celiac disease.[8] Adherence to a gluten-free diet for more than five years reduces the risk of malignancy to that of general population.[9] Weight loss, abdominal pain, change in bowel habits, vomiting, diarrhea, gastrointestinal bleed or an acute abdomen due to ulceration, obstruction or perforation are the most common presentations of EATL.[5,10] The tumor can be multi-focal in distribution; hence presence of synchronous lesions should be evaluated before therapy. Our patient presented with gastrointestinal bleeding and multi-focal disease.

Although CD is being reported with increasing frequency from our country, malignancy associated with it has been sparsely reported. In a series from a cancer institute, 2 cases of EATL associated with celiac disease have been reported amongst 170 gastrointestinal lymphomas.[6] In our cohort of 750 CD patients, this is the first case of EATL we have encountered. This case highlights the development of malignancy, especially EATL in patients with CD. With increasing prevalence of CD, clinicians dealing with this disease should be made aware of this complication.

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Biliary atresia with cytomegalovirus infection and its response to ganciclovir

Introduction

Biliary atresia (BA) is a condition in which the normal extra hepatic biliary system is disrupted. Biliary atresia affects approximately 1 in 10,000 – 15,000 births and occurs in 2 distinct clinical forms: fetal-embryonic (or syndromic) and perinatal (or acquired). The fetal embryonic form is characterized by early cholestasis, appears in the first 2 weeks of life, and accounts for 10-35% of all cases. In this form, the bile ducts are discontinuous at birth and 10-20% of affected neonates have associated congenital defects including situs inversus, polysplenia, malrotation, intestinal atresia and cardiac anomalies, among others. The perinatal form of biliary atresia accounts for the remaining 65-90% cases. This form is typically found in neonates and infants aged 2-8 weeks. Progressive inflammation and obliteration of the extra hepatic bile ducts occurs after birth. This form is not associated with congenital anomalies. Infection with cytomegalovirus (CMV), group C rotavirus and reovirus type 3 have been implicated in these patients.[1] Immune mediated ductal injury are important in the

pathogenesis of BA, and CMV infection may trigger such inflammation.[2] CMV is the most common cause of congenital infection in humans.[3] CMV infection is ubiquitous in India with high seroendemicity and nearly 99% adults showing IgG antibodies.[4] Neonatal CMV infection may occur due to intrauterine or perinatal exposure to CMV infected cervicovaginal secretions and breast milk.[5] Transmission occurs predominantly via primary maternal infection but can also occur due to reactivation of a CMV infection during pregnancy.[4] Neonatal hepatitis and biliary atresia have been reported as isolated clinical manifestations of congenital CMV infection.[2,3,5,6-9] Patients with neonatal hepatitis and CMV have been treated with IV ganciclovir and its oral prodrug, demonstrating variable outcomes.[3,5,6,7,9] However, the effect of ganciclovir in patients with biliary atresia and associated CMV infection has rarely been reported.[10] Fischler et al in 2002 treated two patients with biliary atresia and ongoing CMV infection for 3-7 weeks, of which one patient responded and another did not.[10] We report for the first time, two patients with biliary atresia and CMV infection who had a variable response to ganciclovir.

Case reports

Case 1

A 2½-month-old girl born of non-consanguineous marriage presented with jaundice since four days along with high colored urine but no clay colored stools. She was born full term with a birth weight of 2.5 kg. She was fully immunized for date and was on breast feeds. The mother experienced no antenatal problems. On examination, the child's weight was 4 kg, length 57 cm, and her vital parameters were normal. She had jaundice, rickets and hepatomegaly. Other systems were normal. Ultrasound abdomen showed hepatomegaly with non-visualization of gall bladder. HIDA scan showed normal uptake of tracer but no excretion of dye in the intestines even after 24 hours. An intraoperative cholangiogam showed no passage of dye in the intestines and the patient underwent a Kasai's surgery at 3 months of age. Operative liver biopsy showed an atretic gall bladder with multinucleate giant cells without inclusion bodies; and biliary atresia with bile channels measuring 150 microns at porta. Postoperatively the child continued to have jaundice. At six months of age, bilirubin was 11.3 mg/dl (direct bilirubin = 5.8 mg/dl) with elevated transaminases (SGOT = 266 IU/L, SGPT = 148 IU/L). A TORCH titre was done at that time which showed CMV IgM positive and CMV IgG level of 100 AU/ml. The child was started on oral ganciclovir (10 mg/kg/day) twice a day for 21 days and 5 mg/kg/day for next 21 days but there was no response. At 10½ months of age, the child showed failure to thrive (weight = 6 kg) with hyperbilirubinemia (bilirubin = 19.7 mg/dl), bleeding per rectum, portal hypertension and liver cell failure. The parents were advised a liver transplant for the child but the parents refused.

Case 2

A 1½-month-old girl born of non-consanguineous marriage presented with jaundice and clay coloured stools since birth. On examination, her weight was 3.5 kg and length was 60 cm. Her vital parameters were normal. She had jaundice with hepatosplenomegaly. Other systems were normal. Ultrasound abdomen showed hepatosplenomegaly with presence of gall bladder. TORCH titres showed positive CMV IgG (7.7 IU/L). In view of clay coloured stools, an intraoperative cholangiogram was done that showed absence of dye in intestines and the patient underwent Kasai's portoenterostomy at two months of age. Operative liver biopsy showed biliary atresia with periportal fibrosis and biliary channels sized 150 microns. Postoperatively, the child continued to have jaundice and clay coloured stools. At 4 months of age, the child was treated with intravenous antibiotics for sclerosing cholangitis and pneumonia. A repeat CMV IgG showed four fold elevation (30 IU/L). The child was then treated with oral ganciclovir for 6 weeks following which bilirubin decreased to 5.5 mg/dl and she started passing green coloured stools. At seven months of age, the child has chronic liver disease with portal hypertension and CMV IgG was 260 IU/ml.

Discussion

Treatment of CMV infection in children is controversial. However increasing number of studies indicate the necessity of treatment, especially in cases with symptoms of acute or chronic cholestatic hepatitis.[5-7,9,10] Ganciclovir has been used in only two patients with biliary atresia and CMV infection, of which one patient had a good outcome while the other did not.[10] Both our patients underwent Kasai operation at three and two months of age, respectively. However both had bile duct size of 150 microns which suggested poor prognosis.[11] Both patients continued to pass clay-colored stools postoperatively. Infact the first child still had jaundice at six

months of age (bilirubin = 11.3 mg/dl) suggestive of failed Kasai's operation. Thus, a beneficial effect of ganciclovir was unexpected as was eventually noted. This child developed liver cell failure at 10½ months. In the second child, ganciclovir was started two months after Kasai surgery because of active CMV disease and four-fold rise in CMV IgG, besides presence of pneumonia and sclerosing cholangitis. On treatment with ganciclovir, clay-colored stools disappeared and jaundice also decreased. However, since there was already significant liver damage, the child subsequently developed chronic liver disease with portal hypertension. Though the treatment was started late, but it seemed be partially beneficial in this patient. In a study by Fischler et al, of 28 patients with biliary atresia who underwent Kasai procedure, 11 had CMV infection. The authors found that survival with native liver among CMV infected and non-infected was 50% and 36% at four and 10 years follow-up, respectively, with no significant difference in long term outcome with respect to early CMV infection.[8] However the role of ganciclovir in treatment of biliary atresia with CMV needs further investigation and may offer different outcome for these patients. Thus randomized controlled trials are needed in patients with biliary atresia and associated CMV to examine the effect of ganciclovir treatment.

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A case of postcholecystectomy benign biliary stricture with atrophy-hypertrophy complex, mimicking a type IV Bismuth stricture

Introduction

Prolonged unilateral biliary obstruction or vascular occlusion can lead to atrophy of the ipsilateral lobe of liver with contralateral lobe hyperplasia. This is known as the 'atrophy-hypertrophy complex' (AHC). In the absence of a gall bladder, this phenomenon can be missed on preoperative magnetic resonance cholangiography (MRC). Performing a hepaticojejunostomy in the presence of AHC is technically difficult and is associated with greater blood loss and a higher incidence of stricture at the anastomotic site.[1] Hence preoperative diagnosis is important. We describe here a patient with AHC which went unrecognized during preoperative evaluation.

Case report

A46-year-old male patient who suffered an attack of acute cholecystitis 18 months earlier, underwent interval laparoscopic