U/L, serum alkaline phosphatise 360 IU/L, gamma GT 47 U/L, INR 1.4, total proteins 5.5g/dL, and serum albumin 2.5g/dL. Doppler ultrasongraphy showed attenuation of all three hepatic veins, with irregular hepatic outline and coarse echogenicity. The portal vein was 9 mm wide, IVC was patent and the patient had gross ascites with grossly dilated colon. Ascitic fluid analysis showed high serum-ascites albumin gradient (SAAG) of 1.3g/dL with no evidence of spontaneous bacterial peritonitis. Other etiological work up for chronic liver disease including autoimmune,Wilson's disease, chronic hepatitis B and C was negative. After ascites was controlled, the bowel loops became prominent and erect x-ray abdomen along with single contrast barium enema suggested HD (**Figure 1**). A triple phase contrast-enhanced CT abdomen suggested BCS (**Figure 2**). Anorectal

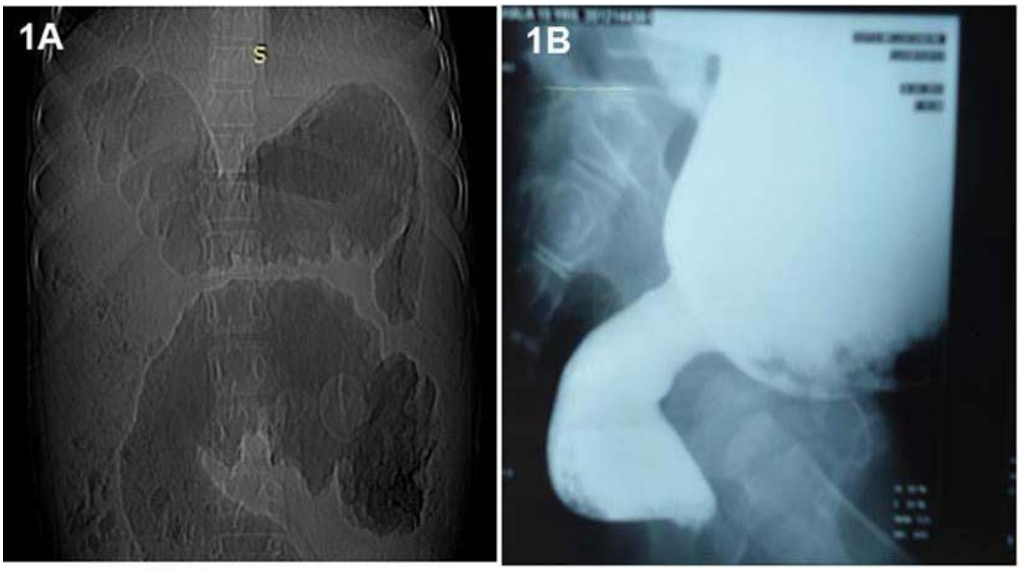

Figure 1: A) Plain x-ray abdomen (erect) showing grossly dilated colon; B) barium enema showing contracted rectum and grossly dilated sigmoid colon with a transition zone at the rectosigmoid junction.

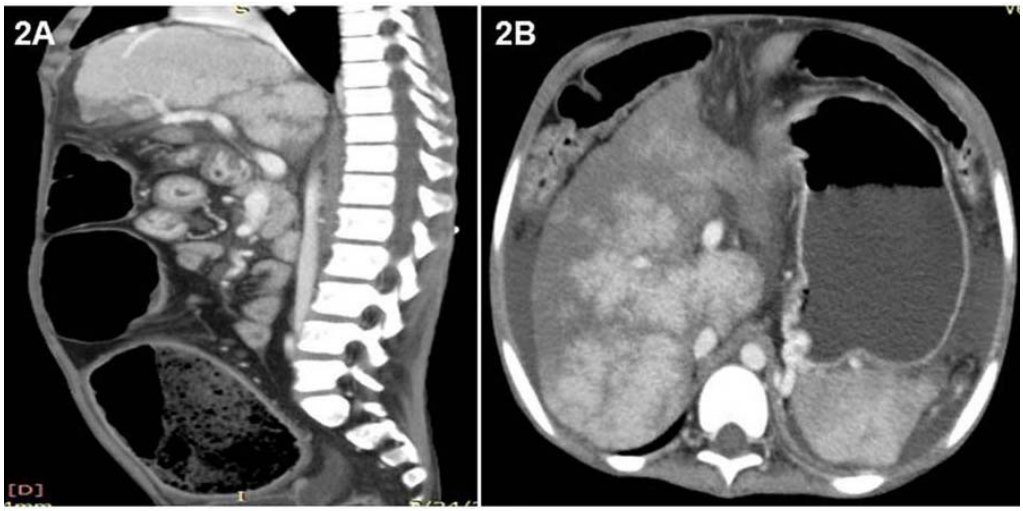

Figure 2: A) Longitudinal and, B) transverse views of thevenous phase of triple phase CT abdomen, showing attenuation of all three hepatic veins with a patent IVC and patchy uptake of contrast and a hypertrophied caudate lobe

manometry showed increased resting anal sphincter pressure (26mm Hg) and absent rectoanal inhibitory reflex (RAIR). Thus, confirming the diagnosis of Hirschsprung's disease with BCS. The child was started on regular rectal wash. She was discharged with a plan for hepatic venous stenting followed by definitive surgery for HD.

Discussion

The patient had Hirschsprung's disease since birth and she developed Budd-Chiari syndrome subsequently at 12 years of age. An extensive literature search did not reveal any similar case. It is highly unlikely that these two diseases are inter-related as one had a onset at birth (congenital disorder) and the other started almost a decade later (likely to be an acquired condition). However, long-standing HD (of 15 years duration) with repeated enterocolitis can give rise to a hypercoagulable state which in turn can produce BCS. Since most cases of HD get operated in early infancy, such late complications due to delayed referral of a symptomatic case have not been documented before. It has been observed that IVC obstruction with or without hepatic vein obstruction may be related to IVC web. In contrast, pure hepatic vein (HV) obstructions are mainly due to hypercoagulable state as coagulation factors are produced in the liver and their concentration is highest in HV.[5] This case had obstruction of hepatic veins and her IVC was patent, thereby suggesting hypercoagulability as a cause of BCS.

The association of total colonic aganglionosis with ileal atresia and congenital thrombophilia due to a mutation in the MTHFR C677T (methylenetetrahydrofolatereductase) gene has been reported. It has been postulated that ileal atresia due to vascular thrombosis in-utero might be responsible for prevention of cranio-caudal migration of neuronal cell from the neural crest, giving rise to total colonic aganglionosis.[6,7] However, in our case the child had classical ano-rectal HD and not total colonic aganglionosis. Though we could not investigate her for hereditary thrombophilia, the temporal correlation of events suggests she might have had acquired thrombophilia due to enterocolitis. Despite lack of reports this case highlights the need for early intervention in HD to prevent such complications.

VIKAS JAIN[1]
UJJAL PODDAR[1]
SHEO KUMAR[2]
SURENDER KUMAR YACHHA[1]
ANSHU SRIVASTAVA[1]

Correspondence: Dr. UjjalPoddar
Departments of Pediatric Gastroenterology[1] and Radiology,[2]
Sanjay Gandhi Postgraduate Institute of Medical Sciences (SGPGIMS),
Lucknow - 226014, India
Email: ujjalpoddar@hotmail.com

References

1. Belknap WM. The pathogenesis of Hirschsprung disease.*Curr Opin Gastroenterol*. 2002;**18**:74–81.
2. Roy CC, Silverman A, Alagille D. Congenital aganglionicmegacolon (Hirschsprung's disease). In: Roy CC, Silverman A, Alagille D, editors. Pediatric clinical gastroenterology. 4th ed. St. Louis (Mo): Mosby; 1995.p.503–15.
3. Valla DC. Hepatic venous outflow obstruction etiopathogenesis: Asia versus the West. *J Gastroenterol Hepatol*. 2004;**19**:S204–11.
4. Okuda K. Inferior vena cava thrombosis at its hepatic portion (obliterativehepatocavopathy). *Semin Liver Dis*. 2002;**22**:15–26.
5. Dilawari JB, Bambery P, Chawla Y, Kaur U, Bhusnurmath SR, Malhotra HS, et al. Hepatic outflow obstruction (Budd-Chiari syndrome). Experience with 177 patients and a review of the literature. *Medicine (Baltimore)*. 1994;**73**:21–36.
6. Guler N, Yapici O, Erdem D, Ogur G, Ozatli D, Bakir T. Acute complete splenic infarction in a patient with Hirschsprung's disease and literature review of complete splenic infarction. *Pathophysiol Haemost Thromb*. 2008;**36**:102–4.
7. deChadarevian JP, Bouie SM, Peddinghaus ME, Luck LR, Schwartz MZ, Prasad R, et al. Terminal ileal atresia, total colonic aganglionosis, and thrombophilia. *Pediatr Dev Pathol*. 2009;**12**:394–7.

An uncommon etiology of adrenal enlargement and fever in a patient of cirrhosis diagnosed with EUS guided FNAC

Introduction

We present an interesting case of alcoholic cirrhosis with bilateral adrenal enlargement, presenting with pyrexia of