Caroli’s Disease

Sir,

Caroli’s disease, a congenital biliary ductal disease, is a rare cause of cholestatic jaundice. With refinement in various noninvasive imaging techniques the diagnosis can be suspected early which can help in avoiding use of more invasive investigations.

We have a 17 years girl who had presented in August 2003 with progressively increasing jaundice of 30 days duration. She had prodromal symptoms of poor appetite and low-grade fever. Her past medical history was unremarkable. There was also no significant family history of any disease. Physical examination revealed deep icterus and hepatomegaly with liver span of 18 cm. Rest of the general physical and systemic examination was normal. Laboratory investigations showed normal hemogram and renal function, total serum bilirubin 33 mg/dl (0-1.0 mg/dl) with conjugated fraction of 28 mg/dl (0-0.25 mg/dl), alanine aminotransferase 950 U/L (0-41 U/L), aspartate aminotransferase 895 U/L (0-38 U/L), alkaline phosphatase 247 U/L (36-117U/L) and serum albumin 4 g/dl (3.5-5.0 g/dl). Investigations for hepatotropic viruses such as hepatitis A, B, C and E were negative. Autoimmune hepatitis markers were non-reactive. Serum ceruloplasmin was 68 mg/dl (20-60 mg/dl) and 24 hour urine copper was 124 mg/L. A diagnosis of acute viral hepatitis was considered and she was managed with symptomatic treatment. Jaundice started declining gradually initially and she showed recovery over initial 6 weeks but later her jaundice showed fluctuations and she developed ascites.

She was hospitalized in Jan 2004 with yellowness of eyes, moderate to severe pain in right hypochondrium and intermittent mild to moderate grade fever of 1 month duration. On admission she was anemic, jaundiced with no peripheral signs of chronic liver disease. She had hepatosplenomegaly but no ascites. Blood tests revealed mild iron deficiency anemia with leucocytosis. Liver biochemistry included total serum bilirubin 7.6 mg/dl (0-1.0 mg/dl) with direct fraction of 3.9 mg/dl (0-0.25 mg/dl), alanine aminotransferase 57 U/L (0-41 U/L), aspartate aminotransferase 68 U/L (0-38 U/L), alkaline phosphatase 795 U/L (36-117 U/L), serum albumin of 3.9 g/dl (3.5-5.0 g/dl) with total protein 7.4 g/dl (6.6-8.7 g/dl). Ultrasound abdomen showed hepatosplenomegaly. There were multiple cystic spaces of varying sizes in both lobes of liver with a classic central dot sign. However, IHBR were not dilated. Upper gastrointestinal endoscopy was normal. Liver biopsy showed prominent bile ductular proliferation, canicular cholestasis, portal fibrosis with bridging fibrosis and an attempt to nodule formation. MRCP revealed well defined saccular dilatations of the right and left hepatic ducts with early macronodular changes in the liver parenchyma suggestive of Caroli’s disease. (Fig. 1).

Caroli’s disease is a rare congenital cystic dilatation of the intrahepatic bile ducts. This ‘pure form’ was originally described by Jacques Caroli in 1958 and is associated with intraductal lithiasis and recurrent bacterial cholangitis, as was observed in our patient. Cholangiocarcinoma may occur as a late complication. In contrast, Caroli’s syndrome is associated with congenital hepatic fibrosis, portal hypertension and liver failure. In this case, the dilatation of intrahepatic biliary ducts is usually less prominent. These occur due to inadequate modeling of the ductal plates at all levels of the biliary tree.¹ These entities belong to the group of hepatic fibropolycystic disease which are usually associated with renal polycystic disease both infantile and adult.

Till date, sonography has been the modality of choice for noninvasive assessment of the biliary tree, with adjunct modalities being needed occasionally.² The classic form shows segmental saccular dilatations of the intrahepatic biliary radicals which may be interconnecting. No vascularity is usually identified within these spaces but central dot sign resulting from tiny vessels or portal radicals within the dilated biliary ducts has been reported. The demonstration of communication of these cyst-like structures with the hepatic or cystic ducts is cardinal for the diagnosis and is possible in most instances with ultrasound. CT scan is an invaluable adjunct that complements the above tests. It can identify cholangiocarcinoma and hepatic masses not visualized by ultrasound. Magnetic resonance cholangiopancreatography is a new, non-invasive imaging technique with proven diagnosis for Caroli’s disease.
disease. It is the first choice diagnostic tool especially whenever a purely diagnostic approach to the bile ducts is anticipated. In our case, though ultrasound abdomen showed cystic lesions but diagnosis of Caroli’s disease could not be firmly established and liver biopsy had to be done. The present case highlights the complementary role of ultrasound and MRCP in establishing diagnosis of Caroli’s disease.

A Sood*, Vandana Midha**, Neena Sood***, M Bansal**

*Department of Gastroenterology, **Department of Medicine, ***Department of Pathology, Dayanand Medical College and Hospital, Ludhiana – 141 001, Punjab, India.

Received: 11.8.2005; Revised: 16.6.2006; Accepted: 11.9.2006

REFERENCES


Granulocytic Sarcoma of Spine: An Unusual Initial Presentation of Acute Myeloid Leukemia

Sir,

Reported incidence of Granulocytic Sarcoma (GS) in AML is 2.9 –3.1%. However, granulocytic sarcoma of spine presenting with neurological symptoms as initial presentations of acute myeloid leukemia is rare. We would like to report three cases of GS who presented with neurological manifestations and managed with combination of chemotherapy and local radiotherapy.

Case 1: A 24 years old male presented with a history of weakness of all four limbs and retention of urine. Three weeks later he became anemic requiring blood transfusion and referred to our center. Examination revealed moderate pallor and sternal tenderness. Neurological examination showed the muscle power of grade 4 in all the limbs with extensor planter reflex and no sensory deficit. His hemoglobin was 8 g/dl; total leucocyte count (TLC) of 17,000/mm³ with 16 % blasts. Bone marrow examination confirmed the diagnosis of AML – M4. Magnetic resonance imaging (MRI) of spine showed extensive extradural deposits at multiple levels (C4-C6, T2-T4, T8-T10, L1-L5 and sacral roots). He received local radiation and induction chemotherapy. Post-chemotherapy bone marrow was in remission and patient had a complete neurological recovery. On last follow up at one year, patient was asymptomatic.

Case 2: An 18 years old male presented with low back pain of one-month duration followed by sudden onset paraplegia with retention of urine. Neurological examination showed grade 0 muscle power in both lower limbs with loss of deep tendon reflexes and complete loss of sensation below T₁, spinal segment. Complete blood count showed hemoglobin of 6.4 g/dl, TLC of 52,800/mm³ with 82% blasts and platelet count of 26,000/mm³. Bone marrow examination confirmed AML – M4. MRI spine showed soft tissue mass extending from T4-T9 with cord compression. He developed febrile neutropenia on day 14 of chemotherapy and expired.

Case 3: A 24-year-old male presented with back pain followed by weakness of both the lower limbs. Neurological examination revealed grade 3 muscle power with brisk deep tendon reflexes and bilateral extensor planter reflex. There was hypoesthesia below T₅ dermatome. Hemogram and spine radiogram was normal. MRI spine revealed epidural soft tissue mass extending from T₄ to T₆ with cord compression. Biopsy of spinal mass showed infiltration with blasts. Bone marrow revealed the diagnosis of AML – M4. He had complete neurological recovery following induction chemotherapy and radiotherapy.

Although the prevalence of GS is rare, factors such as certain chromosomal abnormalities t (8; 21), inv-16 and FAB subtype M2, M4 and M5 are associated with higher incidence of GS. All three of our patients were of AML-M4. Commonest site of cord involvement is thoracic spine. Spinal involvement in almost all of the cases reported has been of extradural in origin. Without the clue of circulating myeloblasts, the diagnosis of GS is usually delayed or not considered. Therapy of GS is not well defined because of rarity of cases. Therapeutic options available are chemotherapy, local irradiation and surgical decompression. GS are usually associated with poor prognosis even in otherwise favorable subgroup of AML patients.

DR Chouchary, M Bhattacharya, P Mishtra, M Mahapatra, R Kumar

Department of Hematology, All India Institute of Medical Science, New Delhi, India.

Received: 17.11.2005; Revised: 5.9.2005; Accepted: 21.9.2006

REFERENCES


Chikunguniya

Sir,

Re: The review article on chikunguniya by Sandhya Kamath et al, J Assoc Physician India, Vol. 54, Sept. 2006, P 725-26. The article on chikunguniya warrants
reference to an epidemic of Chikungunya in Tamilnadu, particularly in Madras Metropolitan area in 1964. A large study of the disease was conducted by my colleagues and myself in Govt. Stanley Hospital in collaboration with the Virus Research Lab of ICMR at Pune. 242 subjects of the disease were studied in detail: In 86 subjects, there was lab confirmation by isolation of the virus from acute phase sera or rising titre of HI antibodies or both. In 35 serology was negative. In 121 subjects the diagnosis was clinical.

Besides the clinical profile of the current epidemic described in the review article, our study had shown that the disease could produce serious and sometimes fatal results.

Hyperpyrexia occurred in 2 cases, one of which ended fatally. Haemorrhagic manifestations occurred in 11.6% of laboratory proven cases and in 2.6% of the lab negative cases. None were fatal. Neurological complications occurred in 5 cases: Encephalitis with External Ophthalmoplegia, Polynuropathy of the Guillaine Barre type in one case, transient Dysarthria in another case were observed.

Electrocardiographic changes suggestive of Myocarditis (excluding other causes like electrolyte abnormalities, hyperpyrexia etc) were seen in 3 out of 18 lab proven cases and 2 out of 37 lab negative cases.

The study had shown the possibility of the disease presenting with serious life threatening complications - besides the transient morbidity due to articular disease.

In a parallel study by the ICMR Virology unit from Pune and ourselves - a serological survey of asymptomatic subjects - 38.4% showed HI antibodies to Chikunguniya virus - indicating that a much larger population were infected, though had not developed the clinical disease.

Studies of haemagglutination inhibition tests on paired sera of febrile subjects showed 60.7% to positive for Chikunguniya. An interesting finding was that 6.9% of the sera showed positivity to Group B arboviruses and in 4.8% to both Chikunguniya and Group B Viruses. This may be expected as both the viruses have a common vector for transmission.

This letter may supplement the observation of the authors of the review article, in which this study of the 1964 epidemic in Madras could have found a place.

KV Thiruvengadam
Formerly Professor and Head of Department of Medicine, Madras Medical College and Physician, Govt. General Hospital, Chennai. Honorary Physician to the President of India. Received : 9.10.2006; Accepted : 13.10.2006

REFERENCES

Sir,

In Andhra Pradesh, we are witnessing an unprecedented rush of cases of fever with acute severe polyarthritis and we are treating all of them as Chikungunya. Some of the cases have been serologically confirmed.

Now Chikungunya has become so common in this area (coastal Andhra Pradesh) not a single family has been spared. I would like to make the following comments based on my own experience of treating >150 cases of Chikungunya fever over the past 6 months.

The onset of illness is so acute that the patient is disabled within hours with severe crippling polyarthritis and high grade fever with rash. Most of the patients don’t respond to routine analgesics or NSAIDs. In this situation, I see no reason why steroids should not be given, particularly as immune complexes are said to be involved in the pathophysiology of Chikungunya arthritis. In fact, dramatic improvement is seen with steroids, especially with high dose IV Hydrocortisone. The dosage can be quickly tapered off and substituted with oral steroids.

Many patients are having recurrent acute arthritis. Most probably the virus triggers an autoimmune type of reaction with pts presenting symptoms mimicking acute rheumatic fever and rheumatoid arthritis.

Uric acid levels tend to be high in all these patients. We have given them a trial of Allopurinol with variable results.

Blood Widal test and VDRL are positive, 48 hrs after the acute symptoms, but quickly become negative after 1 week.

Use of Hydroxychloroquine in these patients has not been giving good results.

I would like to get views from all our colleagues in South India, treating Chikungunya fever. We find that it is more rampant than reported and as correctly mentioned by the authors, the epidemic is continuing unabated in Andhra Pradesh, both in the urban and the rural areas.

AV Subba Rao
Senior Consultant Cardiologist, SV Heart Care Centre, 78 10 9 Syamala Nagar, Rajahmundry 533103.

Received : 13.9.2006; Accepted : 13.10.2006

REFERENCES