Symptomatic treatment

There is no globally accepted protocol to treat SC. Agents that affect the neurotransmitters dopamine and GABA are used as symptomatic treatment. Dopamine receptor antagonists include haloperidol, which is an effective symptomatic medication but which must be titrated slowly to reach maximum effect with minimal toxic manifestations.

In South Africa haloperidol is first-line therapy as it is cost effective and readily available. Use of haloperidol should be based on a regimen of 'start low and go slow'; 0.025 mg/kg/day in divided doses going up to a maximum of 0.05 mg/kg/day in divided doses.[12]

GABA is a neurotransmitter which inhibits dopaminergic over-activity.[8] Sodium valproate enhances the action on GABA; hence it is sometimes used to treat chorea.[8] Recommended doses vary between 15 mg/kg/day to 20 mg/kg/day in 2 or 3 divided doses. Sodium valproate has been recommended as treatment where trials with haloperidol have failed and the chorea remains severe.[11] Phenobarbitone is not recommended for symptomatic treatment.

There are detailed descriptions of the psychiatric manifestations in SC but no reports of treatments specifically targeting these symptoms.

Therapeutic interventions: immunological treatments

Antibodies reactive with neuronal tissue in the serum of patients with SC demonstrate that the condition is a humorally mediated autoimmune condition.[10] Imunomodulatory therapies are described using corticosteroids, intravenous immunoglobulins (IVIG) and plasma exchange.[14] Controlled studies are needed to definitively clarify efficacy of these interventions.

Supportive measures

Management of SC requires a multi-disciplinary approach. Management of co-morbid psychopathologies is important. Supportive psychotherapy and family therapy are recommended. Regular attendance should be encouraged for follow-up assessments and to receive benzathine benzylpenicillin injections. Helping educators to understand the condition will promote tolerance and understanding and help them to ensure that children with SC are exposed to minimal teasing and bullying.

Every child with SC must be notified and must undergo echocardiography.

Conclusion

SC has a clinical spectrum from severe chorea with mild psychiatric symptoms to minimal chorea with severe psychiatric symptoms. It is better to refer to SC as a post-streptococcal, autoimmune, neuropsychiatric movement disorder, which encompasses all aspects of the disease.

As SC is synonymous with acute rheumatic fever it must always be notified. The main goal of treatment should be primary prevention, which requires eradication of poverty and poor living conditions. Until this millennium goal is achieved strategies should target management which reduces the burden of disease in children living with SC.

References available at www.cmej.org.za

Approach to a single granuloma on CT scan

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The increasing availability and use of neuro-imaging in children with new-onset partial seizures has resulted in increased identification of intracranial granulomas.

Neurocysticercosis is the most common parasitic infection of the central nervous system and a major cause of seizures in resource-poor countries.[1] Humans develop neurocysticercosis when they accidentally ingest eggs of the pork tapeworm. It is not related to eating undercooked pork and therefore may occur in vegetarians and persons of religions that prohibit the eating of pork. In South Africa, neurocysticercosis is the most common cause of granulomas detected by computed tomography (CT) scanning. The second most common cause of intracranial granuloma is tuberculoma, which may occur in isolation or in association with tuberculous meningitis. Lymphoma and toxoplasmosis should be considered as possible causes for intracranial ring-enhancing lesions in the immune-compromised child.

Radiological differentiation between a tuberculoma and a single cysticercal granuloma is difficult.[2] Correct diagnosis is important because misdiagnosis of a tuberculoma in a patient with neurocysticercosis can result in 6 months' unnecessary high-dose antituberculosis therapy. Similarly, not identifying tuberculosis in a patient could be disastrous. We present an approach to the diagnosis and management of a single granuloma.

Clinical features

Clinically, children with neurocysticercosis often present with seizures. Headaches, altered level of consciousness, focal neurological signs and cerebellar signs present less frequently. If a tuberculous origin is suspected, the following clinical information is informative:

- positive tuberculosis contact
- features of systemic tuberculosis, including failure to thrive, night sweats and coughing for longer than 2 weeks
- focal neurological signs.

Exclusion of active tuberculosis is a priority and has the most impact on patient outcome and management.

Diagnosis

The radiological appearance of neurocysticercosis may range from non-enhancing cysts (viable cyst with no host immune response) to enhancing cysts or granulomas (degenerating cyst with surrounding immune response) to a calcified lesion (dead cyst with total resolution).[3] Contrast CT imaging is adequate in most cases of suspected neurocysticercosis. CT is superior for the detection of calcified granulomas, while magnetic resonance imaging (MRI) is better for visualising parasitic cysts. In a minority of cases, a scolex (small, eccentric internal speck of enhancement or calcification within the cyst) may be visualised. This finding is pathognomonic of neurocysticercosis (Fig. 1).

The second most common CT-enhancing lesion is a tuberculoma (Fig. 2). Imaging features favouring a diagnosis of tuberculoma rather than parasitic granuloma include:

- size >2 cm
- lobulated, irregular shape
- presence of marked oedema
- persistent, focal neurological deficit.

Because of the difficulty of differentiating between a tuberculoma and cysticercus

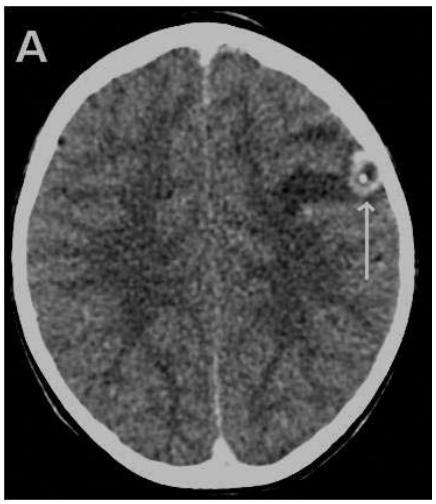

Fig. 1. A CT scan showing neurocysticercosis with a calcified nodule representing a scolex, and surrounding oedema.

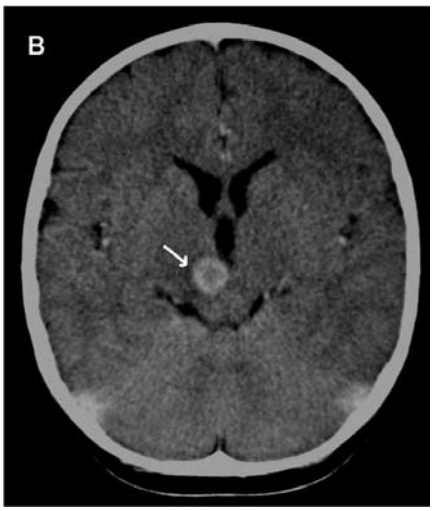

Fig. 2. Tuberculoma in the right midbrain showing ring enhancement after contrast administration.

granuloma, most radiologists do not opt for a specific diagnosis. Depending on clinical findings, the differential diagnosis includes glioma, brain abscess and toxoplasmosis.

Investigations supporting a tuberculous aetiology include a positive tuberculin skin test, a chest radiograph suggestive of pulmonary tuberculosis and a cerebrospinal fluid (CSF) picture characteristic of tuberculous meningitis. Identification of acid-fast bacilli and culture of *Mycobacterium tuberculosis* from gastric washings and/or CSF specimens confirm a tuberculous aetiology. Serological tests are positive in only a small number of cases of neurocysticercosis and are generally not helpful in the diagnosis.

Management

If a diagnosis of tuberculoma is made, the treatment is 6 months of high-dose anti-tuberculosis treatment, i.e. rifampicin 20 mg/kg/day, isoniazid 20 mg/kg/day, ethionamide 20 mg/kg/day and pyrazinamide 40 mg/kg/day. Prednisone 2 mg/kg/day is given for 1 month. It has previously been documented that most tuberculomas disappear after 6 months of antituberculosis therapy. A repeat CT scan is therefore advocated after 6 months of therapy.[2] Treatment is complete once the lesion is no longer visible, is calcified or shows no contrast enhancement. It is important to remember that tuberculomas may paradoxically enlarge on treatment before reducing in size.

Treatment of neurocysticercosis should be individualised and the decision to use anthelmintics should be based on the number, location and viability of the parasite in the central nervous system. There is no consensus regarding the use of anthelmintics in children with neurocysticercosis.[1] Arguments against treatment include CT-demonstrated enhancing lesions that represent dying parasites and lesions that tend to resolve spontaneously. There is some evidence that radiological clearance may be accelerated in children with viable lesions. However, this does not affect seizure recurrence. Albendazole has become the anthelmintic of choice and offers good CSF penetration.[4] There is not enough evidence regarding the optimal dose or duration of treatment with albendazole. The published dose schedule is 15 mg/kg/day and short courses of 7 days or less are as effective as longer courses.[5] Adverse events during treatment are common and include headache, dizziness and gastrointestinal symptoms. Corticosteroids are frequently used in children with multiple neurocysticercosis lesions because of the possibility of cerebral oedema after drug-induced cyst death. The drug of choice is either dexamethazone 0.5 mg/kg/day or prednisone 1 mg/kg/day 24 hours before anthelmintic treatment and for the duration of such treatment. There is some evidence that corticosteroids reduce adverse effects associated with albendazole. Children presenting with single lesions do not require steroid cover.

There is consensus that anti-epileptic therapy is indicated in all children with symptomatic seizures and carbamazepine is the drug of choice. The starting dosage is 5 mg/kg/day in 2 - 3 divided doses, increasing by increments of 5 mg/kg/day, if clinically indicated, to a recommended dosage of 15 - 20 mg/kg/day in 2 - 3 divided doses. Often therapy can be withdrawn after 6 months if the child remains seizure free. In such cases, repeat imaging is not recommended. If seizures recur after the acute phase of inflammation and involution of the cyst, a diagnosis of epilepsy should be considered, and anticonvulsant therapy should continue until a 2-year seizure-free period has elapsed.

References available at www.cmej.org.za

Approach to headaches in children

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Headache is a common problem in childhood – up to 25% of schoolchildren suffer from chronic, recurrent headaches. Although primary headaches are far more common than those with a secondary cause, it is the latter that result in the most anxiety for families.[1] A logical approach to investigating and managing headaches is needed.

Classification

According to the *International Classification of Headache Disorders,* 2nd ed., headaches are divided into those with primary and those with secondary causes. Primary headaches occur independently of any other medical condition, while secondary headaches are directly attributed to an underlying medical cause.[2]

History

A detailed history is vital to identify the characteristics of the headache and to exclude secondary causes. Pertinent questions that need to be answered are set out in Table I.[1]

Examination

The general examination should include vital signs, including blood pressure and temperature. Meningitis should be excluded in all children with pyrexia. The skin must be examined for features of neurocutaneous syndromes. An increased head circumference may be a reflection of chronically raised intracranial pressure. Exclusion of other non-neurological causes entails examination of the ears, nose and throat (ENT), sinuses, teeth, temperomandibular joint and cervical spine. It is useful to evaluate visual acuity by means of a Snellen's chart to exclude refractive errors that may cause headache.