Hydatid Cyst of the Brain
Report of a Case

Leslie E. Geiger, M.D.
Departments of Neurosurgery, University of Southern California and Los Angeles County General Hospital, Los Angeles, California

Hydatid infestation of the brain is an unusual manifestation of a disease which in any anatomical location is rare. Katz and Pan,9 in a collective review of 541 instances of echinococcosis diagnosed in the United States between 1892 and 1956, report that only 5 new cases were found annually. The majority of these individuals acquired the disease outside the borders of the continental United States. Incidence of the cerebral form in this series was 1 per cent contrasted with 70 per cent hepatic and 10 per cent pulmonary infections. A comparable figure of 2 per cent cerebral infections is quoted by Araña-Iniguez and San Julián1 from Uruguay, where the frequency of echinococcosis is considerably higher. Statistics regarding association of cerebral cysts with infection in other organs vary widely.2 As can be surmised, familiarity with the treatment of the intracranial form of hydatid disease has been most extensive in countries where sheep raising is common, yet, even in Roumania, intracranial hydatidosis represents only 1.6 per cent of 2,286 intracranial mass lesions.2

Consistent with the rarity of intracranial hydatid cyst formation, the number of reports of operated cases in North America is small. Only 4 have been published. The first by Mudd5 in 1892 is of considerable historical interest representing, possibly, the first successful removal of an intracranial hydatid cyst. Another patient was reported by Craig and Kernohan6 in 1934 and a third instance was recently published by Ayres, et al.7 A brief abstract concerning a patient found at operation to have a multilocular cerebral cyst was included in a study of the ecology of hydatidosis in the Northwest by Rausch and Schiller.16

Another instance of cerebral hydatid cyst is presented here. Preoperatively, it was erroneously diagnosed as an avascular neoplasm on the basis of carotid angiography. A greater awareness of the possibility of echinococcosis might have led to a better planned operative approach with consequent avoidance of rupture of the cyst and spilling of its contents. The cyst was, however, successfully resected and the patient has returned to her usual activities.

Received for publication November 16, 1964.

Case Report

Mrs. D.A. (Los Angeles County General Hospital P0223-34-42) was born in Mexico in 1910. Except for a month-long visit to her birthplace in 1957, she had lived in Los Angeles since 1922.

In March 1961 her right arm suddenly became weak and speech was noted to be slurred. This disturbance cleared within 3 days. The patient made a brief stay at a psychiatric hospital where a diagnosis of involutional depression was recorded. Similar episodes recurred twice during the next 18 months. Beginning in August, 1962, the right upper and lower extremities became progressively weaker. The paresis was accompanied by right parieto-occipital headaches. Her right arm was burned in September, 1962, but no pain was experienced. Repeated questioning of the patient and her family failed to reveal any history of related prior illness. She had been pregnant 3 times and delivered without complications in 1930, 1936 and 1946.

Examination. When seen at the Los Angeles General Hospital on October 24, 1962, she seemed a well-developed and well-nourished woman with a blood pressure of 130/80, and a pulse of 80. The left parieto-occipital region of the skull was tender but no exostoses were palpable nor were any bruits audible.

The patient was oriented as to person and place but not as to time. Although bilingual, she had considerable trouble with word selection even in Spanish. There was equivocal blurring of the temporal margins of both optic discs. Right homonymous hemianopia was demonstrated by confrontation testing. There was hypesthesia in the right trigeminal distribution. The right lower facial muscles were weak. Paresis was also demonstrated in the right arm and to a lesser extent in the right leg. There was apraxia in the right hand. Sensory examination revealed right hemihypesthesia and hypalgesia as well as extinction of simultaneously applied tactile stimuli on the right side. Proprioceptive and vibratory sensibilities were diminished in the right leg. There were hyperactive tendon reflexes on the right side, normal abdominal reflexes and upgoing plantar responses bilaterally.

Laboratory examination. The hemoglobin was 14 gm. and urinalysis was normal. The lumbar cerebrospinal fluid pressure was 200 mm. of water. The fluid was clear, colorless and without cells; protein content was 53 mg. per cent, sugar 61 mg. per cent and chloride 121 mg. per cent. Roentgenogram of the chest was normal. Bilateral carotid arteriography revealed an avascular left frontal opercular and parietal lesion with a square shift of the anterior cerebral and pericallosal arteries to the right as well as a shift of the internal cerebral vein (Fig. 1a, b).

Operation. On October 27, 1962, a left parietotemporal
Hydatid Cyst of the Brain

Fig. 1. A. Left carotid arteriogram antero-posterior projection with shift of pericallosal vessels to the right under the falx and outline of a superficial avascular lesion by posterior middle cerebral branches. B. Lateral projection of carotid arteriogram with outline of avascular mass by branches of the middle cerebral artery.

craniotomy was performed. The bone appeared irregular and thin; atrophic dura was adherent to the bone in the mid-parietotemporal region. Immediately subdurally, a cyst was observed which ruptured at the time of reflection of the dural flap. Approximately 50 to 60 ml. of clear fluid escaped. This thin-walled cyst occupied the mid-portion of the left side of the calvarium and extended into the Sylvian fissure spreading it to expose the posterior insula. In the midst of the cyst, a 1/2 cm. yellow nubbin of firm tissue was found. The cyst wall was totally removed with the exception of a few adherent strands about the Sylvian vessels. The subdural space was drained with a #14 catheter. The dura was incompletely closed due to its extreme thinness and friability. An epidural drain was also placed because of persistent oozing of the dural surface in the vicinity of the lesion.

Postoperative Course. The patient awoke immediately postoperatively but a deepening comatose state and the appearance of a speech disturbance 3 days later led to reopening the craniotomy; a 2 cm. thick epidural hematoma was found and evacuated. Following the 2nd operation the patient’s postoperative recovery was gratifying with progressive clearing of the weakness in the right hand and the speech disturbance. Sensory findings and hemipanopia soon disappeared and she returned to work as a seamstress. At her most recent examination in April, 1964, 18 months after surgery, the only residual was minimal weakness of the right hand. Focal motor seizures involving the right thumb had occurred 3 times. Two of the seizures were preceded by an ill-defined visual aura. Lumbar puncture performed 16 months following operation revealed fluid under pressure of 75 mm. of water, protein 69 mg. per cent and normal sugar and chloride contents. No cells were seen in the fluid.

Histological examination of the cyst wall revealed a layer of fibrous tissue investing a laminated, acellular membrane to which were attached germinal cells. Sclerions were numerous within the inner membrane and fluid (Fig. 2).

Discussion

Coincident with the increasing population of Western Canada and Alaska and the improvement of medical service to the native population, echinococcosis may become a more frequently encountered disease entity in North America. 16 14 In one region of the Northwestern territory, 28 per cent of dogs were found to carry the tapeworm, E. granulosus. 12 In addition to the dog, definitive hosts of the adult tapeworm may be other carnivores such as wolves, foxes and coyotes. Sweatman 19 reports that 36 of 58 wolf carcasses were found infected. The herbivorous wildlife such as the deer, moose, caribou and rodents in addition to domestic hogs and cattle may serve as intermediate hosts. 15 Cross infections may occur within these groups and the disease is widely spread by migratory species of wildlife. In other climates the usual intermediate host is the sheep. Certain of the wildlife may be specifically infected by E. multilocularis or sibiricensis which cause alveolar hydatidosis, a variant of the disease which may also lead to cerebral infection. 12

Man enters the cycle when food contaminated by the excreta of infected carnivores, usually canines, is ingested. The high frequency of this disease in children in localities where sheep raising is common is probably explained by intimate contact with pet sheep dogs. The ova which are excreted in huge numbers by the infected dog hatch in the duodenum of the intermediate host and are then spread hematogenously. The first barrier, and the most common site of infection, is the liver. The lung forms a second possible sieve. Primary cerebral infection is caused by hexaneanth embryos which have bypassed both
these filters. Secondary cerebral cysts arise from the rupture of a fertile cyst in the lung with subsequent spilling of scolices into the pulmonary vein or from the rupture of a cyst in the left side of the heart. Because of the embolic nature of the infestation, the favored site of cyst formation is the region of the brain supplied by the middle cerebral artery.

Growth of the cerebral cyst tends to be extremely slow; 3 to 4 years or more pass before the mass becomes symptomatic. The cysts tend to be single, though multiple cyst formation is presumed from the rupture of fertile cysts within the arterial side of the circulation. Multiple cyst formation may also follow a head injury which ruptures a viable primary cyst. The reaction of the surrounding brain is usually negligible and the only contribution of the host to the cyst wall is a thin gliotic membrane. When the cyst impinges on the surface of the brain, the arachnoid and dura may become locally adherent, as apparently occurred in this case.

Clinically, the cyst acts as any other slowly growing mass with ultimate development of focal signs and evidence of increased intracranial pressure. Tarcan mentions that the overlying skull may be tender, a preoperative finding in the present report.

Laboratory tests for echinococcal disease are of little value in establishing the diagnosis of intracranial infection. In systemic infection, eosinophilia of approximately 5 per cent of the total leukocyte count is found in about 50 per cent of cases. The Casoni intradermal allergic reaction utilizing cyst fluid is stated by some to be positive in 75–95 per cent of infected individuals but is often falsely positive. The complement fixation reaction is positive in 50–60 per cent, and other serological tests may prove to be even more accurate. Cerebrospinal fluid may show a minor degree of lymphocytosis but its analysis is of little help.

Carotid angiography is the preferred neurosurgical diagnostic technique. The appearance of the cyst is that of a totally avascular lesion smoothly displacing surrounding vessels which exhibit none of the abnormalities often found on the fringes of an avascular neoplasm. Pneumoencephalography may be hazardous in the presence of a supratentorial mass lesion and ventriculography carries the risk of puncture of the cyst with dissemination of viable scolices. Fortunately, angiography is the procedure of choice for the localization of supratentorial masses in most clinics. Electroencephalography may be helpful if a characteristic pattern is found. This is usually an area of localized electrical flattening surrounded by a fringe of delta activity.

Treatment of the intracranial cyst is surgical. It is preferable not to rupture the cyst by means of needle biopsy or during delivery in order to prevent dissemination of viable scolices. Recur-
Hydatid Cyst of the Brain

449

rence following rupture and multiple scolex implantation is rapid. As an aid to delivery various techniques have been described. Arseni and Samitca recommend elevation of intracranial pressure by means of jugular compression or coughing after incision of the cortex. Araña-Iniguez and San Julián describe inflation of the contralateral ventricle and injection of saline between the cyst wall and the brain. Although some wash the cyst bed with formalin to sterilize the area, others fear damage to the underlying brain by this procedure. Attempts to find a less toxic sclocide are in progress.

In the present case no dissemination, apparent to this date, followed the inadvertent rupture of the cyst. Whether the surgical field was sufficiently confined to prevent enclodgement of viable scloces or whether this particular cyst was sterile is not known.

Summary

This is the case report of a cerebral hydatid cyst successfully treated by operation. Correct preoperative diagnosis would have been facilitated by a greater awareness of the possibility of echinococcus infection. The diagnosis of echinococcus disease and the mechanism, pathology and treatment of its cerebral form are discussed.

The author gratefully acknowledges the help of Dr. M. Biddle and Dr. T. Kurze in the preparation of this manuscript.

References