FROM THE TREASURE TROVE OF NEUROLOGY INDIA

Cysticercosis of the brain

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Introduction

Cysticercosis of the brain is caused by the presence of the larval form of *Taenia solium* in the parenchyma of the brain, the meninges, or in the ventriculosubarachnoid pathway.

According to Olive and Angulo-Rivero,[10] cysticerci were found by Rumler in 1588 in the dura of an epileptic patient. An account of meningeal racemose cysticercosis of the base of the brain was given by Virchow in 1860. In 1862, Griesinger drew attention to cysticercosis as a cause of convulsive disorder. The epoch making study of MacArthur[8] firmly established the role of cysticercosis in the causation of chronic cerebral seizures.

The earliest to report cerebral cysticercosis in India was Armstrong according to Singh *et al*.[12] Subsequently, Eliott and Ingram in 1911, Krishnaswami in 1912, Menon and Velliath,[9] and Gault and Balasubramaniam[5] among others reported cases of cerebral cysticercosis.

Material

This presentation is a study of 25 proved cases of cerebral cysticercosis treated in the department. The recorded history, physical signs, clinical course of the disease, and operative findings were analyzed. Histopathological confirmation of the intracranial lesions was obtained in 16 patients, of whom 5 were subjected to autopsy. In 9 other cases, the diagnosis was established on the basis of biopsy of subcutaneous nodules or by the presence of intestinal taeniasis associated with radiological changes such as ventricular filling defects in the pneumoencephalogram [Table 1].

Table 1: Diagnostic basis of cerebral cysticercosis

Diagnosis based on	No. of cases
Cysticerci in cerebral lesions	16
Cysticerci in subcutaneous nodules	7
Intestinal taeniasis plus Radiological changes	2

Age

The youngest patient was aged 5 years and the oldest was 56 years. The average age of the patients was 25 years. In Gonis series, a majority of patients were between 20 and 40 years of age. Among 132 cases reported by Stepien,[14] the age of the patients ranged from 7 to 63 years. About 72% of the cases were between 21 and 50 years. Table 2 shows age incidence in this present series.

Table 2: Age incidence of 25 cases of cerebral cysticercosis

Age (years)	Male	Female
0-10	1	1
11-20	5	4
21-30	4	2
31-40	3	2
41-50	1	1
51-60	0	1

Sex

There were 14 males and 11 females in the present series. Both sexes were equally affected according to Goni[6] and Lombardo and Mateos.[7] In Stepien's[14] series, there were 59.8% females and 40.2% males. Among 65 cases of Arseni and Samitca,[1] 37 were males and 20 were females.

Dietary habits

Ten patients were nonvegetarian and two were vegetarian among the 12 cases where this information was recorded. There was no mention regarding pork consumption, except in two cases in whom it was negative. Though in the literature considerable stress is laid on pork consumption as an etiological factor, this has not been substantiated by this study. It is known that *Taenia* solium ova can be consumed through contaminated water and vegetables, and it is not necessary for the patient to be a pork eater. Evidence of tapeworm infestation was present in five cases in this series.

Duration of symptoms

The shortest duration of symptoms was 3 weeks and the longest was 11 years. The average duration was 1.9 years. The corresponding figures were 2 weeks and 16 years in Stepien's[14] series. About 60% of his cases sought medical help within 1 year of onset of symptoms. Fourteen of 25 cases in this series (56%) reported for treatment within 1 year of onset of symptoms.

Symptoms and signs

Epilepsy, headache, and diminished vision were the most common symptoms. Vomiting and mental symptoms were reported by a few.

Epilepsy was the commonest symptom being present in 20 patients (80%). Seizures were generalized in 11 and focal in 5 cases. The exact nature of seizures was not known in four cases. In 15 of these patients, epilepsy was the first symptom. The longest duration of fits was 11 years and the shortest was 19 days. In 450 cases described by Dixon and Lipscomb,[4] epilepsy was present in 92% of cases, and it was the first symptom in 76% of cases. In Stepien's[14] series, epilepsy was present in 36.5% of cases. In all, 31 of 65 cases of Arseni and Samitca[1] had epilepsy.

Headache was present in 18 cases (72%). In Stepien's[14] series, headache occurred in 97.7% of cases; 30 of 31 cases of Lombardo and Mateos complained of headache and 52 of 65 cases of Arseni[1] had headache.

Diminished vision was complained of by 18 patients in the present series. Vomiting was the next in frequency (40%). Two patients had mental symptoms.

Papilledema was present in 13 patients, and in 3 there was optic atrophy. Papilledema occurred in 84% of Stephien's[14] patients and in 25 of 31 cases of Lombardo and Mateos.[7]

Focal neurologic deficits such as motor weakness, sensory changes, and abnormalities in the reflexes were present in 15 cases.

Subcutaneous nodules were present in nine patients. In one patient, subcutaneous nodules developed after his discharge from hospital. In one patient, in whom cysticerci were excised from the right temporal lobe, subsequent autopsy showed diffuse multiple cysticerci in the brain. There were also numerous cutaneous nodules. Six of the cases with cutaneous nodules were in the group presenting mainly with seizures. Diagnosis was made after demonstration of cysticerci in these nodules.

Blood and cerebrospinal fluid

Blood eosinophil count of 10% or more was found in five cases. Eosinophilia has been reported by different kind of workers in a varying proportion of patients. Dixon and Hargreaves[3] found eosinophilia in about 10% of their 284 cases. Arseni and Samitca[1] found eosinophilia of over 6% in 13 of their 65 cases.

Cerebrospinal fluid studies were done in 21 cases. Biochemical abnormality was present in nine of these. In 12 cases, the spinal fluid was normal. In nine cases showing cerebrospinal fluid abnormality, the protein content ranged from 55 to 400 mg%. The range of glucose levels was 10 and 90 mg%. White cell count of more than 5/mm^3 was found in six cases out of these nine. There were five additional cases showing raised cell count in the cerebrospinal fluid with protein and sugar values remaining within the normal limits. There was no eosinophilia in the cerebrospinal fluid in this series. Changes in cerebrospinal fluid consisted of lymphocytic pleocytosis which occurred in 18.9% of cases in Stepien's[14] series. Eosinophilia in the spinal fluid is said to indicate the presence of cysticercus lesion in the nervous system. However, failure to find spinal fluid eosinophilia is not against the diagnosis of cerebral cysticercosis. According to MacArthur,[8] the cerebrospinal fluid may remain unaffected even with profound cerebral disturbance. Dixon and Hargreaves[3] agree with this view.

Classification

Stepien and Chorobski,[13] in a study of 23 personal cases and 71 cases reported in the literature, grouped cases of cerebral cysticercosis into three categories. Group I comprised case with cysts often single, which caused symptoms similar to brain tumor. The focal symptoms varied depending on the area involved. Group II cases had cysts, which were usually numerous and produced diffuse swelling of the brain or multiple focal symptomatology. In the third group, the parasites caused chiefly hydrocephalus. Dixon and Lipocomb[4] stated that three distinct clinical patterns related to pathological factors at work emerged from their study of 450 cases. In Group I, the illness started soon after the probable date of infection, and the course was that of an acute febrile disorder during or soon after which signs of raised intracranial pressure developed. In Group II cases, signs of intracranial hypertension developed gradually during the course of the disease. The clinical picture was that of intracranial hypertension with fits. Patients in Group III developed hydrocephalus gradually with characteristic recurrent attacks of acute rise in intracranial pressure, when ventricles contained free lying cysts, and without such attacks when arachnoiditis was the feature. Goni,[6] in his Mexican series, found it convenient to group them into three categories: (a) parenchymatous type of two forms, acute and chronic, the acute resembling encephalitis with raised intracranial pressure, when diffuse, and without raised pressure when localized, (b) ventriculosubarachnoid type, which may be purely ventricular, purely subarachnoid, or ventriculosubarachnoid, and (c) mixed type. All the authors have mentioned that it is difficult to group the cases into watertight compartments, and many cases are on the borderline between the two groups. It is also noted that the classification of cerebral cysticercosis is not yet standardized. In this study, cerebral cysticercosis has been catergorized into the following four groups:

1. **Meningocephalitis:** Cases in this group presented with relatively short history with rapid deterioration of consciousness and some signs of meningeal involvement. Most of these ran a fatal course. This group is comparable to Group II of Stepien and Chrobski[13] and Stepien.[14] In the latter series of 132 cases, this group constituted 25%. These cases also correspond to Goni's diffuse, acute parenchymatous form. Eighteen of 65 cases of Arseni and Samitca[1] simulated meningoencephalitis. In the present series, there were seven cases in this group. The age of these patients ranged from 5 to 42 years. Only one was a child. All the remaining were between 18 and 42 years of age. There were four females and three males. The duration of symptoms was short, ranging from 3 weeks to 6 months. One patient had episodes of headache for 5 years which became severe and constant 4 months before admission. All patients had fever, altered level of consciousness, and papilledema. Focal neurologic deficits were present in five cases. The cerebrospinal fluid showed raised protein and lowered sugar in five cases, but the white cell count was elevated only in four of them, ranging from 11 to 500/mm^3. Four of these cases were operated upon because of radiological localization of a possible mass. During operation, two of them had cysticerci in the fourth ventricle. Both expired in the postoperative period. Autopsy was done on one of them, and there was cerebral edema, with cysts in different parts of the brain. Of the other two operated patients, one had cysts in the left frontal region which were excised. She could not be followed subsequently. The other patient remained unconscious and died 2 months after craniotomy and excision of multiple cysts from the right temporal lobe. Autopsy showed multiple cysticerci scattered throughout the brain. Among the three unexplored cases, two patients died in the ward before a definite diagnosis was arrived at. Autopsy showed generalized cerebral cysticercosis in both cases, one of them having in addition cysts in the liver and a tapeworm in the intestines. One patient who presented with signs and symptoms of meningoencephalitis did not reveal any evidence of cysticerci during his stay in the hospital. He was treated with anticonvulsants and dehydration measures with which he improved. A few weeks after discharge from the hospital he developed cutaneous nodules and they were excised and were reported to be cysticerci
2. **Brain tumor syndrome** (nine patients): These cases showed signs and symptoms of raised intracranial pressure with or without seizures and usually exhibited focal neurologic deficits. Investigations revealed cysticerci, usually forming a mass lesion located in the cerebral hemispheres or cerebellum, or a single cyst occupying the fourth ventricle causing obstruction. This group corresponds to Stepien's[14] Group I. About 42% of his cases belonged to this group. This also corresponds to Group II of Dixon and Lipscomb[4] and to chronic parenchymatous form of Goni.[4] Nine cases in our series represented this group. The age of the patients ranged from 9 to 56 years. Six were males and three were females. The duration of symptoms ranged from 1 month to 11 years. Seven of these patients had headache, eight had fits, eight had focal neurological deficits, and seven had papilledema with signs of raised intracranial pressure. The location of cysts as found during operation was cerebral hemispheres in six, cerebellum in two, and fourth ventricle in one
3. **Basal arachnoiditis** (one patient): There was one patient in this category who presented initially with chronic hydrocephalus and neurologic deficits. A third ventricular tumor was radiologically suspected. Exploration of the lateral and third ventricles was negative and a ventriculoatrial shunt was done. On a subsequent admission, her condition deteriorated and she expired. Autopsy revealed basal cysticercus arachnoiditis
4. **Convulsive disorder** (eight patients): Patients representing this group had mainly seizures. They did not exhibit signs of focal neurologic deficits or raised intracranial pressure. The diagnosis was confirmed by demonstration of cutaneous cysticerci in six cases and intestinal taeniasis in two.

Surgical treatment and operative findings

Verco was probably the first to remove a cysticercus from brain.[14] There were many subsequent attempts on removal of these cysts at operation, but many surgeons expressed themselves against surgical removal of cysticerci. Dandy[2] felt that surgical treatment was of no value. In 1949, Stepien and Chorobski[13] published the results of operation of 23 personal cases and 71 from the literature. They concluded that operative treatment was followed by good results in 50% of cases.

Fourteen patients were operated upon in this series. Multiple cysts forming mass lesions were excised from different regions of the cerebral hemispheres in eight cases. These were two cases in which multiple cysts were found in the cerebellum. A solitary cyst in the fourth ventricle was found in three cases. In one case, exploration through a right frontal craniotomy was negative, and a ventriculoatrial shunt was done. The patient subsequently expired and autopsy revealed hydrocephalus due to basal meningitis with cysticerci.

Results of surgery

Four of the 14 operated patients expired during the postoperative period in the hospital. Of these, three belonged to the meningoencephalitis group and one belonged to the basal arachnoiditis group. Of the other 10 operated cases, 9 belonged to Group II. There was no postoperative mortality in this group. One patient expired subsequently 9 years after operation. Five patients were followed up for periods ranging from 4 to 16 years. Three of them have shown improvement in terms of freedom from fits and ability to lead an independent life. Two patients have remained unchanged and they continued to have seizures though they are on anticonvulsants. No follow-up is available for the other three patients. The overall operative mortality is about 28%. It is significant that there is no operative mortality among the brain tumor syndrome group, who apparently have a localized mass of cysts or a single cyst, and who do well after surgical excision of the lesion. On the contrary, those who have a diffuse distribution of cysts and present as meningoencephalitis have a higher operative mortality (75%).

Pathology and pathogenesis

The gross and microscopic pathology of neurocysticercosis is fairly well established. The live larvae produce very little reaction and are generally neurologically silent. After the death of the larva, the cyst is surrounded by a thick zone of infiltrate with polymorphs, lymphocytes, macrophages, epithelioid cells, and foreign body giant cells. This inflammatory process ultimately progresses to the formation of scars. It is the formation of multiple scars in the brain and meninges which causes seizures.

Fibrosis and inflammatory reaction were minimal in silent cases of cysticercosis cerebri. Showramma and Reddy[11] found that in contrast to silent cases, tissue reaction was intense in symptomatic cases. Fibrosis and inflammatory reaction according to these authors are directly proportional to the degree of degeneration of cysts. Dixon and Lipscomb[4] found the main types of lesions described below:

1. Thin-walled cysts with living larvae
2. Cysts with degenerated larvae and caseous material with surrounding inflammatory host tissues
3. Calcareous nodules of hardly recognizable structure.

A diffuse distribution of cysts in many areas of the brain was found in four cases at autopsy. Cysticerci forming a mass lesion in the cerebral hemisphere was found in seven cases, and in the cerebellum alone in two cases. There were two cases with solitary cysts in the fourth ventricle. There was one case of basal cysticercus with arachnoiditis. The histopathology has been reviewed in 15 cases, but the larval parts could be seen only in seven cases. Inflammatory changes in the cyst wall consisted of lymphocytes, plasma cells, eosinophilic cells, epithelioid cells, and foreign body giant cells. Fibrosis was present in all cases. Microscopic calcification was seen only in one case.

Summary and Conclusion

In all, 25 cases of neurocysticercosis have been reviewed with regard to clinical features, biochemical abnormalities in the blood and cerebrospinal fluid, and operative and autopsy findings. Patients with cerebral cysticercosis present in one of the four following forms: (1) meningoencephalitis, (2) brain tumor syndrome, (3) basal arachnoiditis, and 4) convulsive disorder. Eosinophila in blood and cerebrospinal fluid has not been a prominent feature in this series.

Prognosis in the meningoencephalitis group is poor. Patients presenting as brain tumor have a good prognosis if the space occupying lesion formed by cysticerci is excised.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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