Artículo original

Hepatic and pulmonary hydatidosis in children. Report of three cases

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Resumen

La hidatidosis es una zoonosis parasitaria transmitida al hombre mediante la ingestión de huevos de *Echinococcus granulosus*, helminto que parasita el intestino delgado del perro. Esta enfermedad afecta principalmente las regiones agrícolas y ganaderas. La enfermedad es poco frecuente en México y la mayor parte de los casos se han diagnosticado en adultos. Los órganos más afectados son hígado y pulmón. Las manifestaciones clínicas dependen de la localización, tamaño del quiste y compresión de estructuras vecinas. El diagnóstico es difícil de establecer debido a que las pruebas serológicas son difíciles de interpretar y la radiología en muchos casos no es concluyente. El tratamiento puede ser quirúrgico, mediante punción-aspiración-inyección-reaspiración o por quimioterapia con albendazol o prazicuantel. Se presentan tres casos pediátricos en los que el diagnóstico fue problemático, y el éxito terapéutico con albendazol.

Palabras clave: Hidatidosis, *Echinococcus granulosus*, albendazol.

Abstract

Hydatidosis is a parasitic zoonosis of humans caused by the ingestión of *Echynococcus granulosus* eggs, a helmint parasite of dogs' intestine. The disease is prevalent in agricultural and cattle-raising areas. It is unusual in Mexico, where most of the cases reported have been in adults. The most frequently involved organs are the liver and the lung. Clinical manifestations depend on the presence of cyst and the pressure it exerts on surrounding structures. Diagnosis is difficult to establish owing to the fact that serologic tests are not easy to interpret and radiologic studies are often non-conclusive. Treatment includes surgery, punction-aspiration-injection-reaspiration of the cyst or chemotherapy with albendazole or praziquantel. We present three children in whom the diagnosis posed a problem; but that were successfully treated with albendazole.

Key words: Hydatidosis, *Echynococcus granulosus*, albendazole.

Introduction

Hydatidosis is a parasitic zoonosis caused by the larval form of *Echynococcus granulosus*, a cestode whose adult stage is a parasite of the dogs' intestine. In its larval stage it affects man and herbivorous animals [1,2].

Whereas the disease is cosmopolitan, it is prevalent in cattle-raising countries. Endemic areas include Asia, Africa, Latinamerica, Australia, New Zealand and the Mediterranean countries. In Latinamerica it has been reported in Argentina, Chile, Uruguay, Peru, Brazil, Colombia and Paraguay. Incidence of the disease in endemic areas ranges from 1 to 220 cases per 100,000 inhabitants. In Southamerica, over 2,000 new cases are reported annually [2-6].

In Mexico only ten cases in human adults have been reported; there are no reports of hydatidosis in children. However, a serologic study to ascertain the prevalence of antiechynococcus antibodies in susceptible populations yielded a 15% positive rate. This figure is higher than expected given the fact that Mexico has large cattle-raising areas and numerous stray dogs.

Humans acquire hydatidosis by the ingestion of *E. granulosus* eggs present in fecal matter of infested dogs.

Hydatidosis symptoms depend on the location of the cysts. Symptoms are not characteristic; they are caused by the pressure exerted by the cyst on the surrounding organs.

When the cyst is located in the lung it may cause chronic cough, hemoptysis, pneumothorax, pleurisy, abscess and other pulmonary symptoms. The cyst may rupture and empty

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its contents in the surrounding space; this causes fever, hypersensibility syndrome expressed by urticaria, pruritus, asthma, hypotension, shock and death. The liver is the most commonly involved organ, followed by the lung [9-14].

Reports of this problem in our country are sparse in adult populations and exceptional in children. The purpose of this paper is to report three children with hydatidosis studied at the INP.

Case 1. A six-year-eight months girl from the state of Guanajuato (Mexico) was the third child of a 30 year old mother. She was born spontaneously at home. There was no prenatal control. Height, weight and APGAR not available. She received all her vaccinations. The family home lacked minimal hygienic conditions; outdoors open fecalism. Poor nourishment. Different animals roamed freely in the house: chickens, dogs, sheep, goats.

Past history includes a blood transfusion at age 6 because of a low blood count. Her problem began one year before with gradual weight loss for a total of six kilograms in one year; she complained of oppressive intermittent abdominal pain in the upper right quadrant radiating to the entire abdomen of ten months duration at different hours. It subsided spontaneously off and on without the use of medications. Four months prior to admission abdominal pain increased and became more frequent. For the last ten months the patient had become pale, asthenic, prostrate and anorexic. She complained of generalized headache at different hours. Subsequently dyspnea appeared which progressed and presented with minimal exertion. Five months prior to admission she had two episodes of hematoquezia.

An hepatic ultrasound was done in her town which showed a hepatic cystic image containing detritus. She was sent to the INP where she was hospitalized on March 2, 1994. Physical examination: The patient was unclean; responsive, there was generalized paleness and wasting i.e., considerable weight loss. Her scalp was full of lice. Numerous caries. Normal chest sounds. There was a grade II/IV systolic murmur in the midprecordial area. The abdomen was somewhat distended and exhibited a superficial venous network. The spleen was palpable 5 cm below the right costal margin. The liver was palpable 4 cm below the right costal margin. Upper and lower extremities were very thin. There were numerous axillary and inguinal lymph nodes of about 2 cm in diameter. The rest of the examination was non-contributory.

The admitting diagnosis was hepatosplenomegaly under study. The patient was seen at the Department of Infectious Disease, Hematology and Gastroenterology. The following laboratory tests and examination were requested to discard the presence of a hydatidic cyst.

Complete blood count on March 3, 1994 reported a hemoglobin of 7.4 g/dL; white blood count, 8,300; segmented neutrophils 66%; lymphocytes, 12%; eosinophils, 16%; (total count, 1328); platelets 303,000; prothrombin time and TPT 71.4% and 71 seconds respectively. A control CBC a week later showed a hemoglobin of 6.6 g/dL which required a blood transfusion. Immunologic tests for VHA, VHB and Toxoplasma were negative. Indirect hemagglutination exam for hydatidic cyst was positive on March 16, 1994 with dilution titers of 1:256. An abdominal US on March 3 showed liver and spleen enlargement and the presence of a round area in the left lobe of the liver measuring 6.0 by 5.0 cm (Figure 1).

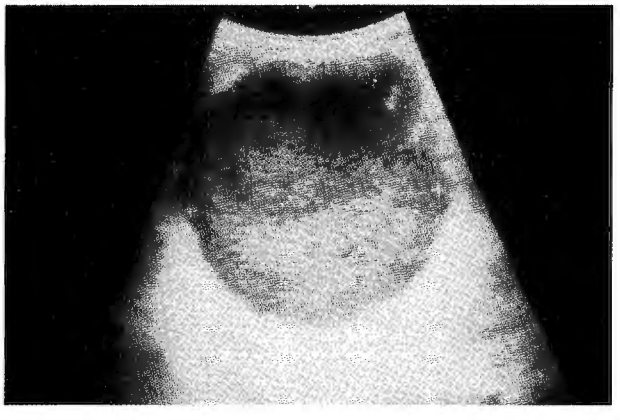

Figure 1. Hepatic ultrasound. A hypoecoic image of a hydatid cyst measuring 6 x 5 cm in diameter.

Treatment was initiated with 15 mg/kg/day of albendazole for 28 days with resting periods of 14 days, for a total of three courses. One month after treatment the patient was stable, complaining of mild abdominal discomfort. An endoscopy was performed on June 8, 1994 which revealed the presence of grade I esophageal varices.

A control hepatic US showed a predominantly liquid cystic image in the left lobe measuring 6.4 x 5.9 cm. A splenoportogram was done on June 21, 1994 which showed a cavernomatous tortuous portal degeneration with a pressure of 36 mmHg. An abdominal CAT scan reported the presence of a cyst in the left hepatic lobe measuring 6.3 x 5.7 cm; there was portal hypertension related to the cavernomatous degeneration and the hepatosplenomegaly.

It was decided that the antiparasitic treatment was not successful. On June 27 an exploratory laparotomy was done; 60% of the cyst was removed. Microscopic examination of the liquid content was negative for the presence hydatidic sediment.

The patient made a successful recovery and was discharged on August 1, 1994. Her only complaint was occasional abdominal pain. The liver and the spleen recovered their normal size. Hepatic function tests were normal.

Five months later (Jan. 3, 1995) portal hypertension was still present as well as a superficial collateral venous network; the liver was palpable 3 cm below the right costal margin and the spleen, 2 cm below the left costal margin. In May 1995 an esophageal endoscopy revealed grade II varices. A control indirect hemagglutination test was negative for hydatidic cyst. A control abdominal CAT scan indicated the disappearance of a cystic image with persistence of hepatomegaly and splenomegaly. A second exploratory laparotomy was performed on July 5, 1995; a liver biopsy was taken; the histopathologic report was focal fibrosis of the capsule. Propranolol was prescribed at the dose of 2 mg/kg/day. On Jan 3, 1996 a control endoscopy showed grade I-II varices in the upper third and grade III-IV in the middle and lower thirds of the esophagus; esophageal varices in the gastric fundus and congestive gastritis were present. On March 1998 another endoscopy showed grade I esophageal varices and congestive uncomplicated gastritis. One more endoscopic study on Jan. 17, 2002 was reported as normal. On September 17, 2002, the patient was asymptomatic and her laboratory and specialized studies were normal.

Case 2. An eleven year old girl from the State of Mexico, born to a 34 year old mother in her eighth pregnancy. There was no prenatal control. A cesarean section was done following a 48 h history of membrane rupture. The child breathed and cried spontaneously at birth. APGAR, weight and height not available. The child received all her vaccines. At age 7 she developed generalized tonic clonic convulsive seizures for which she was not treated. They subsided spontaneously at the age of nine years. The patient lived in a rural home with no hygienic conditions. She had poor nourishement. Dogs and cats lived with the family.

Four years prior to admission to our Institution the patient complained of intermittent moderate stabbing abdominal pain in the right upper quadrant which subsided spontaneously. Three days prior to admission she coughed up a large amount of mucous salty-tasting matter on two occasions during a severe bout of right upper abdominal quadrant and urticaria. She was then hospitalized in the capital city of her state.

She was admitted to our Institution on August 1, 1996. A chest X ray showed a radiopaque image with well defined borders in the base of the right lung (Figure 2). A US showed a cystic image in the left hepatic lobe which suggested and amoebic liver abscess for which she was given metronidazole. She was discharged a week later. A control US showed the same cystic image. A CAT scan done on August 23, 1996 showed a right posterior basal lung lesion measuring 6.5 x 4.5 x 5 cm and a hepatic cystic lesion in the left lobe measuring about 6 cm in diameter (Figure 3).

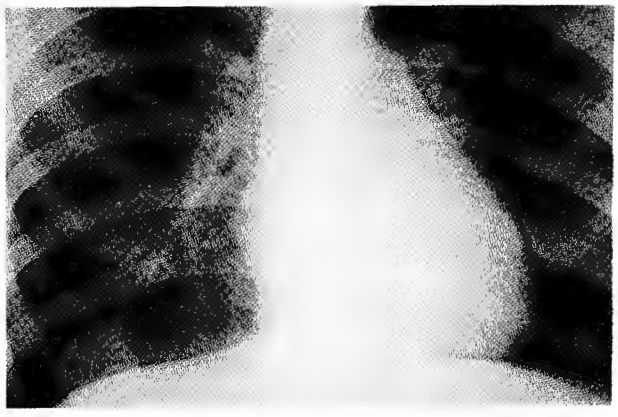

Figure 2. Chest X ray showing a round radiopaque with a well-defined borders image in the right basal region.

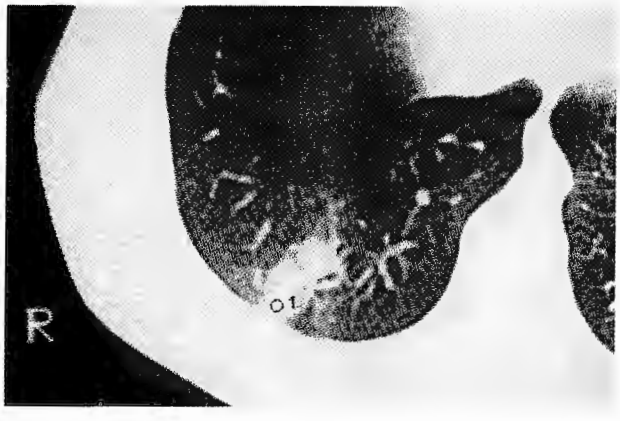

Figure 3. Chest CAT scan . There is a right lung posterior basal lesion measuring 6.5 x 4.5 x 5 cm in diameter.

At the end of the treatment she developed a fever of 38°C and productive cough. She weighed 29,800 kg; height, 1.44 m. Heart rate 80/min; temperature 37°C. The patient was

alert. Her throat showed hyperemia and whitish spots. There was right basal chest hypoventilation. The abdomen was normal.

Tests were done to confirm a probable hydatidosis. Hemoglobin, 12.8 g/dL; white blood cells, 6,800; neutrophils 42.2%; lymphocytes 49.3%; eosinophils under 0.7%; platelets 268,000. Liver function tests were normal. ELISA was negative for amoeba. On September 19, HAI was positive for *Echinococcus granulosus* with a 1:2084 dilution, which confirmed the diagnosis of hydatidosis. A lung US on September 18, 1996, showed a cystic image with poorly defined contours at the right pulmonary base of 3 x 4 x 2.8 cm and a cystic image in liver segments 6 and 7 with a thick wall and a well defined contour. Treatment was then initiated with 10 mg/kg/day of albendazole for 30 days.

She was discharged clinically asymptomatic on September 23, 1996 and was advised to return for consultation in the Departments of Gastroenterology, Pneumology, Thoracic Surgery and Parasitology.

On September 22, 1996 she remained asymptomatic with the exception of mild transient headaches. A second course of albendazole was well tolerated by the patient. Radiological findings were normal. On January 27, 1997, the HAI gave a 1:228 dilution result following a third course of albendazole. The poor response to this medication necessitated the contribution of antiparasitic treatment. On March 02, 1998 she was asymptomatic and completed her eighth course of albendazole. A liver US showed a reduced lesion measuring 2 cm in diameter. A pulmonary TAC exhibited a 3 cm hollow fibrotic lesion (Figure 4).

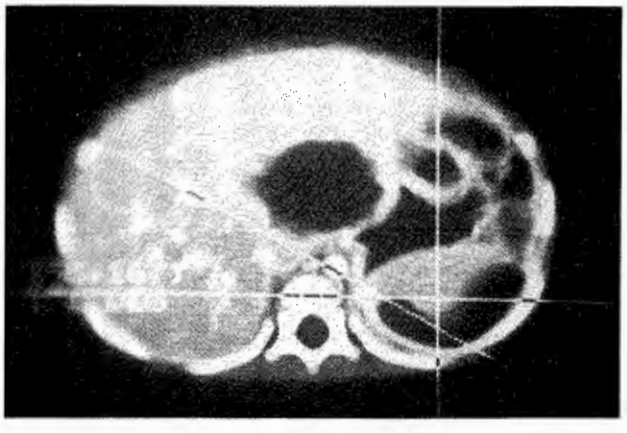

Figure 4. Abdominal CAT scan with an hepatic cyst measuring 4.6 x 6.5 x 6.0 cm in the right hepatic lobe.

The patient remained asymptomatic by June 23, 1999. The control HAI gave a 1:320 dilution titer. Pulmonary and hepatic X rays showed small cystic lesions.

May 21, 2002. The patient continued to be asymptomatic and in good condition. A lung X ray showed a fibrotic cystic lesion; the liver X ray showed a healed cystic lesion. The patient remains under observation.

Case 3. A two-year-three-month old boy from the state of Guerrero, Mexico was born of the second pregnancy of a 24 year old mother. Following an episode of threatened abortion the child was prematurely born on the 7th month pregnancy, weighing 3,400 grams with a height of 52 cm. APGAR was not determined. The child received every vaccination. The family lived in a rural unhygienic home with one dog and several chickens. Poor eating habits.

Past medical history was non-contributory. Two weeks prior to admission he had a temperature of 38.5° C. He was diagnosed as having an upper respiratory infection for which he was given a febrifuge; he had loss of appetite, nausea, weight loss and a distended abdomen. Hepatosplenomegally was detected. The diagnosis of hepatic amoebic abscess was made based on a US study. He was treated with metronidazole and ceftriaxone for 7 days. He developed disseminated petechiae at the site of needle puncture. His condition did not improve. Two weeks later a CAT scan showed a cystic area in the left hepatic lobe. A conglomerate of lymph nodes was seen in the spleen and the peritoneum; they formed a mass which displaced the large intestine.

He was hospitalized at the Instituto Nacional de Pediatría on October 4, 2001. Physical exam. The patient was conscious, afebrile, with pale skin and mucosae. Large abdomen; the liver was felt 7, 8 and 8 cm below the right costal margin; it was tender to palpation. The spleen was palpable 5 cm below the left costal margin. Peristaltic movements were present. The rest of the examination was non-contributory.

Heart rate 100/min; respiratory rate 20/min. Blood pressure 100/60. Temperature 37 ° C; weight 13 kg; height, 86 cm.

The admission diagnosis was hepatosplenomegally under study. He was evaluated by the Departments of Oncology, Surgery and Parasitology. Several tests were done to determine the diagnosis of hydatidosis vs amoebic abscess.

On October 4, 2001 hemoglobin was 9 g/dL; leukocytes 20,000; segmented neutrophiles 58%; lymphocytes 36%; eosinophils under 0.7%; platelets 180,000.

On October 4 an abdominal CAT scan showed an area of diminished density in the left hepatic lobe measuring 4.6 x 6.5 x 6.0 cm (Figure 4).

A CBC on October 8 reported 9.5 g/dL hemoglobin; 11,400 leukocytes; segmented neutrophils 44%; lymphocytes 42%; eosinophils under 0.7%; platelets 465,000. Three serial coproparasitoscopic studies were negative. Blood chemistry and liver function tests were normal.

A Casoni intradermal reaction on October 19 was negative at 48 h. ELISA assays for *Toxocara* and for amoeba were negative. HAI for hydatidic cyst was also negative. Because of these negative results, new tests were done in order to discard the diagnosis of hydatidosis. Counter immunoelectrophoresis showed a positive precipitation band. These findings and the absence of daughter hydatidic vesicles and of sediment within the cystic mass supported the diagnosis of a sterile hydatidic cyst. The patient was treated with albendazole at the dose of 10 mg/kg/day for 28 days in three courses with resting periods of 14 days between each course.

He was asymptomatic, discharged on October 30 and adviced to return for consultarion in the Outpatient Departament. The patient remained asymptomatic on December 12, 2001. A CAT scan done on December 10 reported a decrease in the size of the cyst. A counter immunoelectrophoresis control test for hydatidic cyst was negative. Serologic controls in February of 2002 for hydatidic cyst were negative. Three courses of albendazole were completed with no side effects.

On July 2, 2002 the patient remained asymptomatic. A serologic test for hydatidic cyst was negative; radiologic studies did not show a cystic lesion.

Discussion

Hydatidosis is a polymorphic disease not easy to diagnose, especially if an epidemiologic assessment has not been done properly and when the diagnosis is not suspected. Indeed, symptoms may be easily mistaken for bacterial infectious, parasitic or neoplastic conditions as was the case in the patients described above.

An epidemiologic assessment is of the utmost importance to suspect the diagnosis. Living in close contact with dogs carrying the parasite is the main source of infection. This was the case in the three patients under discussion.

Notwithstanding the low incidence of pediatric hydatidosis in non-endemic areas this disease must be discarded in the differential diagnosis in the presence of an abdominal mass with cystic features in the liver and/or spleen detected by ultrasound or CAT scan studies.

The same is true for cystic masses in the chest, especially when in addition to the mass, there is a hypersensitivity reaction for parasitic antigens, such as urticaria, bronchial spasm, sibilant rales and expectoration of egg-white consistency salty matter, which is in fact a hydatidic vomica; this occurred in one of our patients and it provided the clue for the diagnosis.

The most common symptoms in our patients were fever, abdominal pain, hepatosplenomegaly, cough, asthenia, adynamia and loss of appetite.

Laboratory tests are difficult to interpret; they require a correlation with the clinical history and with other specialized studies. This was apparent in the tests done in our three patients. Casoni's intradermal reaction test was only done in one patient and it was negative. The reason for this is that the sensitivity of the test varies from 40 to 90% depending on the purity of the antigen [15].

Sensitivity of serum antibodies against the parasite with the HIA and ELISA tests is 80%. However, especially in chronic cases when the host has developed a fibrous layer around the cyst there is no contact with the parasite for which reason, the measurement of antibodies may be negative as in one of our patients, who had a negative hemagglutination test and required more sensitive tests such as counter immunoelectrophoresis which was positive [15,16].

Eosinophilia was present only in one of our patients, as has been reported in the literature: only 15 to 25% of the cases have it.

Hepatic enzymes were slightly elevated in our patients, especially alcaline phosphatase which did not reach abnormal levels during treatment.

Treatment with albendazole must be assessed based on the radiologic features and not on the level of antibodies, since these have great variations depending on the sensitivity of the patient, his immunologic condition and his age. Thus in patients under 15 year of age, the immunologic response is lower than in adults. Most of the reports assess the response to treatment on the basis of the reduction in size of the cyst and its changes, such as fibrosis and scarring. Our three patients were considered cured on the basis of radiological findings: disappearance of abnormal images or the presence of a scar and/or residual fibrosis.

Chemotherapy with albendazole or praziquantel is indicated for inoperable patients with cysts in two or more organs; for cases with rupture of the cyst into the biliary ducts or the bronchi; for peritoneal hydatidosis and as an adjuvant of surgical treatment [17].

Surgical treatment has a 2% mortality rate and recurrence rates of 2 to 25%. It is indicated in large hepatic cysts with multiple secondary cysts; in superficial hepatic cysts at risk of rupture by trauma or sponteaneously; in hepatic cysts draining into the biliary ducts, or pressing vital organs; in infected cysts [1,5].

Puncture-aspiration-injection-reaspiration is an alternative for inoperable patients; for patients who refuse operation; for multiple cysts in hepatic segments I, II and III; for recurrences following surgical treatment or chemotherapy [19].

Hydatidosis can be prevented with adequate control and disposal of dog feces; with periodic antiparasitic treatment of these animals who should not be fed raw viscerae. These measures will significantly diminish the risk of children to acquire the disease.

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