

## RENAL ECHINOCOCCUS

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Echinococcosis, or cystic hydatid disease, is a parasitic infestation endemic to most parts of Asia, Europe, South America, the Near East, Australia, New Zealand, Canada and Alaska.<sup>1-6</sup> Although liver and lungs are most frequently involved, it can invade almost any organ. Its incidence in the kidney is 2 to 3 per cent.<sup>7-9</sup> Parasitologists have warned that with increased travel, conditions are ripe for its spread to the United States.<sup>10</sup> Already there is some autochthonous Echinococcus in the southeast United States and California.<sup>4,11</sup> The more malignant varieties frequently have been misdiagnosed as cancer even in areas of the world where suspicion of the disease runs high.<sup>1, 5, 6</sup> There is no effective chemotherapy for the disease and public health measures to date have been unable to control it.<sup>12</sup> The only treatment is surgical.

### CASE REPORT

A 28-year-old man from Michigan had severe left flank pain for the first time 2 weeks prior to

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<sup>1</sup>Smith, E. M. G. and Hanson, S.: Alveolar echinococcosis: a case report with a discussion of the ecology of the disease. *Amer. J. Clin. Path.*, **35**: 160, 1961.

<sup>2</sup>Bakir, F.: Serious complications of hydatid cyst of the lung. *Amer. Rev. Resp. Dis.*, **96**: 483, 1967.

<sup>3</sup>Papadimitriou, J. and Mandrekas, A.: The surgical treatment of hydatid disease of the liver. *Brit. J. Surg.*, **57**: 431, 1970.

<sup>4</sup>Wilson, J. F., Diddams, A. C. and Rausch, R. L.: Cystic hydatid disease in Alaska: a review of 101 autochthonous cases of Echinococcus granulosus infection. *Amer. Rev. Resp. Dis.*, **98**: 1, 1968.

<sup>5</sup>Mathur, S. C., Bhatt, J., Gangwal, K. C. and Sharma, M. L.: Primary retroperitoneal hydatid cyst. *Int. Surg.*, **51**: 54, 1969.

<sup>6</sup>West, J. T., Hillman, F. J. and Rausch, R. L.: Alveolar hydatid disease of the liver: rationale and technique of surgical treatment. *Ann. Surg.*, **157**: 548, 1963.

<sup>7</sup>Deliveliotis, A., Kehayas, P. and Varkarakis, M.: The diagnostic problem of the hydatid disease of the kidney. *J. Urol.*, **99**: 139, 1968.

<sup>8</sup>Henry, J. D., Utz, D. C., Hahn, R. G., Thompson, J. H., Jr. and Stilwell, G. G.: Echinococcal disease of the kidney: report of case. *J. Urol.*, **96**: 431, 1966.

<sup>9</sup>Sharma, S. K. and Eggleston, F. C.: Management of hydatid disease. *Arch. Surg.*, **99**: 59, 1969.

<sup>10</sup>Rausch, R.: Studies on the helminth fauna of Alaska. XXX. The occurrence of Echinococcus multilocularis Leuckart, 1863, on the mainland of Alaska. *Amer. J. Trop. Med. Hyg.*, **5**: 1086, 1956.

<sup>11</sup>Schantz, P. M., Clérout, R. P., Liu, I. K. M. and Schwabe, C. W.: Hydatid disease in the central valley of California. *Amer. J. Trop. Med. Hyg.*, **19**: 823, 1970.

<sup>12</sup>Schwabe, C. W.: Epidemiology of echinococcosis. *Bull. WHO*, **39**: 131, 1968.

admission to the hospital. He had no hematuria, fever, voiding difficulty or bladder irritative symptoms. He had spent 2 years in the army in southern Spain and 6 weeks in North Africa. Physical examination was normal. Urinalysis demonstrated no white cells, red cells, bacteria, protein or sugar and the pH was 6. Serum creatinine was 1.0 mg. and blood urea nitrogen 11 mg. per 100 ml. Hematocrit was 44 per cent and white blood count 6,500 mm.<sup>3</sup> with a normal differential. Alkaline phosphatase was 51 international units, bilirubin 0.5 mg. per cent, serum glutamic oxaloacetic transaminase 38 and serum glutamic pyruvic transaminase 20 international units (all normal values). Urine culture for acid-fast bacillus was negative. Chest x-ray and bone survey were negative. An excretory urogram (IVP) showed a left renal mass (fig. 1). Renal arteriography showed a lucent tumor deformity in the mid portion of the left kidney. Cyst puncture was performed and chocolate-colored fluid was obtained. No acid-fast bacilli, fungi or tumor cells were found in the necrotic fluid.

A transabdominal left nephrectomy was performed. Several cystic areas in the kidney were noted from which dark bloody fluid was removed. There were no other lesions in the abdomen. Convalescence was unremarkable.

Microscopic section of the resected kidney demonstrated echinococcosis. There was 1 multiloculated cyst about 2 cm. in diameter with central necrosis. Microscopically there were numerous brood capsules invading surrounding tissue exogenously with a granulomatous reaction of fibroblasts, plasma cells, giant cells and epithelioid cells (figs. 2 and 3).

### DISCUSSION

The literature on echinococcosis is confusing because too often a distinction is not made between the different forms. There are 3 basic types: 1) sylvatic Echinococcus granulosus is a part of a wild moose, caribou or deer—wolf, dog cycle and usually forms a benign self-limited unilocular cyst,<sup>4, 13</sup> 2) pastoral Echinococcus granulosus is part of a domestic sheep, swine or cattle—dog cycle and forms a rapidly expanding unilocular cyst that carries a 3 to 10 per cent mortality<sup>3, 4</sup> and 3) Echinococcus multilocularis is a less common but invasive multicystic process that behaves like cancer and is almost uniformly fatal.<sup>1, 6, 13, 14</sup> There are also gradations between these forms that are difficult to classify.<sup>1</sup> E. granulosus and E. multilocularis are

<sup>13</sup>West, J. T.: Malignant echinococcosis disease of the liver. *Alaska Med.*, **1**: 107, 1959.

<sup>14</sup>James, E. and Boyd, W.: Echinococcus alveolaris (with the report of a case). *Pathology*, **35**: 160, 1961.



FIG. 1. IVP reveals cystic tumor deformity in lower pole of left kidney

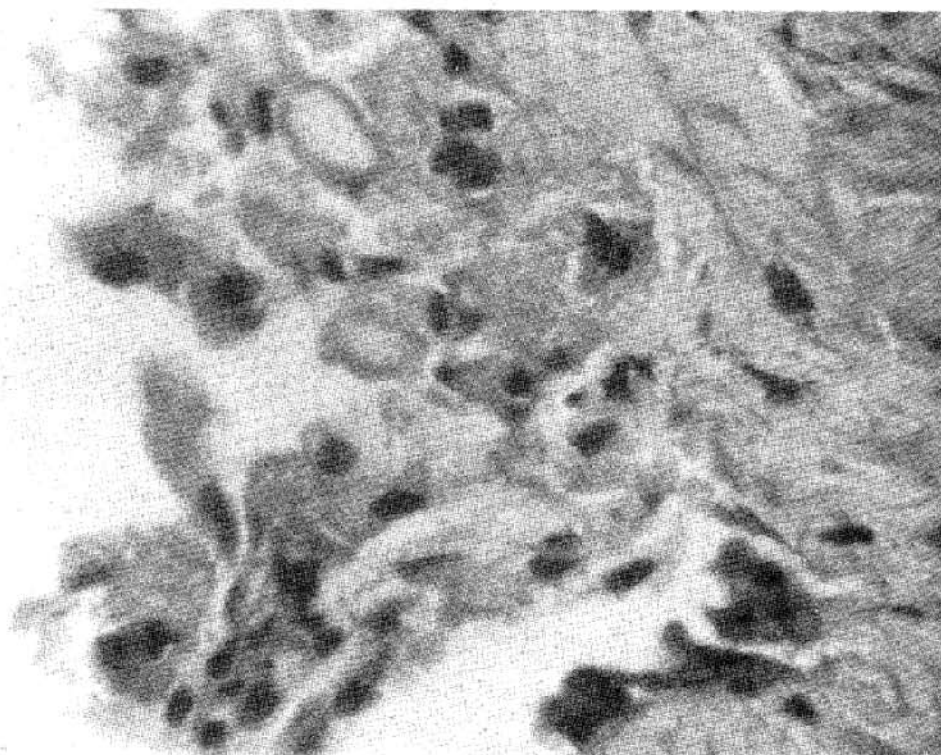


FIG. 2. Brood capsules seen to invade irregular wall of cyst. Reduced from  $\times 215$

indigenous to central and southern Europe, where our patient had visited.<sup>10</sup>

*E. granulosus* forms a single cyst typically surrounded by a dense layer of connective tissue with endogenous budding. The cyst wall consists of 2 layers: 1) a thick, white outer layer and 2) a thin, granular inner layer which buds off brood capsules

and scolices internally. These may form daughter cysts within the major walled-off cyst. The cyst fluid is clear. The indigenous *E. granulosus* in Canada and Alaska is sylvatic and quite benign.<sup>4</sup> However, the cases of *E. granulosus* that are imported from other areas of the world and those indigenous to California and the southeast United States are most

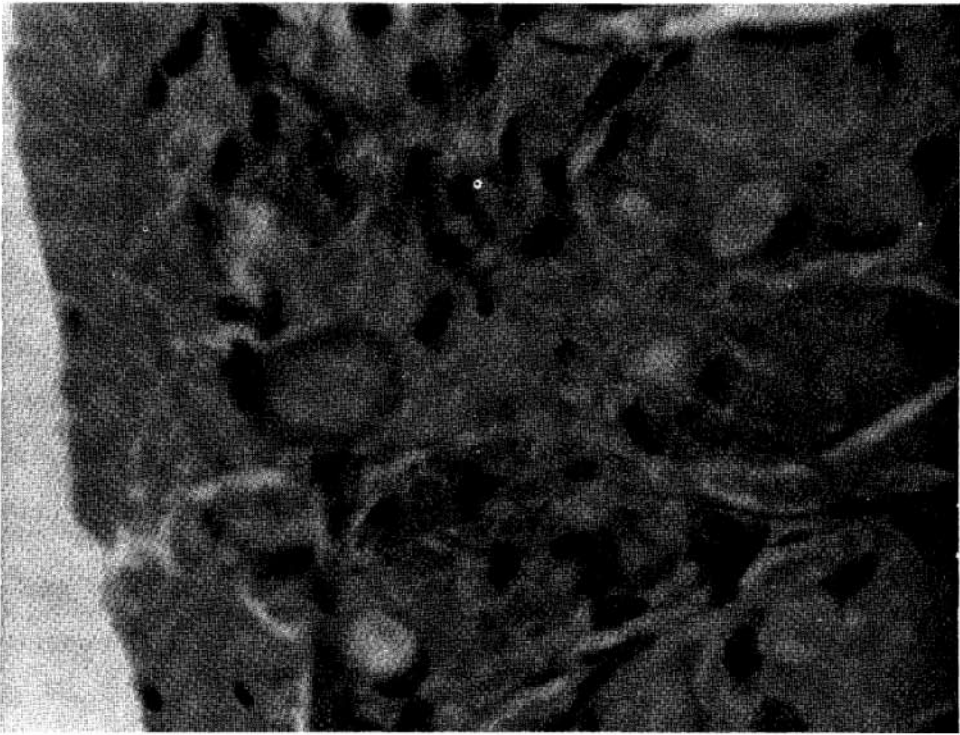


FIG. 3. Brood capsules actively invading surrounding renal tissue and producing intense granulomatous reaction. Reduced from  $\times 215$ .



FIG. 4. Typical spherical calcification seen in *E. granulosus*.



FIG. 5. Typical feathery calcification seen in *E. multilocularis*.

often pastoral. This form can be aggressive and cause surrounding pressure necrosis with rupture

into open body spaces such as the peritoneal or pleural cavity, bronchi and so forth, producing local symptoms or sometimes anaphylactic-type reactions. Simple surgical excision, drainage or formalization is usually adequate treatment.

*Echinococcus multilocularis*, on the other hand, behaves like a malignant neoplasm and grows by exogenous budding.<sup>15, 16</sup> Our experience with 19 cases in northwest Alaska led us to conclude that an early, aggressive surgical approach is warranted.<sup>17</sup> Grossly, it looks much like neoplasm and even on frozen section has been mistaken for undifferentiated carcinoma.<sup>1</sup> The larvae actively invade surrounding tissue and blood vessels and can metastasize to any part of the body. There is no clear plane for tissue dissection and a radical extirpative operation is the only hope for cure.<sup>1, 6</sup>

Mathur reported on a case of *Echinococcus* (probably *multilocularis* but not documented as such), which presented clinically as a massive right hypernephroma. At operation the diagnosis was confirmed, the "tumor" was biopsied and it was only after examination of microscopic sections that the correct diagnosis was made of *Echinococcus*.<sup>5</sup> The lesion was unresectable.

Henry reported a typical case of *E. granulosus* of the right kidney, 6.5 cm. and circular, calcified around the periphery. There were daughter cysts within the main unilocular cyst formed by breaking off of scolices from the inner germinal layer by trauma but there was no invasion of surrounding tissue.<sup>8</sup> A segmental nephrectomy was performed.

Deliveliotis reported on 12 cases involving the kidney and pointed out that it is often difficult to differentiate these lesions from malignant tumors or tuberculosis.<sup>7</sup>

There are some helpful diagnostic points. Occasionally echinococcosis of the kidney can be heralded by hydatiduria or the passage of membranes or cyst material in the urine.<sup>7</sup> Less than half the patients have eosinophilia.<sup>7, 8</sup> The Casoni skin test and serum hemagglutination titer are positive about 90 per cent of the time except in the benign sylvatic variety, in which they are almost always negative.<sup>4, 7, 8, 12, 17</sup> More than 50 per cent have typical calcifications seen on x-ray. In *E. granulosus*, there is an unmistakably round, homogeneous shadow (fig. 4). In *E. multilocularis*, however, calcifications are feathery or stippled (fig. 5).<sup>7, 14, 17</sup>

The case presented herein probably represents *E. multilocularis* or pseudo-multilocularis.<sup>1</sup> Liquefaction in the necrotic portion of the larval mass may make the lesion superficially resemble a unilocular cyst of *E. granulosus* but exogenous budding, the invasive nature of the parasitic tissue at the periphery of the lesion, the irregular pattern and the intense surrounding granulomatous response

distinguish it from the more benign form of the disease.<sup>14</sup> There was no eosinophilia or calcification. Urinalysis was normal and there was no hydatiduria. Neither skin tests nor hemagglutination titers were performed because of lack of suspicion of the disease.

To appreciate how *Echinococcus* can become more prevalent in the United States, one must understand its life cycle. The different types of echinococcosis are caused by closely related parasites that spend their adult life in the intestine of a definitive host, which can be a dog, fox, wolf, coyote or even cat, depending on the locale. These definitive hosts are in no way harmed by the parasite.<sup>16</sup> The adult parasite sheds eggs into the stool of these hosts.

These eggs are then ingested by an intermediate host usually by eating contaminated vegetation. The intermediate host is usually a grazing animal such as deer, moose and caribou in the sylvatic *E. granulosus* life cycle or cows and sheep in the pastoral *E. granulosus* cycle. In *E. multilocularis*, the major intermediate hosts are microtine rodents, such as the field vole or lemming.<sup>16</sup>

From the intestine of the intermediate host, the larval forms find their way through the portal system to the liver as a first filter, the lungs as a second filter and from there to any other site in the body. About 15 to 20 per cent of the scolices thus enter the general circulation and are distributed according to blood flow and density of local capillary beds.<sup>9</sup> It is this larval form that causes the disease that may ultimately kill its intermediate host. At that point the life cycle would end except that a new definitive host may then ingest the larvae in the infected viscera of the intermediate host. The larvae mature there into adults and the cycle begins again. Man in this case is just another intermediate host.

Since among the intermediate hosts or victims, man is the only carnivore, it should be noted that eating the infected liver or any other infected organ of an intermediate host will not produce the disease. Only the intact eggs shed in the stool of the definitive host such as a dog, can pass the disease on to man.

Since rats could easily substitute for field voles, the condition is obviously favorable for establishment of *E. multilocularis* in urban as well as rural areas if pet dogs or cats are brought from endemic regions.<sup>10</sup> It would be difficult to keep these pets from passing the parasite to local rodents which would then be eaten by other local dogs or cats. *E. granulosus* could also be easily established if dogs are allowed to eat the viscera of dead sheep or game animals, a custom deeply ingrained in many rural areas.

#### SUMMARY

A case of *E. multilocularis* of the kidney is presented. More *Echinococcus* may be seen in the United States in the future. The life cycle, pathology, diagnosis and management of echinococcal disease are discussed.

<sup>15</sup> Rausch, R. and Jentoft, V. L.: Studies on the helminth fauna of Alaska. XXXI. Observations on the propagation of the larval *Echinococcus multilocularis* Leuckart, 1863, in vitro. *J. Parasit.*, **43**: 1, 1957.

<sup>16</sup> Rausch, R. and Schiller, E. L.: Studies on the helminth fauna of Alaska. XXIV. *Echinococcus sibiricensis* N. Sp., from St. Lawrence Island. *J. Parasit.*, **40**: 659, 1954.

<sup>17</sup> Silber, S. J. and Wilson, J.: Surgical treatment of *Echinococcus multilocularis*. To be published.