

INTRACRANIAL CYSTICERCOSIS

Report of a Case with Operation

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THE possibility of infestation of the alimentary tract by the pork tapeworm (*Taenia solium*) is well recognized, particularly in those areas and countries where poor sanitation and lack of meat inspection exist. Less commonly recognized clinically is the fact that the larval form of *Taenia solium*, i. e., *Cysticercus cellulosae*, may also infest the nervous system and its coverings.

Africa and Cruz¹ quote Vosgien who reviewed 107 cases of *Taenia solium* infestation and found that the nervous system was involved in approximately 40 per cent. Dressel² observed that the brain was affected 72 times in 87 cases, or 82 per cent. Hennenberg,³ in reviewing 1408 autopsies at the Charité in Berlin, reports only 3 cases of cysticerci of the brain having been found. Hare⁴ of this country relates 2 cases in a large series of autopsies from the Presbyterian Hospital and the Neurological Institute of New York. Arana and Asenjo⁵ of Chile found 25 cases of brain cysticercosis in a total of 202 intracranial tumors.

While the relative paucity of intracranial infestation in this country offers testimony to the sanitary systems in effect, such infestation is nevertheless possible.

MacArthur⁶ referred to this condition as a possible cause of epilepsy and Forster⁷ reported a case of insanity with epilepsy following infestation by *Cysticercus cellulosae*. Brailsford,⁸ in 1925, showed that evidence of the parasite could often be obtained by radiography. He intimated that four or five years may elapse before the deposition of calcium in the cysts occurs, if indeed such deposition occurs at all.

The diagnosis of intracranial cysticercus may be difficult, largely because the infestation may be limited to one lesion or due to multiple lesions. Likewise, the infestation may involve the meninges, the cortex, or the white matter. The lesion may be found in the ventricular system as well and cause obstructive symptoms.

Kuchenmeister, as reported by Krause,⁹ noted that in 88 cases of brain cysticercosis, the meninges were involved in 49, the cortex in 59, the ventricles in 18, and the white matter in 19 cases.

The symptomatology is extremely variable since it is dependent upon the location of the lesion or lesions. The number of the cysts and/or their location may vary to influence the clinical picture. While racemose clusters of the cysticerci are most common in the basilar cisternae, those in the supratentorial regions are usually intraparenchymal⁶ and may give signs referable to these areas.

The state of the larva may also alter the clinical findings. MacArthur believes a patient may live for years with cysts within the cerebrum and experience little disturbance. However, when the parasite dies, symptoms may be caused by toxic effects or by an increase in the size of the cyst. The flaccid and unpalpable cyst absorbs fluid at that time and tends to become enlarged and tense.

When the cysticercus develops in the skin or muscle, the demonstration of the larval form of *T. solium* in the excised cyst may clarify the diagnosis quickly in a patient suspected of this condition. Since calcification of the cysts occurs more rapidly in the skin and muscle when these organs are involved, it may be possible to demonstrate the presence of this condition by the roentgenogram. Skin tests and complement fixation tests are generally regarded as unsatisfactory.

The difficulties encountered in arriving at this diagnosis in one patient are related here. Until the lesion was disclosed at operation, it was believed the findings were consistent with a diagnosis of meningo-encephalitis. The chronicity and the clinical course appeared to substantiate this impression.

Case Report

A 32 year old woman, born in Italy, had emigrated to the United States at 11 years of age. She lived in a nearby large city and had not left the country at any time. She was first seen at the Cleveland Clinic on August 19, 1944 complaining of pain on the left side of the face and neck and in the suboccipital region, of three months' duration. The strain of a difficult domestic situation caused her to be extremely nervous. There was a complete lack of objective findings. A functional basis for her complaints was entertained at that time by one examiner.

She was again seen on June 22, 1946 complaining of headaches, pain in the left face and left ear and in the occipital region. There was an associated stiffness of the neck. She had developed a lateral diplopia in the previous three weeks. There had been some nausea but no vomiting. A general physical examination disclosed no abnormalities. Pertinent findings in the neurological examination showed bilateral papilledema of 2 to 3 diopters. The ophthalmologist was of the opinion that the fundus had the appearance of a severe neuroretinitis. There was a hyperreflexia of the left lower extremity only. The Babinski test was positive on the left while the Romberg test was negative. X-ray of the skull showed no abnormality. It was believed that, while a meningo-encephalitis must be considered, the lesion was more likely an intracranial neoplasm affecting the right parietal area. To differentiate between an inflammatory or neoplastic lesion, a pneumo-encephalogram was done on June 14, 1946. The ventricular system was well outlined and was normal in shape and position with no displacement. The third and fourth ventricles as well as the aqueduct were normal. The basilar cisternae were normal. There was no evidence of a space-filling lesion. Examination of the fluid showed 300 white cells of which 60 per cent were lymphocytes, with no globulin, and a total protein of 41 mg. per cent. It appeared that the patient had an inflammatory lesion in the nature of a meningo-encephalitis. She was given three x-ray treatments during which she experienced discomfort in her left arm. The appearance of the optic disks improved considerably. Before she was discharged from the hospital, another lumbar puncture was made on June 22, 1946. Although she seemed clinically improved, there were 280 white blood cells in her spinal fluid. The protein was 45 mg. per cent. Special cultures were made on the fluid which were all negative for bacteria or fungi.

She was readmitted to the hospital on July 15, 1946 stating that the frequency of pain on the left side of her head was less but the intensity was greater. Lumbar puncture showed increased pressure with a high Ayala index. On July 18, 1946, a left subtemporal decom-

pression was done, and following this operation, the patient was relatively free of headache. A pneumo-encephalogram was again made on July 24, 1946, and showed no evidence of an intracranial lesion. Postoperative visual fields indicated some enlargement of the blind spots bilaterally. There was no change in the neurological examination. The spinal fluid contained 30 lymphocytes. Following some x-ray treatment, the patient was completely free of pain. At the time of discharge on August 5, 1946, the spinal fluid pressure was normal.

Subsequently, she was readmitted to the hospital for several short periods during which lumbar punctures were done. The pressure remained in the vicinity of 190 mm. of water and contained from 65 to 150 white blood cells. She was given a series of six typhoid shocks and had a good reaction to this treatment. She was dismissed from the hospital on October 6, 1946, apparently improved.

She was seen as an out-patient on December 4, 1946. Her headache was gone. She was troubled only by a slight intermittent pain behind the left ear. Neurological examination was entirely within normal limits.

On August 8, 1948, she was again admitted to the hospital because of postauricular pain on the left as well as a swishing noise in her left ear. Neurological examination revealed the presence of a low-grade papilledema, and a moderate degree of tension in the left subtemporal decompression. Lumbar puncture revealed an initial pressure of 220 mm. of water. Twenty-four lymphocytes were present in this fluid and 33 mg. of protein. The smear and culture were negative for organisms. The stained sediment showed no tumor cells. The spinal fluid sugar was 49 mg. per cent with 693 mg. per cent chloride. A pneumo-encephalogram again was made which showed a diffuse dilatation of the ventricular system with no evidence of an obstructing lesion. Because of the papilledema and increased pressure, a right subtemporal decompression was performed on August 12, 1948. Histoplasmin and coccidioidin

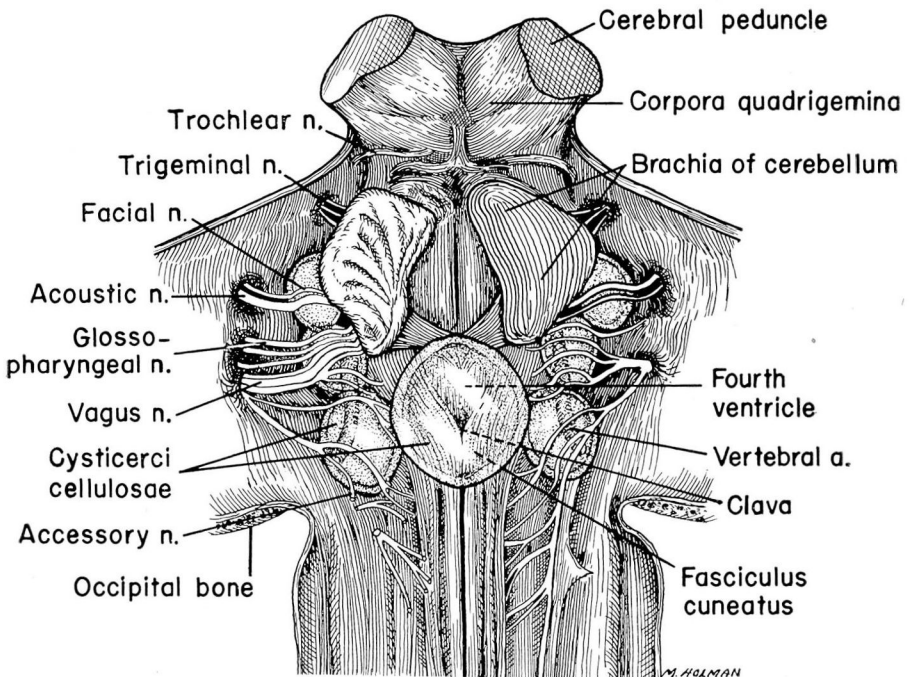


FIG. 1. Schematic drawing indicating position of cysts found at operation.

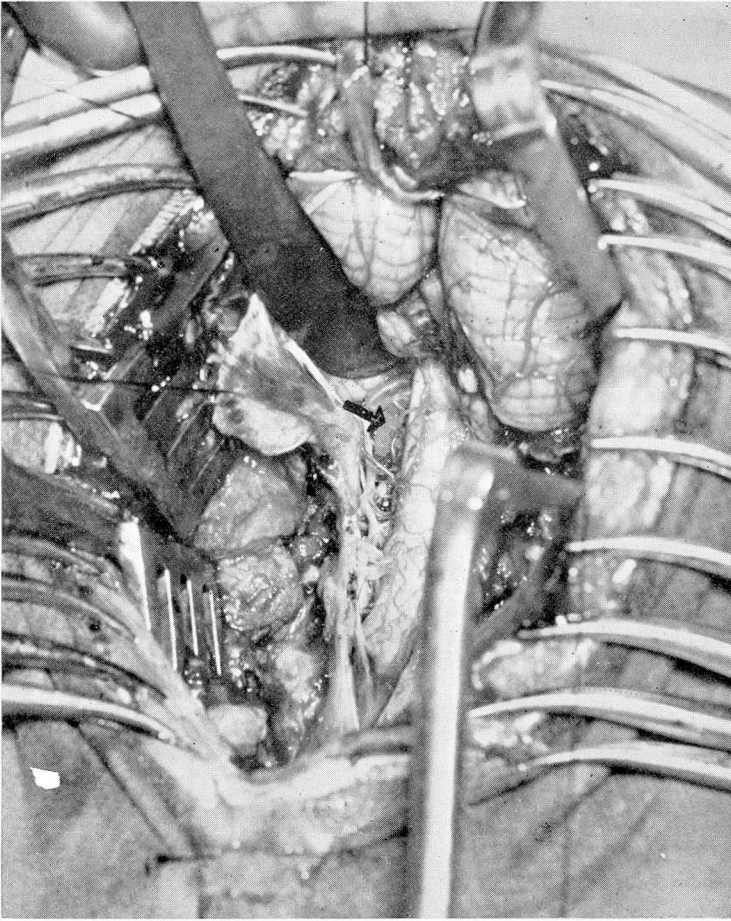


FIG. 2. Operative exposure on left showing one cyst (indicated by arrow) beneath the vagal root filaments.

skin tests were negative. The patient made a satisfactory recovery and was discharged on August 21, 1948. The diagnosis was still considered to be chronic meningo-encephalitis.

She was symptom-free until October, 1948, when, while vacationing, she developed a severe headache. There was nausea but no vomiting. The subtemporal decompression areas were extremely tense. She was observed in another hospital where a diagnosis of increased intracranial pressure, cause unknown, was made, and lumbar puncture performed to relieve the pressure.

She was returned again to the Cleveland Clinic Hospital on October 18, 1948. The only pertinent physical finding was the presence of a mild degree of papilledema and a bulging of the decompressions. Lumbar puncture was repeated which showed an initial pressure of 360 mm. of water. A pneumo-encephalogram was again made on October 22, 1948 which indicated the presence of a nonobstructive internal hydrocephalus of considerable degree and a lack of cortical filling. The lateral ventricles as well as the third and fourth ventricles

were well visualized. In the course of the pneumo-encephalogram proceedings, the flow of cerebrospinal fluid appeared to come in spurts of approximately 20 cc. volume as if an intermittent obstruction was present. The dilatation of the ventricles had increased considerably over the encephalogram of August 12, 1948. A suboccipital exploration was elected on October 27, 1948. Upon opening the arachnoid, a large round opalescent cyst was dislodged from the midline in the area of the fourth ventricle. This fluid-containing cyst was approximately 3.0 cm. in diameter, and lay free in the subarachnoid space. It was at that time that the clinical diagnosis of an intracranial parasitic infestation was made. This diagnosis was confirmed by subsequent microscopic study. Numerous other cysts were noted in the posterior fossa as depicted in the drawing (figs. 1 and 2). The pathologic report indicated later these were *cysticercus cellulosae*. A total of seven such cysts were removed at this operation with no further lesion being observed. Exploration of the fourth ventricle revealed a free flow of fluid mixed with air emerging from the aqueduct area. The patient had a relatively uneventful convalescence and was discharged from the hospital on November 7, 1948. Spinal fluid pressure at the time of discharge was 170 mm. of water.

She was examined again on December 13, 1948 and was free of pain. However, she was troubled with vomiting which occurred shortly after eating. The decompressions were soft and the optic disks flat and pallid. The deep reflexes were slightly hyperactive. She was given symptomatic treatment.

When seen again three weeks later, the patient stated that the decompressions had begun to bulge again in both temporal and suboccipital areas. There were short periods of disorientation and some staggering in walking. Attacks suggestive of right Jacksonian type were present. The optic disks showed some slight degree of elevation. The spinal fluid pressure at this time was 270 mm. of water. Her symptomatology increased and she was again admitted to the Cleveland Clinic Hospital on January 17, 1949, the pertinent findings consisting of bulging of the decompression sites, hyperactive deep reflexes, equivocal bilateral Babinskis, and a lateral nystagmus. A lumbar puncture was done again which showed a pressure range from 270 to 400 mm. of water. On January 24, 1949, a pneumo-encephalogram was made which showed a complete block in the posterior fossa. The suboccipital decompression was then reopened and a remarkable degree of scarring was present throughout the posterior fossa. The entire cervical cord, the medulla and the base of the cerebellum were enveloped in a funnel-shaped, fibrinous appearing membrane approximately 1 to 2 mm. thick. This membrane was dissected free until fluid was observed to flow freely from the fourth ventricle region. Further inspection revealed no evidence of cysts. The Torkildsen procedure was elected, the tube being brought subosteally from the posterior portion of the right lateral ventricle into the cisterna magna area. Dye studies before closure indicated the patency of the tube.

The first day postoperatively, the course was satisfactory. The second day postoperatively, the decompression areas were noted to be bulging and lumbar puncture was done. The normal appearing spinal fluid of the day before was replaced with a thick, syrupy, yellowish fluid. Supportive treatment was instituted. On the third postoperative day, a similar finding was made both in the lumbar region and in the ventricular region. The patient lapsed into unconsciousness, grew progressively worse and died on January 28, 1949.

The postmortem examination was limited to a reopening of the incision which disclosed a necrosis of pronounced degree in the inferior cerebellar area. A considerable amount of liquefaction was also found. In the exploration of the posterior fossa, a group of cysts was recovered from the prepontine cistern area. Incisions were made from below into the posterior portion of the ventricles and a limited examination failed to uncover the presence of any cysts in the ventricles although obviously, the examination was inadequate.

Comment

In this patient repeated examinations failed to show any evidence of *Taenia solium* infestation even after the diagnosis had been established. X-ray studies of the soft tissue showed no calcifications. Stool studies disclosed no

abnormalities. From the history, the variable clinical picture, the laboratory findings, the cellular response in the spinal fluid as well as response to fever and x-ray therapy, it was believed that this patient had a meningo-encephalitis of undetermined type until the diagnosis was clarified at operation.

Arana and Asenjo⁵ believed that the presence of an internal hydrocephalus, absence or displacement of the ventricular system, especially of the aqueduct, partial obliteration of the aqueduct and fourth ventricle with the presence of air in both and in the cisterna magna, associated with clinical, biological and laboratory examination were diagnostic of cysticercosis of the brain. The various air studies made in this patient were carefully reviewed and evaluated, but no changes suggestive of those described by these writers were found.

Generally, surgical experiences in this condition have not been encouraging. Arana and Asenjo⁵ as well as Ray¹¹ and Ritchie¹¹ of this country have reported cases benefited by surgery.

Dandy¹² and Cairns¹³ each report 2 cases in which operation was unsuccessful. To this group we must add our own unsuccessful experience.

When an isolated cyst is the cause of a focal disturbance, surgical intervention may be of some benefit. When the obstruction to the cerebrospinal circulation is due to the presence of intraventricular or multiple cysts in the posterior fossa, it is believed that surgical removal of as many cysts as possible in conjunction with the use of the Torkildsen procedure may be of value in an otherwise discouraging situation. Further experience is necessary before the effectiveness of this suggestion can be evaluated. It might well serve as a temporizing measure, if one recalls MacArthur's observation that a later resorption of fluid from the cyst occurs. Whether resorption of fluid takes place in the same manner when the cysts are located in the cerebral parenchyma or sub-arachnoid space as it does when the skin or muscle are involved, remains to be demonstrated.

The clinical course here was suggestive of an inflammatory reaction, i. e., possibly toxic, which led to the diagnosis of meningo-encephalitis. The death of the larva may well have been responsible for the findings. The development and progression of obstructive symptoms and signs may be inferred to have been due to the enlargement of the cysts by fluid absorption. Whether or not a reabsorption of the fluid from the cysts would have occurred later cannot be ascertained.

Conclusion

1. A patient with cysticercosis of the posterior fossa was unsuccessfully treated surgically.
2. The use of the Torkildsen procedure in conjunction with a thorough evacuation of the cysts is suggested for this type of case when multiple cysts are present in the posterior fossa.
3. When the obstructing lesion is intraventricular in position and craniad the fourth ventricle, the Torkildsen procedure alone may suffice.

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