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Extraparenchymal Neurocysticercosis

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1. Introduction

Neurocysticercosis (NCC) is the most common disease causing cystic lesions in the central nervous system, especially in developing and tropical countries (1-5). Although neurocysticercosis is pleomorphic in its presentation, extraparenchymal neurocysticercosis may be challenging to diagnose and treat. With increasing globalization and international travel, NCC is now being reported from many developed countries such as the USA, UK and many European countries. Neurologists and neuroradiologists in these countries are often unaware of the pre-/post-treatment radiographic patterns of extraparenchymal NCC and the potentially poor prognosis if not correctly diagnosed and managed. Herein, we review the literature on extraparenchymal neurocysticercosis as a cause of meningitis and hydrocephalus and we discuss challenges in diagnosis and management of these cases.

2. Definition

Extraparenchymal neurocysticercosis (NCC) is defined as neurocysticercosis involving the subarachnoid, meningeal and intraventricular space (1;6;7). Thus, the clinical manifestations of extraparenchymal NCC range from asymptomatic lesions to meningitis and hydrocephalus (1;6;7).

3. Epidemiology of extraparenchymal NCC

Neurocysticercosis is the most common parasitic infestation of the central nervous system worldwide (1;4-7;11;12). Extraparenchymal NCC is frequently seen in Latin American countries whereas it is less common in the Indian subcontinent. Genetic differences in *T. solium* cysticerci have been reported from different countries (15) and may contribute towards the clinical variations across countries. These variations are perhaps due to complex interactions between the host, parasite and environmental factors (13;16-18). Cysticercal meningitis, although reported to constitute 42-48% of cases in Latin American case-series of

NC, is somewhat uncommon with <8% of cases having meningitis among adult patients (6-9;19). Extraparenchymal NCC is also of emerging importance in developed countries (11;20). This increase likely includes extraparenchymal cases, and a recent study reported an overall frequency of subarachnoid cysts in 2%, ventricular cysts in 6%, and hydrocephalus in 16% of NCC cases (11). These cases represented almost one-third of NCC in a medical center in New Mexico (21). Intraventricular NCC, the presence of *Taenia solium* cysts in the cerebral ventricular system, occurs in 7–30% of patients with NCC (10;21-23). Thus, extraparenchymal NCC is probably more frequent than previously thought (14;18).

4. Pathogenesis of extraparenchymal NCC

4.1. Subarachnoid NCC

The disease occurs when humans ingest eggs of *Taenia solium* from contaminated food (1). Brain parenchyma is most likely seeded through hematogenous dissemination and the ventricular system, subarachnoid space, and basal cisterns are then seeded via the choroid plexus (1). When cysts lodge outside the brain parenchyma (extraparenchymal NCC), they tend to grow irregularly depending on the space available and usually elicit a strong inflammatory response. Subarachnoid cysts can also grow abnormally as a membranous and/or cystic mass called racemose cysticercosis (7). Occasionally the cysts enlarge considerably, become racemose without scolices and cause mass effects. Cysticercus racemose is characterized by larger size (4–12 cm), absence of scolex and a variable appearance. Cysticercus racemosus is a form multilobular grape-like cluster without scolex thought to be a *forme fruste* of cysticercus cellulosae (24), most frequently located in the basal cisterns, Sylvian fissure, or ventricles (10;21-23). These cysts continually grow compared with intraparenchymal NCC and commonly result in basilar arachnoiditis (7) with inflammation and fibrosis in and around critical structures, causing meningeal inflammation, hydrocephalus due to CSF outflow obstruction, or cerebrovascular complications (10;21-23). The cyst often tends to migrate to the fourth ventricle because of gravity and CSF flow patterns (10;21-23). Although neurocysticerci undergo four stages of involution: vesicular, colloidal, granulovacuolar and calcific, this evolution does not occur in the intraventricular or the subarachnoid forms (or racemose type) of NCC (25).

4.2. Cysticercal meningitis

“Racemose” cysticercosis is associated with an intense inflammatory reaction and progressive thickening of the leptomeninges at the base of the brain. Signs of meningitis, cranial nerve palsy and cerebral infarcts secondary to vasculitis may also develop (26). In approximately 50–60% of the cases, there is an obstruction of the CSF circulation, resulting in progressive intracranial hypertension and hydrocephalus and mortality of over 20% of cases (26). When hydrocephalus secondary to cysticercotic meningitis is present, the mortality rate is high (50%) and most patients die within 2 years after CSF shunting(4). Therefore, basal cisternal locations are considered to be malignant forms of NCC (26).

4.3. Intraventricular NCC

Hydrocephalus can result either because of direct obstruction of cerebrospinal fluid (CSF) pathways by intraventricular cysts or secondary to inflammatory obstruction. Intraventricular neurocysticercal cysts occur singly or in multiples and frequently coexist with parenchymal and sub-arachnoid cysts (10;21-23). Intracranial hypertension is a common manifestation of extraparenchymal NCC and the increased intracranial pressure can be from the mass effect of a giant subarachnoid cyst (27), or from obstructive hydrocephalus produced by direct obstruction of the ventricular system by a cyst (28), distortion of ventricular CSF pathways (29), or blockage of CSF pathways within the subarachnoid space from the inflammatory reaction (28). However, a cyst in the fourth ventricle tends to be solitary, without accompanying parenchymal cysts (10;21-23).

4.4. Extramedullary cysticercosis

Involvement of the spinal cord is rare, accounting for 1–5% of all cases of NCC (30;31). Spinal NCC may be intramedullary or extramedullary (intradural or extradural); however these forms are rare, and most disease involves the subarachnoid spaces, which may result from direct CSF dissemination (30;31). It has been suggested that small, or developing larvae present in the subarachnoid space settle to the basal cisterns by way of gravity and then further descend into the subarachnoid spaces of the lower parts of the spine including the lumbosacral space where they find adequate room to grow and develop (30;31). Many of the pathologic descriptions of cysticerci recovered from the spinal canal have a racemose morphology, which consists of membranous cells that proliferate, which may cause seeding of the spine from the base of the brain and subsequent growth (30;31). Cervical involvement consists mostly of unilocular or multilocular cystic forms, primarily due to direct extension of cysts located in the basal subarachnoid cisterns (30;31). Conversely, lumbosacral involvement has a more varied picture including multilocular cystic lesions causing spinal displacement or flattening, and clumping or displacement of the nerve roots most likely due to adhesive arachnoiditis (30;31). Clinical signs can be caused by direct compression of the spinal cord and/or roots by cysticerci or, indirectly, by the inflammatory reaction, including progressive paraparesis and sphincter disturbances.

4.5. Orbital neurocysticercosis

Ocular cysticercosis is caused by the growth of the larvae of *Taenia solium* within the ocular tissues and the cysts may be located in descending order of frequency, subretinal space (35%), vitreous (22%), conjunctiva (22%), anterior segment (5%) and orbit (1%)(32). The extraocular muscle form is the most common type of orbital cysticercosis (33-35). In the ocular form, the most favored sites are the vitreous and the subretinal space whereas the inferior rectus and the medial rectus are the most common extraocular muscles involved (33-35). Intraocular cysticercosis is predominant in the Western countries, whereas extraocular is more common in the Indian population and several authors have attributed geographic and environmental factors to this difference (33-35). The continuation of the

retina with the optic nerve allows direct communication of the subarachnoid space with subretinal space but it remains unclear whether subretinal NCC can be considered another form of subarachnoid NCC (33-35).

There is lack of understanding of the pathophysiology of extraparenchymal NCC. Circulating antigen detection assays could help establish if living parasites are still present after apparent radiographic cyst regression, providing an indication to continue or reuse antiparasitic treatment (36). It is unclear whether chronic inflammation in extraparenchymal NCC is due to continuous cyst degeneration, or continuous antigen release from dead parasitic tissues (37). Newer mechanisms are being explored to understand the complex neuropathology and heterogeneity of NCC.

5. Clinical manifestations of extraparenchymal NCC

The clinical and radiologic manifestations of NCC are pleomorphic. Epilepsy, present in both intraparenchymal and extraparenchymal NCC, focal neurological signs, and headache are the most common clinical manifestations of the disease (1;38). Focal neurological signs that vary according to the size, number and location of the parasites have been described in up to 20% patients with neurocysticercosis (1;38). Pyramidal tract signs predominate, but sensory deficits, involuntary movements, and signs of brainstem dysfunction, may occur in some patients (1;38). These manifestations usually follow a subacute or chronic course resembling that of a brain tumor and are most often seen in patients with large subarachnoid cysts compressing the brain parenchyma (1;38). Thus, several varieties of NCC have been recognized depending upon the number, location, and evolutionary stage of the cysticerci in the human brain (39).

Extraparenchymal NCC occurs mainly in young adult males and is uncommon in children (38;40). The patients typically present with subacute or chronic intracranial hypertension from mass effect or hydrocephalus; chronic meningitis characterized by lack of meningeal signs of exam, often due to chronicity of symptoms, a mild-to-moderate CSF lymphocytic pleocytosis and moderate-high increase in protein; radiographic hydrocephalus with or without obvious cysts; radiographic presence of cysts in the ventricles or subarachnoid space and protracted clinical or radiographic course after antiparasitic treatment (8;21). Stroke can occur secondary to vasculitis caused by inflammatory occlusion of the arteries at the base of the brain secondary to arachnoiditis (4;5;12).

Hydrocephalus develops in approximately 30% of all patients with NCC because of obstruction by intraventricular or subarachnoid lesions (10;21-23). Intraventricular NCC can cause non-communicating hydrocephalus by obstructing the CSF pathway and communicating hydrocephalus by development of ependymitis (10;21-23). Abrupt permanent obstruction can cause sudden death due to brain herniation (10;21-23). Life-threatening acute intermittent hydrocephalus (Brunn syndrome)(41) can occur due to cyst-inducing intermittent CSF obstruction from a ball-valve mechanism (10;22). Overall, extraparenchymal NCC has a more aggressive behavior and a higher morbidity and mortality rate than parenchymal form (10;21-23). Presence of sub-arachnoid cysts can

cause chronic cysticercal meningitis. Cysticercal meningitis (CM) is characterized by inflammatory cerebrospinal fluid (CSF) and negative bacterial and fungal cultures (8). There have been no systematic studies of CM. In a recent study of patients with CM these patients often had intracranial hypertension, meningeal signs, CSF hypoglycorrachia, positive CSF results in an enzyme-linked immunosorbent assay (ELISA) for cysticercal antigens, negative CSF cultures for bacteria, fungi, and mycobacteria and longer clinical course of NCC (8). The management of the chronic inflammation and the complications caused by this meningitis are usually very difficult, and the mortality rate can be up to 33% (10;21-23). It is likely that CM is often not identified and its correct identification may reduce morbidity and risks of unnecessary surgery in patients with chronic neurocysticercosis and CSF shunts (10;21-23).

6. Diagnosis of extraparenchymal NCC

The diagnosis of NCC is often made based on presence of lesion highly suggestive of neurocysticercosis on neuroimaging study, positive serum immunoassay for the detection of anticysticercal antibodies, positive CSF immunoassay for detection of anticysticercal antibodies and epidemiologic criteria including individual coming from an area where cysticercosis is endemic (1). These diagnostic criteria have been stratified in four categories—absolute, major, minor, and epidemiological—on the basis of their individual diagnostic strength (1). Based on a previous consensus, the absolute criterion for the diagnosis of neurocysticercosis that is being considered as pathognomonic of this disease is the detection of a scolex inside a cyst by CT or MRI although, *C. racemosus* doesn't have a scolex (1).

6.1. Clinical presentation

Extraparenchymal NCC is associated with a local inflammatory response with high protein concentration and cell counts in the CSF (8). Clinical manifestations and CSF findings are similar to the more common tuberculous meningitis, (8;23) and other forms of chronic meningitis including chronic HIV-associated meningitis (42;43) since the CSF findings consist of pleocytosis (usually lymphocytic but frequently polymorphonuclear), reduced glucose and elevated protein (8). In one series of cysticercal meningoencephalitis, confusion with tubercular meningitis was present in 61.5% cases (7;8). An important differentiating feature with these other forms of chronic meningitis is the presence of eosinophils in the CSF (7;8) which is usually seen only in the initial phases of the illness (8;21). However, this staining of the CSF is not routinely done in most places (8;23). Although the suspicion of NCC as the cause of chronic meningitis is increased when CSF eosinophils are found, CSF eosinophils (above 5%) occur in only 15% of patients (21). It is often a common practice to attribute chronic meningitis and hydrocephalus to tubercular meningitis in the presence of appropriate epidemiologic history and treat empirically by shunting and anti-tubercular therapy. Thus, an astute clinical acumen is required to make the diagnosis of CM.

6.2. Imaging

Both computed tomography (CT) and magnetic resonance imaging (MRI) are indispensable tools for diagnosis and characterization of NCC (38;44;45); CT is superior for diagnosis of racemose cysticercosis, brain granulomas and calcifications, which are the most frequent finding of NCC, and may be missed by MRI (38;44;45). However, neuroimaging findings of extraparenchymal cysticerci are subtle and are usually not seen by CT. Thus, MRI is more useful than CT for diagnosis of ocular, ventricular, and subarachnoid cysticercosis and for analysis of the inflammatory reaction that accompanies most cases of active NCC (38;44;45).

Neuroimaging findings are variable depending on the stage of the infection. The first stage, described as the larval tissue invasion phase, is not normally imaged owing to lack of symptoms at this very early stage (1;46). During the vesicular stage, cysts and scolex, the "mouth" of the tapeworm that is lined with suckers and hooks, are both imaged without enhancement. However, as observed in one study(47) in which imaging was performed regularly to follow anticysticercal therapy, this phase appears as a localized focus of edema on T2-weighted images and displays nodular tissue enhancement following the administration of gadopentetate dimeglumine (1;46).

The second stage (the vesicular stage) describes the formation of a cyst that encircles the scolex (1;46). These cysts are thin walled, contain clear fluid and are typically 1-2 cm. On imaging, the cyst fluid parallels cerebrospinal fluid intensity. The scolex is approximately 2–4 mm and appears as a mural nodule that is isointense with brain parenchyma (48). The lesion is antigenically inert and therefore does not induce an inflammatory reaction or circumferential edema (49).

During the third stage (the colloidal stage) the parasite dies, and as a result the cysticercus becomes nonviable (1;46). As the scolex dies, the cyst fluid transforms into a colloidal suspension containing protein solutes (1;46) and on MRI imaging this results in T1 shortening while the scolex and cyst capsule are decreased in T2 signal intensity. The surrounding edema suggests that the parasite is in its colloidal state, and, therefore, enhancement of the capsule and scolex will occur avidly. In addition, the proteinaceous nature of the cyst fluid during involution of the cysticercus is appears as hypointense central T1 signal to white matter but hyperintense to cerebrospinal fluid and appears markedly hyperintense on T2-weighted images (1;46). At CT, cystic contents increase in attenuation (50). Thus, in the colloidal vesicular stage, ring enhancement and edema are appreciated by both CT and MRI imagings (1;46).

The fourth stage (the nodular granular stage) represents the degeneration of the cysticercus. The edema begins to subside gradually, the contents begin to mineralize and the cyst involutes. Thus, the lesion becomes isointense with brain parenchyma on T1-weighted MR images and hypointense on T2-weighted MR images. At CT, a thick nodular ring continues to enhance and the lesion becomes isoattenuating (51).

The final stage is the calcified stage, which describes complete involution of the lesion with continued mineralization. The calcifications are obvious at CT as small areas of

hypointensity (1) and show susceptibility at MR imaging, particularly on gradient-echo images (47). However MRI is not the imaging modality of choice for calcified NCC (47). Also, in the nodular calcified stage, when the cystic lesion is mineralized and shrunken and a nonenhanced CT scan is diagnostic, enhancement is unusual (51). The granular nodular phase is characterized by decreased ring enhancement and edema, along with the calcification of cysts (46).

Subarachnoid (racemose) neurocysticercosis usually infiltrates the basal cisterns and sylvian fissure and has different imaging findings compared to parenchymal NCC. Common neuroimaging findings to suspect the diagnosis include hydrocephalus (with or without obvious cysts), cysts obstructing CSF pathways or freely floating inside ventricles, cysts in the basal subarachnoid cisterns, migrating cysts across the cerebral aqueduct, and ependymitis or arachnoiditis (21). The most common CT finding in subarachnoid NCC is hydrocephalus (1;9). Because the cyst membrane is thin and the fluid is isodense with the cerebrospinal fluid, uninflamed extraparenchymal cysticerci are usually not visible on computed tomography scanning and may only reveal subtle, indirect findings on MRI (9). Therefore, neuroimaging may reveal hydrocephalus without noticeable cysts (1;9).

6.3. Laboratory tests for diagnosis of NCC

Taenia antibodies detected by methods such as Western Blot is considered as a major criterion whereas the positive serologic test in the CSF is listed as a minor criterion (19). The development of numerous serodiagnostic tests using different parasitic antigens is indicative of the fact that none of them are 100% sensitive and specific. For multiple lesions, the enzyme-linked immunoelectrotransfer blot (EITB) assay using purified glycoprotein antigens from *T. solium* cysticerci was reported to be highly specific (100%) and nearly 98% sensitive (4;5;12). The sensitivity was less for single lesions (52) and for calcified lesions (2;2;53-56). A comparative study of enzyme-linked immunosorbent assay (ELISA) and dot-blot assay in children found that both were more sensitive in cases with multiple brain lesions (100%) than in those with a single lesion (87%) (5). The sensitivity of antibody detecting EITB assays is not better with the use of CSF samples as compared with serum samples (55). Detection of circulating cysticercosis antigens using ELISA has a modest sensitivity especially for parenchymal lesions. The antigen detecting ELISA has a better sensitivity with the use of CSF samples as compared with serum samples. However, ELISA has less sensitivity as compared with EITB for serum as well as CSF samples, for both intraparenchymal and extraparenchymal NCC (55;57-59).

It has been suggested that the use of excretory secretory (ES) antigens, rather than somatic antigens might improve the serodiagnosis of cases with vesicular stages of the parasite (60;61). The use of ES antigens for the detection of antibodies in serum was found to be more useful than that in urine in patients with enhancing lesions (62). Three ES peptides were reported to have high sensitivity and specificity in both serum and CSF reactivity; (60;61). It has been suggested that synthetic peptide selected by phage display may be useful in the immunodiagnosis of NCC (63;64).

Polymerase chain reaction (PCR) in CSF has also been used for the diagnosis of neurocysticercosis but is not widely available (19). However, extensive and comprehensive revision of the diagnostic criteria of neurocysticercosis, especially of extraparenchymal neurocysticercosis is mandatory according to many recent publications (2;8-11).

7. Treatment

There is still no consensus regarding optimal treatment strategies in patients with extraparenchymal NCC (10;21-23). Various therapeutic modalities include antihelminthic medication, microneurosurgical removal, ventriculoperitoneal shunting, and endoscopic management (8).

7.1. Medical therapy

Although parenchymal cysts have historically been treated quite effectively with antihelminthics such as praziquantel and albendazole, medical therapy alone is not favored for extraparenchymal NCC because of the limited efficacy in such cases, and a risk of developing acute hydrocephalus during the clinical treatment period (10;21-23). Good results for antiparasitic treatment with different albendazole and praziquantel regimens for extraparenchymal NCC including orbital, spinal, intraventricular and subarachnoid NCC, and even for giant cysts have been reported (57;65-69) although resistance has been reported by some (13;16;17). However, although treatment with antihelminthic medication such as albendazole has been shown to improve outcome in live, cystic parenchymal cysticercosis, the benefits of antihelminthic treatment in patients with solitary cystic lesion remain uncertain (70). While it is generally accepted that both praziquantel and albendazole are effective in destroying viable cysts, their use in cases with enhancing lesions has been debated as these lesions are considered to represent degenerating cysts, many of which resolve spontaneously (4;5;12). Thus, the decision whether antiparasitic treatment should be used in these cases is always a clinical decision and should be made on an individual basis.

Antihelminthic agents hasten the evolution of intraventricular viable cysts, which may trigger an inflammatory response similar to that seen with the natural history of the parasite (25). This may result in long-term sequelae (1;6;9;71). Extraparenchymal cysts may regress only after long term and multiple antiparasitic courses (1;6;9;71). The optimal treatment to prevent chronic inflammation is unknown due the lack of understanding of its pathophysiology and lack of controlled trials to help guide management. Likewise, controversy exists regarding the use of corticosteroids, alone or in combination with antihelminthic drugs (1;6;70;71). At a previous consensus meeting, experts agreed that no single treatment approach could be advocated and that management options varied according to the type of clinical presentation (9). However, intraventricular neurocysticercosis has a risk of ependymitis in those treated with antihelminthics such as albendazole and praziquantel regimens. Thus these agents should be used with caution in cases with extraparenchymal NCC as any increase in the inflammatory response may lead to the development of infarct; pretreatment with steroids and management of intracranial

hypertension is warranted and surgical evaluation is often needed prior to medical treatment (8).

7.2. Surgical therapy

In patients presenting with acute hydrocephalus, surgery is the only option (10;21-23). Neurosurgical procedures for NCC are still part of the armamentarium when treating this disease and good results for open craniotomy and rigid endoscopic surgery in patients with intraventricular and subarachnoid NCC have been reported (72;73). Infratentorial intraventricular cysts have been treated with open surgery for excision whereas it is generally suggested that supratentorial cysts, due to not only location but also the need to often treat hydrocephalus in these patients, be removed endoscopically (74). Ependymitis, confirmed by neuroimaging is a relative contraindication for surgical removal of the cysts (10;21-23).

Ventriculoperitoneal CSF shunting is burdened by a high shunt dysfunction rate which leads to worsening obstruction of CSF flow and increased intracranial pressure, risk of infection and thus high mortality rates. Microneurosurgical approaches can be technically demanding and associated with various complications (10;21-23). For these reasons, endoscopic approaches for intraventricular neurocysticercosis have been described in recent years and often allow for cyst removal and hydrocephalus treatment, freeing the patient from shunt procedures (75;76).

Although the literature regarding the use of endoscopic management of intraventricular NCC is scarce, this modality has shown encouraging results in the treatment of intraventricular NCC (10;21-23). In a recent comparative study of 140 patients from Mexico with intraventricular NCC, traditional treatment with albendazole and steroid had similar outcome versus neuroendoscopic surgery in terms of survival, hospitalization (23). However, almost all patients with traditional treatment remained with at least one shunt whereas most of the patients from the neuroendoscopic surgery series did not have any shunts (23). Thus, the neuroendoscopic approach to intraventricular neurocysticercal cysts is safe and effective and offers the additional benefit of avoiding shunt placement (10;21-23). At centres having the required expertise, this should be the treatment of choice. Traditional treatment is a second option where the endoscopic procedure is not available. However, endoscopic cyst excision can be difficult and hazardous in patients with severe ependymitis and dense adhesions and intraventricular bleeding could also report (10;21-23). Thus, despite its many advantages, neuroendoscopy has some limitations even when performed by experienced hands.

Regarding subarachnoid NCC, there are no controlled trials on the management of this form of extraparenchymal NCC (1;6;71). In a series of patients treated with only CSF diversion, 50% died at a median follow-up of 8 years and 11 months (2). More recently, case series using anti-parasitic drugs, corticosteroids, and shunting for hydrocephalus have been associated with an improved prognosis compared with older studies (1;2;6;71). Thus, most experts consider subarachnoid NCC a clear indication for anti-parasitic therapy (2).

However, the optimal dose and duration of anti-parasitic therapy for subarachnoid cysticercosis has not been established (2). In the largest cases series, Proaño and others treated 33 patients with giant cysticerci with albendazole (15 mg/kg/day) for 4 weeks and most patients required several courses of anti-parasitic therapy (23). However, with controversy in the literature in the optimal management of this condition and without further evidence-based guidelines to help management of extraparenchymal NCC, the decision of the total dose and duration of antiparasitic and steroid therapy must be made on a case by case basis.

8. Conclusion

Extraparenchymal NCC may be a more common form of NCC than previously thought and is often difficult to diagnose, more complex to treat, and carries a graver prognosis. The clinical course is protracted and difficult to cure. Different medical (anthelmintics, steroids), surgical (cyst excision, CSF diversion), or medical-surgical approaches have been reported but not adequately studied. Because clinicians in developed countries often unfamiliar with NCC as a cause of chronic meningitis, chronic ventriculitis, or hydrocephalus without obvious cysts, the diagnosis of extraparenchymal NCC often depends on the correct interpretation of neuroimaging which may miss the diagnosis. Thus, extraparenchymal NCC should always be considered by clinicians and radiologists in the differential diagnosis of chronic meningitis and hydrocephalus.

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