Cerebral Hydatidosis
Clinical Case Report with a Review of Pathogenesis

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Cerebral hydatidosis or echinococcal cyst of the brain, uncommon even in endemic areas, is a rarity in North America. However, the prevalence of echinococcosis in the new state of Alaska increases the possibility of diffusion of this condition in the United States. The following case represents the third report of cerebral hydatidosis operated upon in the United States and the only one with metastatic cerebral localization. In this case there was a delay of 11 years between the onset of cerebral symptoms and their correct diagnosis. In our opinion this delay may have been caused by the sparse knowledge of cerebral hydatidosis in this country. To help supply such information this paper will include a review of the pathogenesis, parasitology, diagnosis, prophylaxis, and the treatment of echinococcosis.

Case Report

N.H.H. #A80996. J.P., born in Italy in 1898, arrived in this country in 1914, and never thereafter left the United States. His health apparently was good up to 1937 when he had precordial pain; he was admitted to the New Haven Hospital, where roentgenograms of the chest revealed three large cysts in the pericardium, diagnosed as "calcified echinococcus cysts of the pericardium." Surgical removal of the cysts, recommended by the hospital staff, was refused by the patient. This part of the history has been reported by Tracy.48

In 1940, J.P. fell unconscious twice during a period of 4 months and was treated with Dilantin. Thereafter he suffered from sleeplessness, severe pounding headaches, and grand mal seizures, which recurred two or three times per month. In 1946 right hemiparesis and dysphasia developed, which persisted to the end of the patient's life. In 1948 he suffered from symptoms of acute intestinal obstruction, requiring emergency laparotomy, which revealed three cysts in the jejunal mesentry. Pathological sections of the cysts showed "a dense acellular fibrous tissue plus a layer of looser, more cellular fibrous tissue showing infiltration of a small number of small round cells and eosinophils. Neither these sections nor the preparations of the cystic fluid showed any evidence of parasites. Diagnosis: Multilocular cysts of mesentry with calcification" (see Postmortem Diagnosis below). The patient recovered uneventfully.

On June 18, 1951, J.P. was examined at the Out-patient Clinic of the Connecticut Valley Hospital† by one of the authors (C.M.A.). The patient complained of feelings of depression and persecution superimposed on his long-standing cardiac, gastrointestinal, and neurological symptoms. He had been free of epileptic seizures for about 6 months on an increased dosage of Dilantin.

Psychiatric examination showed unsystematized paranoid delusions and marked depression. However, on the basis of the symptoms of headache and hemiparesis with a history of cardiac echinococcosis, it was firmly believed that a condition of metastatic cerebral hydatidosis was present. The patient therefore was referred to the Neurosurgical Service of the New Haven Hospital with the diagnosis of "cystic infestation of the brain," and was admitted there on June 19, 1951.

Examination. Rate of pulse was 60, temperature was normal, and respiratory rate was 38. Blood pressure was 140/100. He had mild dysphasia, slight cardiac enlargement, systolic precordial murmur, weakness, hyperreflexia, and Babinski's sign on the right.

Laboratory Tests. Counts of red and white blood cells were normal; differential was not recorded. Spinal-fluid pressure was 120 mm. of water. The fluid contained 35 mg. per cent protein, Kahn reaction was negative, and colloidal gold curve was normal.

Electroencephalogram showed a focus of slow waves in the left occipital region extending to the left central leads. Flat roentgenograms of the skull were uninformative, but a pneumoencephalogram revealed a large mass in the left posterior parietal

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region with atrophy of the right hemisphere. An arteriogram showed this mass to be relatively avascular.

Operation. On July 5, 1951, under local anesthesia, a left parieto-occipital craniotomy was performed. Upon elevation of the dura mater a cystic mass was disclosed in the left posterior parietal region and was enucleated intact. No other superficial cysts could be detected.

Postoperative Course. Immediately following the operation the patient's dysphasia improved and his right hemiparesis disappeared totally. The next day, however, these symptoms reappeared and became more severe during the following 2 days. On July 10 and 11 the symptoms again subsided partially, but on July 12 the patient's general condition became critical, and he lapsed into coma. Epidural hemorrhage being suspected, the operative flap was reelevated, but only 10 ml. of blood were found and removed. Probing of the brain for deep-seated hydatid cysts was not done because of the risk of death from anaphylactic shock. The patient failed to regain consciousness and expired on July 14, 1951.

Surgical Pathology. The operative specimen consisted of a globular vitelline cyst, approximately 4 cm. in diameter, having two daughter-cysts, each 1 cm. in diameter attached to one of its edges. Microscopic sections showed that the wall of the cyst was composed of a thick outer amorphous keratinous layer separated by a well-defined basement membrane from a central epithelial layer of granular germinal cells. No brood capsules, scoleces, or hooklets were found in the cystic cavity. The cerebral tissue attached to the cyst showed chronic inflammatory reaction with marked fibrocytic and lymphocytic infiltration and moderate glial proliferation. More peripherally the cortex appeared normal with moderate vascular congestion. Diagnosis: "Parasitic cyst."

Necropsy. The surfaces of both hemispheres showed flattening of the gyri, without pressure cones. The left occipital lobe was occupied by a large cystic mass covered by 2-3 mm. of cortex and separated from the posterior two-thirds of the left lateral ventricle by a thin membranous layer (Fig. 1). The cystic cavity was lined by a membranous wall that contained several small cysts, grossly typical of echinococcal daughter-cysts. The left lateral ventricle was greatly enlarged. The brain stem showed an area of hemorrhage extending from the pons to the left lateral surface at the level of the mammillary bodies.

The left cardiac ventricular wall contained three cystic areas, each approximately 6 cm. in diameter, confluent at their common junction. One of these cysts had a calcified wall. The remaining cysts were lined by a fibrous wall and their cavities contained numerous small transparent cysts grossly typical of echinococcal daughter-cysts (Fig. 2). The myocardium was white and fibrous in the areas surrounding the cysts but elsewhere of normal color and consistency.

The mesentery of the first part of the jejunum contained several hydatid cysts, each approximately 2 cm. in diameter. No other pathological signs were found.

Postmortem Diagnosis. Echinococcal cysts of the mesentery, heart, and brain; probable cause of death, cardiorespiratory paralysis from bulbar hemorrhage.

Review of Pathogenesis

Splanchnic Hydatidosis. The echinococcal embryo burrows its way through the mucous and muscular layers of the intestinal wall and
reaches the lumen of the lymphomesenteric and portal capillaries. The mesenteric lymph nodes and the subserosal lobules of the liver are the usual first stations of implantation of the embryo. There, unless destroyed by cellular reaction, it undergoes cystic development. The cellular reaction consists mainly of mononuclear, eosinophilic, and phagocytic giant cells and results in a hydatid follicle. Those embryos not filtered by the lymphomesenteric and hepatic capillary barriers reach the right cardiac cavities through the inferior vena cava and then the lungs through the pulmonary arteries. Here again, if not destroyed by follicular reaction, they will either evolve into cystic forms or escape the capillary barrier to reach the left cardiac chambers. From the left ventricle the embryos either reach the myocardium through the coronary arteries or enter the aorta to reach other splanchnic organs through the general circulation, or to reach the brain through the carotid arteries.

*Cerebral Hydatidosis.* Two different histogenetic types (embryonal and scolecal) of cerebral hydatidosis having different names and clinical characteristics are recognized as follows:

1. One caused by embryos which escaped lymphomesenteric, hepatic, pulmonary, and coronary barriers (direct or embryonal implantation), termed primary cerebral hydatidosis.

2. The other caused by scoleces from ruptured fertile cysts (indirect or scolecal implantation), called metastatic or secondary cerebral hydatidosis.

Primary cysts, because of their embryonal origin, usually are fertile, that is, they contain numerous echinococcal headpieces or scoleces. Secondary cysts (also called acephaloceysts) usually are sterile because of the typical infertility of scoleces from which they arise.

Primary cysts usually are solitary and occur mostly in children under the age of 15. Their development is rapid and extensive, with abrupt dramatic symptoms, which result in fatality over a course rarely longer than 4 years in the absence of operative treatment.

Metastatic cysts, much rarer than the primary, usually are multiple and generally are accompanied by metastatic cysts in other organs of the aortic circulation, particularly in the spleen and the kidneys. Slow and insidious in development, with a latent course of about 5 years, they usually are fatal within 3 years from the onset of symptoms, in the absence of operative treatment. Their usual multiplicity offers an operative prognosis more guarded than that of primary cysts.
The present case with cerebral symptoms of 11 years' duration, though not so long as the case of Manceaux et al.,24 is definitely exceptional.

Metastatic cerebral hydatidosis usually is caused by myocardial cysts rupturing into the left ventricle. Myocardial cysts generally are multiloculated because of the impact of the ventricular contractions on their walls; however, they cause remarkably slight interference with cardiac function. By the time cardiac or pericardial hydatidosis is diagnosed, metastatic cerebral hydatidosis must be suspected.

Statistical Survey

Among the cases reported in North America in which the nationality of the patients was known, 95 per cent were immigrants.5,31 Of autochthonous human hydatidosis at least 40 cases have been reported in this country.5,28,30,33,46 Of 6 patients reported by Birch and Anast 3 were immigrants and 3 were born in the United States. Hydatid disease has been found to be endemic in the lower Mississippi valley.5,25 Katz and Pan48 found that in most published cases of echinococcosis in this country the patients were immigrants. They indicated that of 556 cases reported between 1900 and 1958, only 38 patients could be considered native.

The incidence of cerebral localization in hydatid disease is difficult to appraise from the literature. It has been reported variously to be from 0.5 per cent to 2.5 per cent with occasional reports giving higher figures.12,37,42 In several sizeable series of echinococcosis (60 to 300 cases), no cerebral cases were found.14,16,18,20,27,44 Among 1,802 patients reported to the Australasian Hydatid Registry from 1930 to 1945, Cole9 found 16 cerebral cases, an incidence of 0.9 per cent. Among 596 cases of hydatid disease reported in North America from 1808 to 1950 Magath25 found 11 cases, or 1.8 per cent, of cerebral cysts. Dew17 found the incidence in a large series to be 2.1 per cent without regard to type or age; but if the patients in this series under the age of 15 were considered separately, brain cysts in them were found to be seven times as frequent as in adults.

Parasitology

Details of the life cycle of Echinococcus granulosus or Taenia echinococcus are available in any text or standard work on the subject,7,12 but the following aspects relating to the disease in North America are not so readily available.

In most countries the usual cycle is dog-sheep-dog and dog-man. In North America, according to Riley,39 the reservoirs of disease are mainly sylvatic, the usual definitive carnivorous host being the wolf, and the usual herbivorous intermediary host the moose; with caribou, deer, and reindeer substituting occasionally.13,33 In some southern and western regions of the United States there occurs a life cycle involving the fox, and, rarely, the dog, as the definitive hosts, with hogs, sheep, and cattle as the intermediary hosts. Magath31 stated that when dogs are the predominant definitive hosts, sheep and man, especially children, will be the cyst-forming hosts; but when wild carnivora are the definitive hosts, wild herbivora and pigs are the likely intermediates. Dorman and Olson13 found the condition most frequent in the regions of the United States where herbivora (sheep, cattle, horses) are herded. With the addition of Alaska as one of the States the dog will assume an increasing importance as a source of infestation,43 and this will be discussed further under the heading of Prophylaxis.

Diagnosis

The symptoms of cerebral hydatidosis are similar to those of brain tumor.22,39,47 In endemic areas this diagnostic possibility must be considered seriously, especially in children, who are affected by this condition more frequently than adults.23 The presence of cardiac echinococcosis should always alert the clinician to the possible occurrence of metastatic cerebral cysts.

Laboratory Tests

The following tests generally are used in hydatid disease: (1) Casoni, or intradermal allergic skin test. (2) Boyden, or tannic acid hemoagglutination. (3) Weinberg, or complement fixation test (blood serum, spinal fluid),
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(4) Ghedini, or precipitation test (blood serum, spinal fluid).

In cerebral hydatidosis the main diagnostic tests are:

1. Electroencephalogram. This test may be helpful in localization, but the presence of a region of electrical silence or low voltage described as characteristic for hydatid cysts of the brain was not present in our case and reportedly has been encountered only rarely by others.

2. Pneumoencephalogram. This test may be used only in patients free from intracranial hypertension.

3. Arteriogram. This is a safe and useful test in expert hands.

Ventriculography has been recommended but experienced students of this problem oppose its use for fear of puncturing the cyst and disseminating the disease. A recent monograph recommends that it be absolutely proscribed.

Plain roentgenograms do not visualize cerebral hydatid cysts because they excite only minimal fibrocystic reaction and are characterized by nearly total absence of ectocystic calcification.

Prophylaxis

Prevention of hydatid disease is based mainly on the following measures:

1. Knowledge by the physician of epidemiological and pathogenetic characteristics and diagnostic criteria of the disease.

2. Periodic Casoni test (see above: Laboratory Tests) and prophylactic antihelminthic treatment of working dogs. According to Robinson there is no known drug to kill mature worms in the intestinal tract of the dog, but a good vermifuge is helpful.

3. Avoidance by farmers of the use of carcasses of infested sheep as food for dogs.

4. Destruction of all infested dogs.

The success of such prophylactic measures has been demonstrated in Iceland where echinococcosis present in one-third to one-half of the population at the beginning of the century is now eradicated completely.

Although Birch and Anast stated the definitive host most commonly is the dog, Magath found reports of only 9 canine cases; Hutchinson of 6. In the United States, contrary to experience in many endemic areas, dogs rarely have been found to harbor Echinococcus granulosus. While Hutchinson and Bryan reported infecting dogs experimentally by feeding them with the larval stage found in the liver of naturally infected swine, Hinman and Baker examined 1,315 dogs from New Orleans and found not a single instance of echinococcal infestation. Quite the opposite is true in Alaska where, in some arctic villages, per cent of the dogs examined were found to harbor Echinococcus granulosus. Because of the progressively increasing traffic to and from arctic regions and the present fashion of importing husky arctic dogs (without subjecting them to the Casoni test) there is danger of echinococcosis becoming more frequent in the North American continent, unless the above-listed preventive measures are instituted firmly.

Treatment

No effective medical treatment in hydatid disease is as yet available. The treatment is neurosurgical whenever cerebral hydatidosis is concerned. Surgical treatment is successful mostly in superficially located solitary cysts, including cerebral cysts. The operative removal of large cysts of the brain may cause sudden release of pressure followed by cerebral edema of serious proportions. Therefore, several operative technics have been evolved which favor gradual release of the intracerebral cyst. Most postoperative deaths in hydatidosis of the brain are caused by cerebral edema, hyperpyrexia, cardiorespiratory failure, or anaphylactic shock, the last occasionally forestalled by the use of adrenocorticoids or antihistaminics. Early diagnosis and early operation are the basic factors for favorable outcome.

Summary

1. A clinical case of cerebral hydatidosis has been described constituting the third reported case in which the disease was diagnosed and the patient was operated upon in
this country. To our knowledge, this is the only clinical case of metastatic cerebral hydatidosis reported in the United States.

2. This case shows the unusual feature of 11 years' course from the onset of cerebral symptoms.

3. The pathogenesis of hydatid disease has been reviewed, and a survey of statistics, parasitology, diagnosis, laboratory tests, prophylaxis, and treatment has been included.

4. The importance of sylvatic reservoirs in North America and the incidence of echinococcosis among dogs in arctic regions has been emphasized.

References


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