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A CASE OF EXTENSIVE NEUROCYSTICERCOSIS WITH DISSEMINATION

Medicine

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ABSTRACT

India reports a large number of cases of cysticercosis, with hygiene and sanitation being the likely influencing factor. Central nervous system (CNS) involvement with Taenia solium cysts is known as neurocysticercosis (NCC). Clinical presentation of NCC is determined by the cyst location, stage, size, number and associated oedema with seizure being the most common presentation. Diagnosis requires identification of parasites in tissues or radiological demonstration of scolex in cystic lesions. In CNS, multiple cysts are the rule. CT or MRI is the mainstay of radiological diagnosis. MRI is better for detection of degenerating and innocuous (viable) cysticerci, while CT being better for calcified lesions. Serological tests tend to perform better in patients with high cyst load (sensitivity>94%) as compared to patients with calcified cysts or solitary cyst (as low as 28%). Recently developed tests Lentil Lectin Glyco-Proteins Western Blot (LLGP-WB) and Enzyme-linked Immuno-electro Transfer Blot (EITB), have high sensitivity and specificity. Albendazole is the drug of choice for cysticidal therapy. Antiepileptics and shunting surgeries for symptomatic hydrocephalus is warranted before, during and after drug therapy. Antiparasitic therapy is contraindicated as initial therapy in disseminated neurocysticercosis, ocular disease or patients with features of increased ICT. Steroids play a key role in combatting inflammation. Neuro-endoscopic removal is the procedure of choice for intraventricular cysts. Hydrocephalus requires ventriculo-peritoneal shunting. Although management includes both surgical and medical modalities, it needs to be individualized to each case.

KEYWORDS

Neurocysticercosis, MRI, Cestode, seizure

INTRODUCTION

Neurocysticercosis (NCC) is the term used for central nervous system (CNS) involvement with *Taenia solium* cysts (1). *T. solium,* also known as the pork tapeworm with its extensive geographic distribution has an enormous impact on human health with more than 50,000 deaths per year attributable to NCC (2).

The parasite requires two hosts, man, the only definitive host and pigs the intermediate host which get infected with cysticerci. Unfortunately, humans can also be readily infected with eggs of *T.solium* which is the cause of all the significant morbidity associated with this parasite.

Clinical manifestations of NCC vary with the size, location of cysts and the degree of the host's immune response. Seizure is the most common presentation (70 – 90%) which usually responds well to antiepileptic drugs. Other clinical manifestations include headache, features of increased intracranial tension due to parenchymal oedema/ hydrocephalus, chronic meningitis, focal neurological deficits, and psychological disorders.

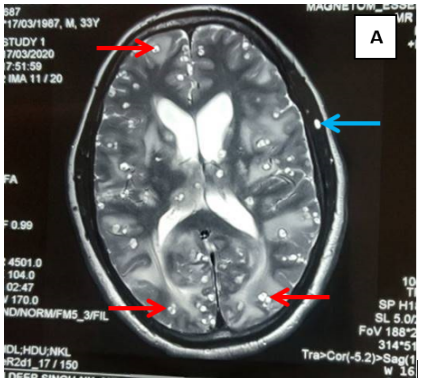
Site of involvement and the symptoms experienced dictate the modalities of diagnosis and treatment. Definitive diagnosis is made by identification of parasites in tissues or by a radiological demonstration of the scolex in cystic lesions.

Management requires antiparasitic therapy, steroids and/or shunt/ surgical intervention decided as per need of the case.

CASE REPORT

A 36 year old male with no known comorbidities was brought in the emergency department with history of recurrent episodes of generalised tonic clonic seizures of half hour duration with transient regain of consciousness in between episodes during which he was confused and behaved abnormally. He had well-formed visual hallucinations and even did not recognise his family members. Detailed history from the next of kin also revealed history of progressive dull headache with no specific characteristics for last 3 months. There were intermittent episodes of transient abnormal behaviour during which he was at times withdrawn and had visual and auditory hallucinations. He became aggressive at other times and had delusions of persecution. Over the last 01 month he had 3 episodes of generalised seizure which were managed symptomatically at a local clinic and was not further evaluated. Relatives also noticed a gradual but distinct change in the personality of the individual. On admission the patient was confused and delirious. Vitals were normal. Pupils were bilateral equal in size and normally reacted to light. Right plantar response was mute and left was flexor. Remaining systemic examination along with hematological and biochemical parameters were essentially normal. MRI Brain revealed multiple round to oval well defined cystic lesions with T1 hyperintense and T2 hypointense focus suggestive of scolices, averaging 5mm and seen diffusely involving all lobes of both cerebral and cerebellar hemispheres with surrounding oedema showing T2 and FLAIR hyperintensities to suggest colloid vesicular stage. Similar findings were also seen in bilateral temporalis, left masseter, tongue, digastric, posterior neck muscles and parotid gland, suggestive of extensive neurocysticercosis with dissemination, however without intraventricular and ocular involvement, raised intra cranial tension or hydrocephalus.

ECG, chest X ray and 2D Echocardiography was normal. In view of high cyst burden, high dose steroid therapy was initially started (dexamethasone 8 mg/day and was given for 28 days with gradual tapering), Tab Albendazole 400 mg twice a day was also administered for 14 days. Anti-epileptics were started and dose was optimised. Patient responded well to therapy and no complications occurred. There was no recurrence of seizures, and psychiatric symptoms improved too. No surgical intervention was required.



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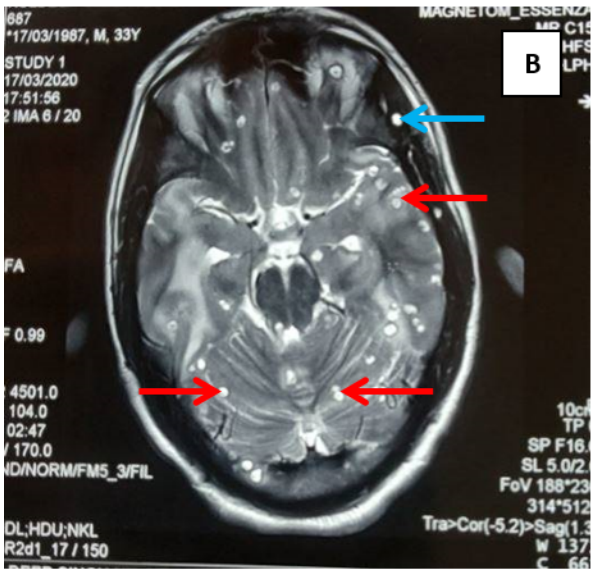

Figure 1: MRI Brain: T2W axial images (A&B) showing extensive involvement of entire brain parenchyma by multiple small cystic lesions with hypointense eccentric focus with surrounding perilesional edema; Frontal, occipital lobe (Red arrows-A) and scalp involvement (Blue arrow-A) & Temporal lobe and cerebellar involvement (Red arrows-B) & scalp (Blue arrow-B).

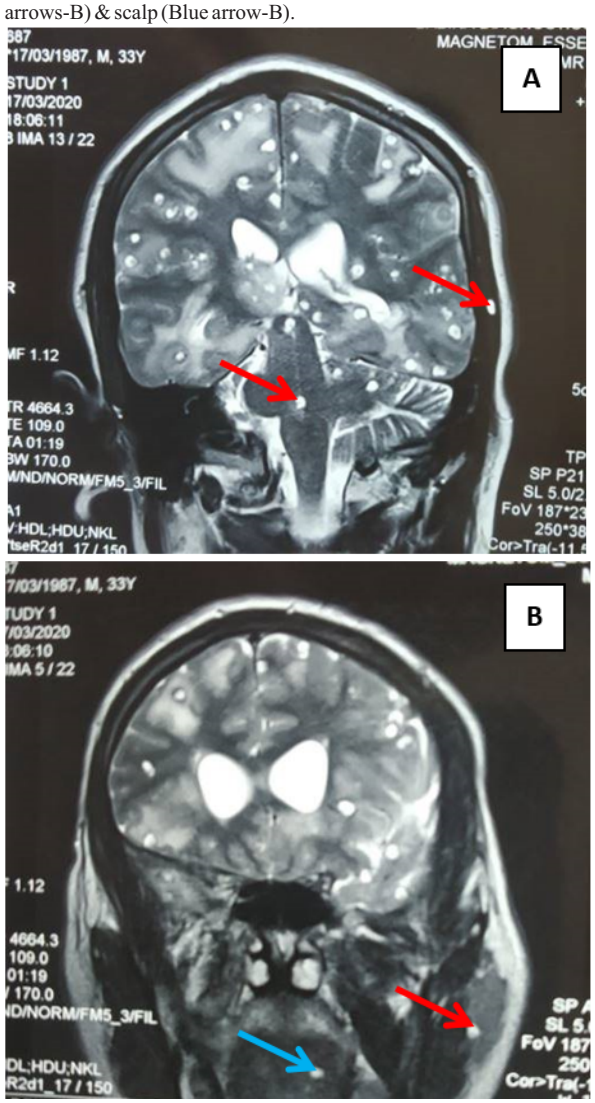

Figure 2: T2W coronal images (A) showing cystic lesions with eccentric nodules and perilesional edema involving brain stem & scalp (Red arrows-A) and tongue (Blue arrow-B) and left parotid gland (Red arrow-B).

DISCUSSION

Tapeworms or cestodes are an ancient class of highly specialized flatworm parasites. Acquired by ingestion of eggs excreted in the faeces, individuals harbouring an adult *T.solium* are at higher risk, probably through faecal–oral autoinfection. Internal autoinfection has also been hypothesised by unhygienic practices or by reverse peristalsis. Infected subjects normally have multiple cysts all over the body. Symptoms may develop because of local inflammation; with neurologic symptoms being most prominent. Any part of the CNS may be involved, but symptomatic disease is often attributable to intracerebral, intraventricular, subarachnoid and spinal cord cysts. Death of intraparenchymal cerebral cysts results in swelling and leaking of the antigenic material which provokes a severe inflammatory response contributing to symptoms of focal or generalized seizures, sensorimotor deficits, intellectual impairment, psychiatric disorders and symptoms of hydrocephalus. Because of their critical location, intraventricular cysts and basilar cysts tend to cause early symptoms pertaining to obstruction of CSF flow or local meningeal irritation (1).

Aggressive forms like *racemose cysticercosis* and cysticercotic encephalitis have been described in which base of the brain and significant brain parenchyma is involved. Intraparenchymal spinal cord lesions cause early symptoms because of direct local pressure effects. In the CNS, multiple cysticerci are the rule. Cysts outside the CNS are usually asymptomatic. Eventually they die and calcify, to be detected incidentally on plain radiographs. Cardiac cysticercosis is a rarely reported entity (3).

Travel to or residence in an endemic area should raise suspicion of NCC. Often confused with a tumour, imaging studies (CT or MRI) are the mainstay of diagnosis, which show multiple enhancing and non-enhancing unilocular cysts mainly at grey-white matter junction and basal ganglia (1). MRI is considered the best neuroimaging tool for the detection of degenerating and innocuous (viable) cysticerci, while CT is better for calcified lesions (7). MRI also helps differentiate the stages of the cyst, which CT fails to do. On MRI, the vesicular cysts show signal properties similar to cerebrospinal fluid (CSF) in both T1- and T2-weighted images. Scolex is visualized within the cyst as a high-intensity nodule. Colloidal stage, which represents dying cyst, appears as a ring-enhancing lesion with surrounding white matter oedema. In the next stage, which is the nodular granular stage, lesions are homogenously enhancing and they finally calcify (7).

CSF examination might reveal lymphocytic or eosinophilic pleocytosis, reduced glucose and elevated protein levels. Tests like Lentil Lectin Glyco-Proteins Western Blot (LLGP-WB) have high sensitivity (>90%) and specificity (100%) (4). Serology tends to perform better for patients with high cyst load (sensitivity>94%) but reduces drastically for patients with calcified cysts or solitary cyst (as low as 28%) (6). Enzyme-linked Immuno-electro Transfer Blot (EITB), which has 100% sensitivity and an overall 98% specificity is used currently in many centres (7). A revised diagnostic criterion helps to standardize the diagnosis in different settings (5). IDSA now recommends anthelmintic and corticosteroid therapy for multicystic parenchymal lesions (6).

Regimens for multicystic parenchymal disease include albendazole as a first-line agent (10 to 15 mg/kg/day divided in two doses for 8 days to 2 weeks) (6). Praziquantel in high doses (50 to 100 mg/kg/day divided in three doses per day for 15 to 30 days) alone or in combination is an alternative (6). CSF penetration causes destruction of subarachnoid and ventricular cysts making albendazole preferable to praziquantel. Seizure control with antiepileptic medications and treatment of symptomatic hydrocephalus by shunting is warranted before, during and after drug therapy as symptoms increase consequent to cyst death provoking local inflammation. The optimal dose of corticosteroids to combat CNS inflammation in cases with heavy cyst load is uncertain and is generally given with tapering over 6 to 8 weeks to avoid rebound symptoms (6). Steroids are warranted for cysticercotic encephalitis also.

Inflammatory response to drug therapy causing disastrous inflammatory response makes anthelminthic treatment contradictory in encephalitic, disseminated and ocular neurocysticercosis (6).

Antiparasitic therapy is never the main stay of treatment especially in a setting of increased ICP. Endoscopic surgery is the procedure of choice for intraventricular cysts. Hydrocephalus requires management with ventriculoperitoneal shunt. Frequent blockage by inflammatory exudates or small cysts necessitates multiple revisions of shunts.

Limited data is available on the role of anthelmintic agents in "single enhancing lesions (SEL)" (6) Anthelmintic agents have no role in the treatment of calcified cysts (6). Treatment failure owing to lower drug levels is likely in intraventricular or cisternal cysts and racemose neurocysticercosis often necessitating a protracted duration of cysticidal treatment with multiple courses (6). Surgical excision is recommended for intraventricular cysts with neuroendoscopic removal being the preferred technique (6). Tailor made therapy, encompassing surgical and medical approach is recommended in difficult to treat cases (6). For symptomatic cysts outside the CNS, surgical resection stays the optimal approach, however lesions involving critical organs and deep seated lesions, make it technically challenging.

CONCLUSION

Disseminated cysticercosis, although not very common, has been chiefly reported from the Indian subcontinent and is related to poor hygiene and sanitation (8). Cyst location, number, size and surrounding inflammatory reaction direct the clinical presentation. Seizure is the commonest presenting symptom. Definitive diagnosis requires identification of parasites in the tissues or radiological demonstration of the scolex in the cystic lesions. MRI is considered the best imaging modality for viable cysts. Serological tests are good for diagnosis in patients with high cyst load. Albendazole is the drug of choice with praziquantel being a good alternative. Antiparasitic therapy is not recommended as first line therapy for disseminated neurocysticercosis, ocular disease or in patients with increased ICT. Steroids are used to prevent inflammatory reaction. Holistic management includes both medical and surgical modalities which are then individualized to each case.

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