Case Report

Hydatid Cyst Disease with Hepatic Encephalopathy and Multi-organ Failure: A Rare Case

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Abstract

Hepatic Encephalopathy is a serious and fatal complication of chronic liver disease and is broadly defined as an alteration in mental status and cognitive functions occurring in presence of liver failure. The diagnosis of hepatic encephalopathy is mainly clinical, sometimes aided by relevant laboratory (biochemical) investigations-if the infrastructural facilities permit. Histopathological findings in the brain and other organs are either meager or absent. Hydatid disease is caused by ingestion of eggs of Echinococcus species. Human beings are accidental intermediate hosts. Two third of hydatid cysts are found in liver. Though obstructive jaundice occurs as a complication of this cestode infestation, encephalopathy is rare or does not happen. The determination of the definitive cause of death may depend on elucidating the histological features of non apparent or equivocal macroscopic lesions. In this case presentation it has been attempted, to illustrate a rare postmortem finding of hydatid disease in a clinical presentation of hepatic encephalopathy where post-mortem histopathological examination shows multi-organ ischemic necrosis.

Key Words: Hepatic Encephalopathy, Hydatid Disease, Autopsy, Histopathology, Multi-organ failure

Introduction:

It is fortunate that all Forensic Pathologists have to deal not only with Criminal, Sudden, Suspicious, Accidental and Suicidal Deaths but with wide range of deaths due to natural causes.

Remember the famous comment of Prof. Bernard Knight "Involvement with natural deaths means frequent professional intercourse with clinicians and non Forensic pathologists with all the consequent benefits of cross fertilization of knowledge and ideas".

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When a team of Forensic and non Forensic pathologists keeps in touch with clinicians to deal a case of death due to natural causes, the result is surprising as we have seen in the present case study.

Case History:

One 27 years old male, Muslim by religion, was admitted to the district hospital on the first week of April'2011, with history of fall on the roadside followed by transient unconsciousness and recovery. He was suffering from chronic ill health, anorexia, vomiting, pain in the upper abdomen and yellowish discoloration of the whole body.

As his condition worsened in spite of treatment, the said patient was transferred to Calcutta National Medical College at Kolkata on the first week of May' 2011. But the patient succumbed to his illness on mid of June 2011.

The cause of death as recorded in the death certificate was "Multi-organ failure in a case of Hepatic Encephalopathy". The dead body of the said subject was sent to NRS Medical College for autopsy.

The clinical entities like "Multi-organ failure" and "Hepatic Encephalopathy" can hardly be documented through routine autopsy procedures. The role of supplementary ancillary investigations is very much relevant in this context. In the present case study the incidental

finding of Hydatid Cyst Disease further complicated the scenario. In this vexing situation we have attempted to establish a causal relationship between the clinical findings, autopsy findings and results of histopathology.

Autopsy Findings:

External Findings:

- Deep yellow discoloration of sclera, conjunctiva and skin of the whole body.
- Surgically made wound at the right flank of the abdomen for drainage of ascitic fluid.

Internal Findings:

- Brain and both lungs were pale and edematous.
- Spleen was blackish in colour and was grossly enlarged-weighing 800 g. multiple small, hard, whitish areas (calcifications) were found over the costal surface. (Fig. 3)
- Liver was grossly enlarged, weighing 2400 g, yellowish coloured and firm in consistency. Multiple small, hard, whitish (calcifications) nodules were found at places over both the lobes.

Over the diaphragmatic surface of the right lobe there was a whitish hard area (calcified) of 1″ x 1″, which on cut section shows multiple small cysts within the substance of the liver. (Fig. 1& 2)

Histological Findings:

- Brain- Congestion, dilatation of meningeal vessels accompanied by focal areas of ischemic necrosis (infarct). (Fig. 4)
- Lungs- Patchy bronchopneumonia with serous fluid and hemosiderin laden macrophages in the alveoli. Foci of ischemic necrosis (infarct) seen at places. (Fig. 5)
- Heart- Heart muscles separated by edema fluid with dilatation of blood vessels. Foci of ischemic necrosis (infarct) seen at places. (Fig. 6)
- Liver- Multiple focal areas of ischemic necrosis (infarct) seen. Rest of the liver tissue shows inflammatory cell infiltration and degenerative changes. (Fig. 7)
- A piece of liver tissue (8cm x 6cm x 4cm) - Cut section shows large cystic areas of size 4cm x 4cm with multiple vesicle-like structure. On microscopic section it shows histology of hydatid cyst with multiple daughter cysts along with pricyst and adjacent liver tissue.

Cause of Death:

Death was due to “Multi-organ Failure in a case of Hepatic Encephalopathy.”

Discussion:

Cestodes are segmented worms. The adults reside in the gastrointestinal tract but the larvae can be found in almost any organ. Human cestode infections are of two major clinical groups. In one group human are the definitive hosts, with the adult tapeworms living in the gastrointestinal tract (e.g. T. saginata, Dyphllobothrium, Hymenolepis etc.). In the other group, human are intermediate hosts with larval stage parasites present in the tissues (e.g. Echinococcus, Spargonium etc.). [1, 2]

Echinococcosis is an infection caused in human by larval stage of the E. granulosus complex. E. granulosus complex parasites are prevalent in areas where livestock is raised in association with dogs. The parasites are found in all continents with areas of high prevalence in China, Central Asia, and Middle East, the Mediterranean region, Eastern Africa and parts of South America. [1, 2]

The definitive hosts for E. granulosus are canines (dogs) that pass egg in their feces. Human are *accidental intermediate hosts* and get infected by ingestion of food contaminated with eggs of E. granulosus shed by dogs. Embryos escape from the eggs, penetrate the intestinal mucosa, enter the portal circulation and are carried to various organs. Two thirds of E. granulosus cysts are found in liver, 5-10% in the lungs and rest in the brain, bones or other organs. Before developing into cysts, the larvae lodge in the capillaries of various organs and incite inflammatory reaction.

Many such larvae are destroyed while some larvae develop into fluid filled unilocular *Hydatid cysts* that consist of an external membrane and an inner germinal layer.

Daughter cysts develop from the inner aspect of germinal layer as do germinating cystic structures called Brood Capsules. New larvae called protoscolices develop in large numbers within the brood capsules. The cysts expand over a period of years while some may degenerate during the process.

The degenerated cysts are replaced by scarring followed by calcification. Initially the Echinococcus cysts are asymptomatic.

But as they expand they produce features of space occupying lesions, impairing functions of the organ. There may be leakage of cystic fluid due to rupture (accidental or otherwise) producing fever, urticaria, pruritus, eosinophilia or anaphylaxis. In the liver it characteristically present as slow growing tumor with progressive destruction of liver tissue, compression of bile duct or leakage of cystic

fluid into the biliary tree (which may mimic recurrent cholelithiasis) and biliary obstruction that can result in jaundice.

The lesion may infiltrate adjoining areas or may metastasize to spleen, lungs or brain. [1-4] Hepatic Encephalopathy is a disturbance in functions of central nervous system because of hepatic insufficiency due to either acute or chronic liver failure.

Alteration of consciousness is its most relevant manifestation and is influenced by concurrent clinic-pathological factors such as inflammation, hypoxemia, and gastrointestinal hemorrhage or electrolyte disturbances. [3-7]

A large body of work points at ammonia as a key factor in the pathogenesis of hepatic encephalopathy. In acute and chronic liver diseases increased arterial levels of ammonia are commonly seen.

Furthermore the abnormalities in the glutaminergic, serotoninergic, GABA-ergic and catecholamine pathways have also described in experimental hepatic encephalopathy.

Depletion of zinc and accumulation of manganese may play some role. But it is a prudent fact that inflammation elsewhere in the body may precipitate encephalopathy through the action of cytokines and bacterial polysaccharides on astrocytes.

It has also been well recognized that bacterial and other infections/inflammations lead to the systemic inflammatory response syndrome, sepsis and Multi-organ failure. [7-12]

In the present case postmortem finding of Hydatid cysts is incidental. But considering the findings all together and after going through authentic texts and reviews it can be proposed that the Hydatid cyst disease may have contributed towards pathogenesis of hepatic encephalopathy and multi-organ failure in the present case. It is noteworthy to mention here that Dulger A C et al, [13] reported one case of Hepatic Encephalopathy where the later was precipitated in connection with Budd-Chiari Syndrome that resulted from infection with Echinococcus Multilocularis.

In that case the worm- damaged hepatic venous system led to biliary cholangitis, sepsis, elevation of liver related transaminases, higher cholestatic enzymes, hyperglobulinemia, eosinophilia, higher C-reactive protein levels and ultimately hepatic failure as a result of massive liver destruction. [12, 13]

Conclusion:

1) Autopsy of cases of death due to diseased condition is basically a challenge for the autopsy surgeon.
2) Methodical external and internal examination (naked eye) and meticulous dissection of the organs may give us vital clues as to the cause of death.
3) Even in smallest doubts histopathology may give us surprising outcomes.
4) In the arena of Forensic Pathology, histological examination may be suggested as compulsory for every autopsy, if infrastructural facilities permit, as to determine specific cause of death.

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Fig. 1: Cut Section of Liver

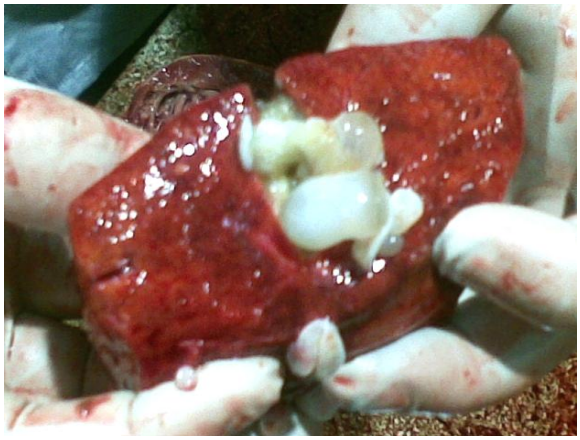

Fig. 1: Calcified Area over Liver

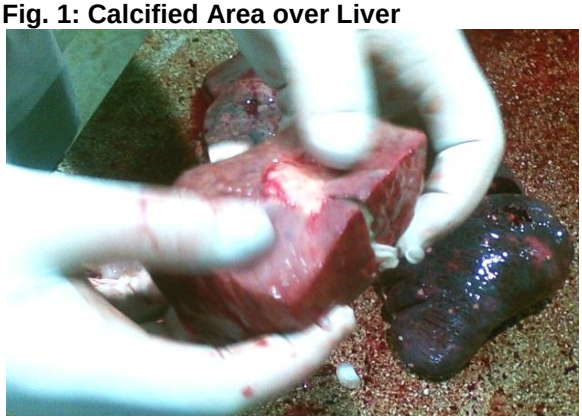

Fig. 3: Calcified Areas over Spleen

Fig. 4: Congestion of Brain with focal areas of ischemic necrosis

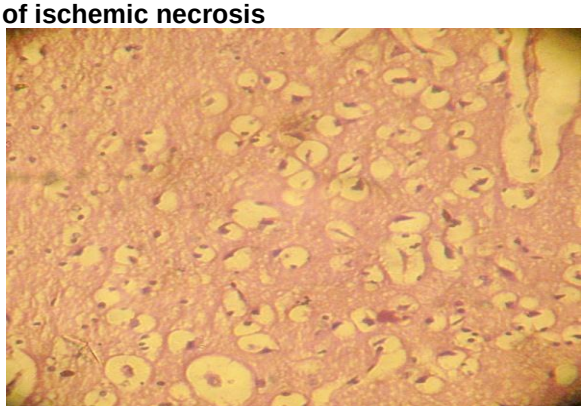

Fig. 5: Lung

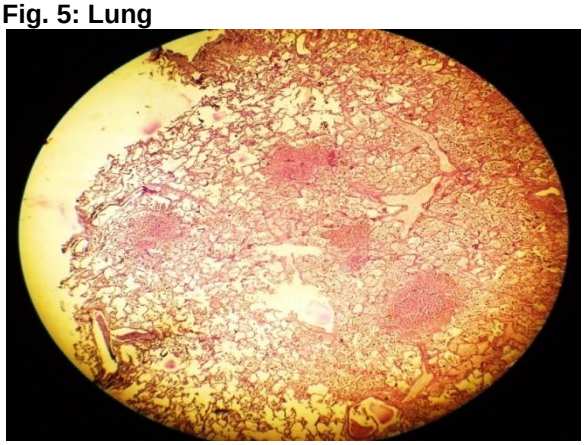

Fig. 6: Heart

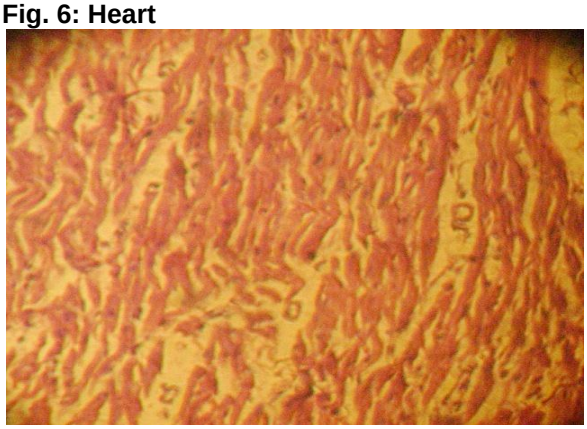

Fig. 7: Liver

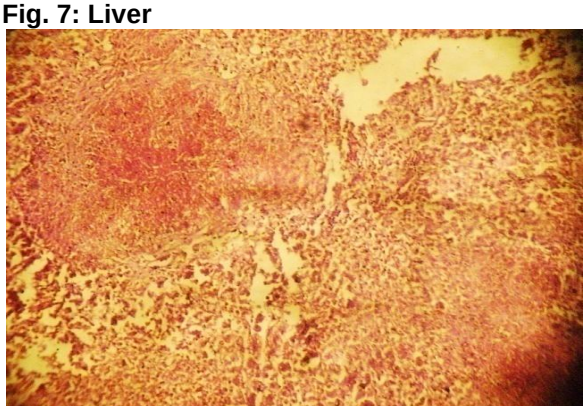

Fig. 8: Hydatid Cyst with Multiple Daughter Cysts along with Pericyst and Adjacent Liver Tissue