Case Reports

Ecchinococcus multilocularis causing alveolar hydatid disease liver: a rare occurrence in Indian subcontinent

Introduction

Alveolar hydatid disease (AHD) in humans is caused by infection with the proliferative larval stage of the fox tapeworm *Echinococcus multilocularis* which is distributed in the northern region of the earth (Europe, Russia, China, Japan and North America). The estimated number of patients worldwide is about 300,000.[1] When a human becomes infected by ingestion of eggs, the eggs develop into the metacestode stage in the liver, proliferate asexually, infiltrate to the peripheral parts of the liver and metastasize to other organs. Once the infection becomes successfully established, AHD is one of the most lethal helminthic diseases in humans.[2] The most effective therapy at present is surgical excision.

Case report

A 31 year old soldier at admission to a peripheral hospital for epigastric pain was found to have tender hepatomegaly with normal LFT, serum AFP and a marginally raised anti Echhinococcus IgG (8.85 U/ml). USG and CECT revealed large- partly cystic and solid SOL in segment VI and VIII of liver. FNAC attempt yielded 250 ml of purulent fluid.

On admission to this centre for persistence of symptoms, he was found to have tender hepatomegaly and normal routine laboratory investigations. ELISA for Echinococcus was positive for *E. granulosus*. USG and CECT imaging revealed non enhancing thin walled mass lesion in segment VI with fluid content and marginal calcification consistent with an abscess. Multiple hypo enhancing complex focal lesions in segment VII and VIII with stippled calcification were also noted. Patient was put on tab albendazole 400mg BD on admission and was taken up for exploratory laparotomy after four weeks with a clinical diagnosis of infected hydatid cyst. Per operatively, he was found to have large thickened cyst occupying complete segment VI and VIII. Excision of cyst with nonanatomical resection of liver involving segment V, VI and VIII was carried out. Patient made an uneventful recovery and histopathology report showed an infected hydatid cyst caused by *E. multilocularis*. He has been put on long term albendazole. Presently, he is asymptomatic and back to his duties.

Discussion

Echinococcus is a small tapeworm (~4mm) which affects humans and causes hydatid cysts. *E.multilocularis* and *E.granulosus* are the two most important species. The parasite requires two mammalian hosts for completion of the life cycle; the definitive hosts, dogs and foxes, and intermediate hosts, mostly rodents. Intermediate hosts become infected by ingestion of eggs, which hatch in the digestive tract, penetrate the small intestine, migrate to the liver and develop into metacestodes. The metacestodes form a huge number of minute cysts which consist of an outer laminated acellular layer and an inner nucleated germinal layer. In some minute cysts, brood capsules contain protoscolices. Connective tissues and inflammatory cells of the host surround the metacestodes. Man is an accidental host. Most common sites are liver (60%) and lung (30%).[3]

In AHD, the liver is the primary site of infection, and it closely mimics cirrhosis or carcinoma. Pain in right upper abdomen is the most common symptom, as seen in our patient, associated with progressive liver dysfunction that ultimately leads to liver failure. The progression can occur over weeks, months, or years. Distant metastasis is possible, and involvement of other organs (e.g., lung, brain, bone) can occur in as many as 13% of the patients.[4] In clinical cases of AE, the mortality rate is 50-60%.[5] This figure reaches 100% for untreated or poorly treated AHD.[6] Sudden death has been reported with AHD in asymptomatic patients (autopsy diagnosis). Specificity of ultrasound is 90% and CT scan is 100%.[7] Almost every serodiagnostic technique has been evaluated for echinococcosis, with variable results. The indirect hemagglutination test and the enzyme-linked immunosorbent assay (ELISA) have a sensitivity of 80% overall (90% in hepatic echinococcosis, 40% in pulmonary echinococcosis) and are the initial screening tests of choice. Immunodiffusion and immunoelectrophoresis demonstrate antibodies to antigen 5 and provide specific confirmation of reactivity. The ELISA test is useful in follow-up to detect recurrence.[8,9]

Radical surgery with complete excision of the lesion is the only chance for cure. In certain cases, total hepatectomy with transplantation has been performed as long as no extra hepatic disease is present.[10] Reemergence of the parasite in the transplanted liver and distant metastasis occurs under immunosuppression.[11] Partial resections of unresectable masses are considered to decrease the parasite load to aid the chemotherapeutic agents. Chemotherapy with benzimidazoles is used perioperatively for approximately 2 years in patients in whom radical resection is feasible because of possible undetected residual parasite tissue. In patients who undergo a partial resection, patients who are inoperable, or patients who have had a liver transplant, long-term chemotherapy is required (3-10 y).[12,13] Prognosis in AE is much worse. Cure is only possible with early detection and complete surgical excision. In patients in whom the latter is not possible, the addition of long-term chemotherapy has decreased 10-year mortality rates from 94% to 10%.[14]

In conclusion, AHD of the liver is endemic in some areas of the world and is rarely encountered in the Indian subcontinent. It remains a continuous public health problem in endemic countries. Although the disease is asymptomatic for many years because of the slow growth of the cyst, it is progressive, may cause life-threatening complications, and has the tendency to recur. Complete surgical excision remains the mainstay of the treatment for AHD.

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Heyde Syndrome

Introduction

The association between aortic valve stenosis and acute gastrointestinal bleeding was described by Heyde in 1958.[1] It has now been reported that enhanced proteolysis of the von Willebrand factor (vWF) by the elevated shear stress originating from passage of blood through the stenosed valve is responsible for this syndrome.[2] This is compatible with the observation that aortic valve replacement can eliminate recurrent blood loss and normalize von Willebrand multimers profiles in these patients.[3] We report a patient of Heyde syndrome in whom angiodysplasia of colon was confirmed as the source of bleeding and immunoblotting analysis revealed a decrease in high molecular weight vWF multimers.

Case Report

A 82 yrs old female was admitted with recurrent episodes of lower gastrointestinal bleed. She had received seven units of blood prior to admission. She had no history of chest pain, breathlessness or other symptoms of heart disease. She was evaluated elsewhere with upper gastrointestinal (GI) endoscopy, colonoscopy and contrast enhanced CT scan of abdomen, which were normal. A repeat colonoscopy in our hospital revealed multiple, discrete, vascular, flat lesions with bleed involving 10 cm of ascending colon, suggestive of angiodysplasia. On clinical examination she had a murmur suggestive of aortic stenosis. A 2-D Echo done revealed severe aortic stenosis. To clarify the association between the colonic hemorrhage and cardiac disorder we analyzed the vWF multimers by electrophoresis. The plasma level of the high molecular weight vWF multimers was significantly decreased. Considering her age and cardiac status, significant risk involved in surgery and anaesthesia and as she had stopped bleeding she has been managed conservatively. She is currently in good health in outpatient care for last six months without any further episodes of bleed.

Discussion

Colonic angiodysplasia is a degenerative ageing disease and accounts for up to 30% of patients presenting with lower-GI bleeding.[4] Most of these lesions are located proximal to the hepatic flexure. Many therapeutic strategies are described to treat angiodysplasia of the colon which include endoscopic methods that require direct contact by using monopolar/ bipolar probes or heat probe, and noncontact methods, eg, argon plasma coagulation. Injection sclerotherapy also appears to be effective.[5] Superselective embolization of the feeding artery at the bleeding site has also been proven to be effective for colonic hemorrhage. However, this procedure is technically difficult, and the risk of bowel infarction associated with embolisation proximal to the mesenteric border of the colon has been reported to exceed 10%.[6] Surgical resection is preferred for acute management of severe hemorrhage or for recurrent hemorrhage over a relatively short period accompanied by a large transfusion requirement.

Heyde syndrome is named after E. C. Heyde who in 1958 described ten cases of aortic stenosis and massive gastrointestinal bleeding for which he could discover no