

Original Investigation

The Surgical Value of Neurocysticercosis: Analyzing 10 Patients in 5 Years

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ABSTRACT

AIM: To investigate the indications and clinical value of the surgical therapy of neurocysticercosis (NCC).

MATERIAL and **METHODS:** We retrospectively reviewed all 10 patients with NCC treated surgically from January 2009 to January 2014 in our institute. The NCC types included parenchymal cysticerci in 6 cases, subarachnoid cysticerci in 3 and ventricular cysticerci in 1.

RESULTS: All 10 patients underwent resection of cysticerci through craniotomy. Parenchymal NCC included an isolated lesion in 4 cases and multi-lesions in 2 cases, who respectively had coexistence of metastasis and cysticerci and a positive history of tuberculosis. In all 3 patients with arachnoid NCC, two cases underwent lesion resection and the other one underwent cyst resection and lamina terminalis fenestration. In one case with intraventricular NCC, a ventriculoperitoneal (VP) shunt was placed as the initial treatment. Due to failure of the shunt, the secondary surgery of cyst excision via open craniotomy and VP shunt placement were performed simultaneously.

CONCLUSION: For parenchymal cysticercosis, the surgical aim is to remove the lesion, treat the medically intractable epilepsy, and establish the diagnosis and subsequent treatment. As for extraparenchymal NCC, surgical therapy is a valuable treatment as an alternative due to its fatal complications. For subarachnoid cysticercosis, because of its higher failure rate in simply VP shunt, the open craniotomy could directly remove the cyst and efficaciously relieve hydrocephalus. For the patient with intraventricular NCC associated with hydrocephalus, the effect of a simple VP shunt seems unsatisfactory, and craniotomy for cyst resection is necessary.

KEYWORDS: Neurosurgery, Cysticercosis, Central nervous system

■ INTRODUCTION

Neurocysticercosis (NCC) is a common intracranial parasitic disease in developing countries, with a predilection for the brain parenchyma, subarachnoid space and ventricles. Due to its different locations, numbers, and developmental stages, the intracranial complications and clinical manifestations vary from person to person. The standardized treatment of NCC is still controversial (10). So far, the medical treatment of cysticercosis in the central nervous system is the primary

choice. However, the data regarding the surgical management of NCC is quite insufficient, especially for extraparenchymal NCC. Many scholars advocate that anti-parasitic therapy should be considered as a priority in parenchymal NCC and steroids and anti-epileptic drugs are also chosen commonly in case of brain edema and seizures caused by the cysticerci (2,5,10). In contrast, extraparenchymal NCC is often associated with more severe complications such as hydrocephalus, arachnoiditis and ventriculitis. Yet, there is still no clear consensus on its treatment in the literature (9,10). The



Corresponding author: Chao YOU E-mail: shionaries@126.com aim of this article was to explore the indications and clinical value of the surgical treatment of NCC through analyzing the patients with NCC treated by surgery in the past 5 years in our institute.

■ MATERIAL and METHODS

We retrospectively reviewed all 10 patients with NCC treated surgically from January 2009 to January 2014, including 5 males and 5 females with an age range of 16-54 years and a mean age of 36 years, in the department of Neurosurgery at West China Hospital, Sichuan University. The clinical presentations included headache in 9 patients, seizures in 3, nausea and vomiting in 3, movement disorders in 3 and consciousness disorders in 3. The lesion locations were based on imaging. Plain computed tomography (CT) of the brain was performed in 4 patients, and magnetic resonance imaging (MRI) in all 10 patients, 7 of whom underwent contrast-enhanced MRI. Among these patients, we found parenchymal cysticerci in 6 cases, subarachnoid cysticerci in 3 cases and ventricular cysticerci in 1 case. Besides, simultaneous parenchymal and subarachnoid cysticerci were found in the patient with intraventricular cysts. Calcifications were observed in 2 cases, arachnoiditis in 1 case, hydrocephalus in 4 cases and cerebral edema in 5 cases.

The Laboratory Examination

Cysticercosis IgG was measured in just 2 cases preoperatively, and only 1 case was positive. A postoperative cysticercosis IgG test was performed in 9 patients and all were positive.

The Medical Treatment

Steroids (methylprednisolone or dexamethasone) were administered in 5 patients because of perifocal edema or leptomeningitis. Anti-epileptic drugs (sodium valproate) were used in 8 patients, among whom 5 patients used the drugs for

prophylaxis. Anti-parasitic drugs (albendazole or praziquantel) were preoperatively used in 1 patient with a definite diagnosis of NCC, and these were administered postoperatively in the other 9 patients after the confirmed pathological results of cvsticerci.

■ RESULTS

The Surgical Treatment

All 10 patients underwent resection of cysticerci through craniotomy, and 1 patient underwent lamina terminalis fenestration simultaneously. In 1 patient, a ventriculoperitoneal (VP) shunt was chosen as the initial surgical procedure.

Intraparenchymal Cysticerci

All 6 patients with parenchymal NCC underwent craniotomy and resection of the cysts. Based on the preoperative imaging, isolated lesion in 4 cases and multi-lesions in 2 cases were demonstrated. In 5 patients, the parenchymal NCC were associated with severe perifocal edema. In the patients with isolated lesions, the cysticerci of 3 cases presented as annular nodular lesions, with obvious ring-like enhancement and a diameter of 0.8 cm, 1 cm, and 2 cm respectively, without apparent scolex. In a lesion 2 cm in size, multiple small annular nodules were found in the enhanced capsule (Figure 1A-C). The surgeries were performed with an initial diagnosis of metastasis. encephalopyosis and glioma. In another case with an isolated lesion, a mass about 4x5x4cm in size was demonstrated as hyper-densities with calcifications on plain CT and low signal intensity on both T1WI and T2WI. The preoperative presumed diagnosis of the lesion was a brain tumor. The pathology result revealed that the mass contained inflammatory, fibrous and keratinized materials; parasitic disease was considered and the cysticercosis IgG test was positive. In one of the two patients with multi-lesions, a mass of 2.5 cm in size, aggregated by numerous small cystic lesions without obvious

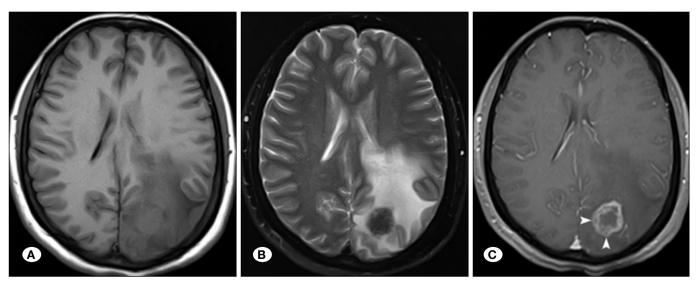


Figure 1: The MR imaging of a 39-year-old male with NCC. A) T1WI, B) T2WI, C) contrast-enhanced T1WI. A lesion with low signal intensity on T1 and T2WI was seen in the left occipital lobe with severe perifocal edema. The ring-like enhancement comprised by numerous annular nodules was demonstrated on contrast-enhanced T1WI (C. Arrowhead).

scolex and with apparent ring-like enhancement, was found. Due to the concomitant tuberculosis and negative result in the preoperative screening of parasite examinations, an intracranial tuberculous granuloma was considered as the initial diagnosis. The postoperative pathology confirmed the diagnosis of NCC. In the other patient, multiple cysts about 0.3-1 cm in size with ring-like enhancement and dispersed distribution were seen in bilateral hemispheres. Meanwhile a huge solid mass in the right occipital lobe, about 5x7x5cm in size, was demonstrated as well (Figure 2A-C). Combined with his history of lung carcinoma, intracranial metastasis was considered preoperatively. During the operation, the huge mass in the occipital lobe and some small nodules nearby

were removed. The pathology confirmed the occipital mass as metastatic adenocarcinoma and the small nodules as parenchymal NCC.

Arachnoid NCC

The cysticerci were found as cystic lesions in the suprasellar cistern in all 3 patients with arachnoid NCC. On MRI, the cysts had thin capsules with mild enhancement but absence of scolexes; meanwhile, an associated communicating hydrocephalus was seen in all 3 patients. The preoperative diagnosis was suspected to be an epidermoid cyst or craniopharyngioma (Figure 3A-C). Two cases underwent lesion resection via a pterion approach. The other one underwent

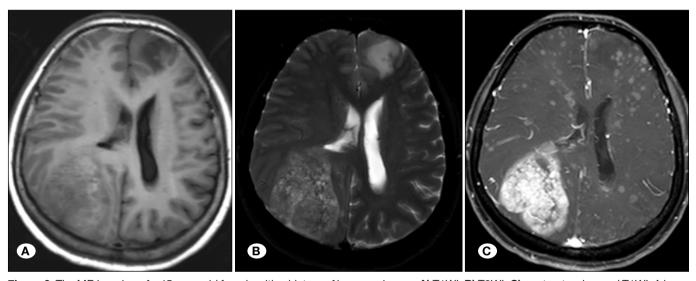


Figure 2: The MR imaging of a 45-year-old female with a history of lung carcinoma. A) T1WI, B) T2WI, C) contrast-enhanced T1WI. A huge mass with heterogeneous hypointensities on T1WI and non-uniform hyperintensities on T2WI was located in the right occipital lobe. On contrast-enhanced T1WI, an enhanced huge mass and multiple dispersed cysts with ring-like enhancement were seen. These lesions were all considered metastases, however, pathology surprisingly revealed the right occipital lesion to be metastatic adenocarcinoma and the adjacent small nodules to be parenchymal NCC.

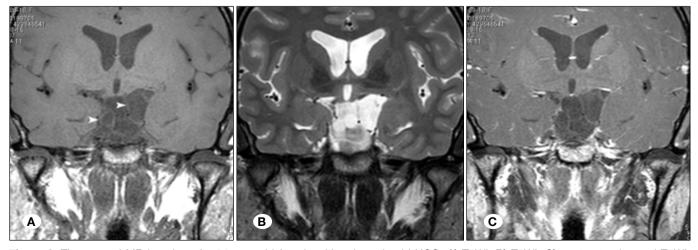


Figure 3: The coronal MR imaging of a 24-year-old female with subarachnoid NCC. A) T1WI, B) T2WI, C) contrast-enhanced T1WI. Multiple cystic lesions with long T1 and T2WI were found in the suprasellar cistern. The thin capsules of the lesions were demonstrated on T1WI with mild enhancement on contrast-enhanced T1WI. No visible scolex was found on imaging.

cyst resection through the falx approach craniotomy with simultaneous lamina terminalis fenestration.

Intraventricular NCC

Intraventricular NCC was found in one case with hydrocephalus. The cyst located in the enlarged left lateral ventricle, about 3 cm in size, adhered to the ependymal layer. Meanwhile, multiple calcified lesions were seen in the bilateral cerebral hemispheres and right lateral ventricle. A VP shunt was performed as the initial treatment but the ventricular shunt was shown to have failed based on the follow-up imaging (Figure 4A-C). Ultimately, the secondary surgery of simultaneous cyst excision via open craniotomy and VP shunt placement was performed. The consequent imaging proved the final improvement in hydrocephalus.

■ DISCUSSION

Neurocysticercosis is a common intracranial parasitic disease in developing countries. The lesions can involve the brain parenchyma or extraparenchymal region. Its clinical features and complications can vary along with its location, number, and stage of development (10). Seizure is the most common symptom of parenchymal cysticerci while hydrocephalus is rarely seen. Anti-parasitic and anti-epileptic therapies are the main treatment so far and standardized use of steroids is sometimes preferred in case of cerebral edema (2,5,10).

Extraparenchymal NCC, namely subarachnoid and intraventricular infection, has different clinical manifestations and prognosis than parenchymal NCC. There is no recognized standard treatment so far. Although some scholars have reported a certain benefit in treating subarachnoid and intraventricular cysticercosis using an anti-parasitic drug (albendazole), medical treatment of extraparenchymal NCC remains controversial. The reason focuses on its fatal complications such as hydrocephalus and leptomeningitis that can hardly be improved using medications only (1). Moreover, even when corticosteroids are used for prevention, anti-parasitic drugs will accelerate the inflammatory response during the process of cystic degeneration and lead to the ventriculitis or arachnoiditis (10). Exploring the value of surgical treatment of extraparenchymal NCC is therefore necessary.

Surgical treatment of parenchymal NCC is commonly chosen in the case of preoperative misdiagnosis as another disorder. In this series, all six cases with parenchymal cysticerci were misdiagnosed. We noticed that a solitary cyst with atypical form has a tendency to be misdiagnosed. In our 3 cases, the imaging findings of solitary cystic nodules included thick walls with ring-like enhancement, severe perifocal edema and no visible scolex. Besides, an irregular pattern of the lesion was seen in one of them. These imaging manifestations are more likely to be considered as metastases, brain abscess or glioma during the diagnostic process. When the solitary lesion with larger size, it is more challenging to identify NCC on imaging, especially in the case of false negative result in parasitic antibody screening, as we found in one case of this group. NCC can also be misdiagnosed with multiple lesions, when there is a big mass due to the aggregation of multiple cysts and associated with an inflammatory response and granulation. These imaging findings are similar to the features of brain tumor or other granulomatous lesions, and unlike the typical presentation of cysticerci with a scattered distribution. In addition, when multiple parenchymal NCC and metastases coexist in the same patient, it is more difficult to identify the lesion and choose proper treatment, as in the case with the coexistence of NCC and metastases we described above. It is worth mentioning that a huge temporal mass was found in 1 patient of our group. On MRI, it presented as hypointensities

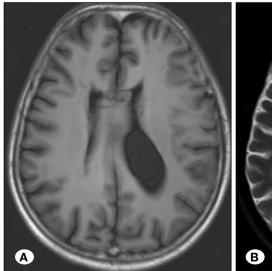






Figure 4: A 40-year-old female with intraventricular NCC. A) T1WI, B) T2WI, C) Postoperative brain CT at the 3rd month after VP shunt. A cystic lesion with long T1 and T2 signals was found in the dilated left ventricle with a thin capsule (B, Arrow). A simple VP shunt without lesion excision was performed as the initial management. 3 months later, the brain CT revealed the original cyst floating in the ventricle (C, Arrow) and aggravated hydrocephalus, suggesting the failure of the VP shunt.

on both T1WI and T2WI, unlike the signal features of most cerebral tumors with long T1 and T2 signal intensity. On CT, calcifications were also seen within the lesion. The intraoperative findings and postoperative pathology revealed abundant glial fibrosis and calcifications. Thus, we hypothesize that the huge isolated mass may have been formed by the aggregation of cysticerci and wrapped formation of adjacent glial and fibrous tissues due to the inflammatory stimuli. The value of surgical treatment of parenchymal NCC not only consists in direct removal of lesions, but also in definitive diagnosis and later proper treatment.

Subarachnoid NCC, as a severe form of the cysticercocis, has a high mortality usually because of a large mass effect, hydrocephalus or leptomeningitis (3). The suprasellar cistern is its common predilection site. The symptoms often include intracranial hypertension, visual impairment and hormone disturbance. Radiologically, these lesions are often misdiagnosed as craniopharyngioma, arachnoid cysts, or epidermoid cysts. Some scholars have proposed prolonged or high-dose anti-parasitic and anti-inflammatory drugs in treating subarachnoid NCC. However, whether simple medical therapy is effective is unclear (6). Currently, the aim of surgical treatment in subarachnoid NCC is to relieve the intracranial hypertension and hydrocephalus. According to the literature, a simple VP shunt has a high failure rate (10). Typically, subfrontal, subtemporal, and pterional approaches are used for suprasellar cysticerci resection (8,10). In our group of subarachnoid NCC, all the patients had suprasellar cysts and communicating hydrocephalus, and one was associated with perifocal arachnoiditis. Craniotomy and lesion excision were performed in all 3 patients, but a VP shunt was not preferred. After the surgery, the hydrocephalus was relieved without recurrence or serious postoperative complications in all the patients during the follow-up period. Based on the surgical results, the prognosis is optimistic. Under the premise of protecting brain function, craniotomy resection of subarachnoid NCC could directly remove the lesion and relieve the hydrocephalus on the one hand and the surgical findings are helpful in clearing the diagnosis and determining subsequent therapy on the other hand.

Intraventricular NCC can adhere to the ependymal layer or float freely in the ventricle (11). Lasting cystic obstruction of the ventricle could cause obstructive hydrocephalus, while intermittent obstruction may lead to Brun's syndrome (11). Apart from direct obstruction of the ventricular system, inflammation or fibrosis could also cause obstruction of cerebrospinal fluid pathways (10). Although it has been reported that a case of 4th ventricular cysticercosis was cured by medical therapy, the effect of medical treatment is still not ideal in general (10). A VP shunt is the most common means of treating cysticercal hydrocephalus. However, according to the literature, the failure rate of a simple VP shunt is not low (4). Some scholars believe the reason is the cyst being sucked into the ventricular catheter, or the obstruction of the drainage tube by inflammatory debris (10). In addition, another potential complication in patients undergoing VP shunt without lesion excision is ependymitis or arachnoiditis provoked by the cystic deterioration, and these may worsen the prognosis considerably (10). Thus, a substantial proportion of patients who first received a VP shunt later underwent craniotomy and lesion resection. Similarly, the hydrocephalus recurred after an initial VP shunt, and lesion resection through craniotomy had to be performed in the ventricular NCC described in our group.

The standard procedures of traditional craniotomy for intraventricular NCC are transcallosal and transcortical-transventricular approaches to the lateral and third ventricle. In recent years, endoscopic surgery has been widely used in the resection of intraventricular NCC. According to reports, the endoscopic microsurgery has a better effect and prognosis compared to the traditional craniotomy as regards shortening the operation time and avoiding complications of corpus callosum transection in open craniotomy (7,12). Moreover, septum pellucidotomy can be performed with microscopic surgery to establish communication between the lateral ventricles in case of occlusion of the foramen of Monro caused by scarring (10). Still, its long-term effect needs further observation and research.

■ CONCLUSION

The surgical treatment of NCC is still in discussion. For now, the medical therapy for parenchymal cysticercosis is still the preferred treatment plan. The aim of craniotomy is to remove the lesion, treat the medically intractable epilepsy, and establish the diagnosis and subsequent treatment. As for extraparenchymal NCC, due to its fatal complications including hydrocephalus, arachnoiditis and ependymitis, medical treatment by itself has risks and surgical treatment is a valuable alternative. For subarachnoid cysticercosis with a higher failure rate of a VP shunt by itself, open craniotomy can help directly remove the cyst and efficaciously relieve the hydrocephalus. For the patient with intraventricular NCC associated with hydrocephalus, the effect of a VP shunt by itself seems unsatisfactory, and craniotomy for cyst resection is necessary.

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