LETTERS TO THE EDITOR

Neurocysticercotic versus idiopathic epilepsy: a comparative study of 175 patients

Epilepsy seems to be the most common clinical manifestation of neurocysticercosis (NC),^{1,2} while NC has been regarded as the most commonly identifiable cause of epilepsy in some developing countries.^{3,4}

Two groups of chronic epileptics, one with NC (61 patients) and the other with idiopathic epilepsy (in whom no cause of epilepsy could be identified) and normal CT scan (114 patients), were compared for sex, age, age of onset of epilepsy, type of epilepsy (International Classification 1981),⁵ and EEG findings. These patients were prospectively and randomly selected from a group of 580 epileptic outpatients who were evaluated and studied during the past four years. Single convulsions and patients less than 10 years old were excluded. A normal neurological examination was a criterion of inclusion in both groups. CT was performed in each case, and CSF examination when indicated. Only patients with a normal CT scan were included in the idiopathic epilepsy group. A standard 8-channel EEG was obtained in all patients. An EEG was considered abnormal if generalised or focal spike discharges or sharp waves were present or if paroxysmal slow activity (delta or theta; generalised or focal) was observed in the absence of spike discharges or sharp waves. The diagnosis of NC was made with the following criteria: 1) CT scan findings compatible with NC;⁶ 2) Positive CSF immunological test (indirect immunofluorescence or indirect haemagglutination) for NC; and 3) Surgical diagnosis of NC. For the control of seizures, a patient was considered free of seizures when epileptic fits did not occur after one year of drug therapy and follow up. All the patients were taking anticonvulsant drugs regularly at the time of this study.

The most common tomographic finding in

the NC group was multiple parenchymal calcifications (n = 55, 90.1%), followed by parenchymal cysts (n = 10, 16.4%), and hydrocephalus (n = 3, 4.9%).

CSF examination was performed in 18 (29.5%) patients of the NC group. Ten patients (16.4%) with neurocysticercosis showed alterations in the CSF, as follows: elevated protein levels (more than 50 mg/dl) 6, pleocytosis (more than five cells/cu mm) 7 (all with mononuclear preponderance), positive immunological test for NC in nine cases. Low CSF glucose levels were not observed in any case. Eosinophils (more than 5%) were observed in two cases. Twelve patients (19.7%) had the active form of NC, according to the classification of Sotelo *et al.*² A spinal tap was performed in 32 (28.0%) patients with idiopathic epilepsy, as part of another protocol of investigation not related to this work. All CSF examinations of the idiopathic group were normal.

There was a higher proportion of late onset epilepsy (onset over 25 years old) in the NC group (28) (46.6%), compared with the idiopathic group (33) (29.4%) [Chi square test = 5.0515; p < 0.05]. Normal EEGs were more prevalent in the NC group. Among the patients with the active form of NC, only one (8.3%) had an abnormal EEG, which consisted of continuous slowing of the right posterior temporal area. In both groups, there was a high proportion of focal EEG abnormalities. No type of epilepsy was characteristic of either group of patients. The modality of drug therapy and the control of seizures did not differ significantly between both groups (table).

The only features distinguishing the NC epileptics from the idiopathic epileptics were the increased prevalence of late onset epilepsy and the higher number of normal EEGs in the NC group. The greater proportion of late onset epilepsy may be due to the average large interval from the initial infestation to onset of symptoms (up to 30 years). The great proportion of normal EEGs in patients with NC has already been observed by other authors,^{1,7} even in the active forms. In addition, focal EEG abnormalities are more prevalent than diffuse.⁷ The number of normal EEGs in the idiopathic group is in keeping with previous reports.⁸ Although there was a tendency for more female epileptics in the NC group, this difference did not reach statistical significance. This observation may be due to sex related

differences in the intensity of the host inflammatory response to cysticerci in the central nervous system.⁹

In conclusion, any patient with late onset epilepsy, with a normal neurological examination, and living in an endemic area of taeniasis/cysticercosis, should be considered as having neurocysticercosis, regardless of the seizure type and the EEG findings.

W O ARRUDA
Serviço de Neurologia,
Centro de Saúde Metropolitano,
Secretaria de Saúde do Estado,
Curitiba PR, Brazil

Correspondence: Dr Arruda, Rua Gonçalves Dias 713, 80-240 Curitiba PR, Brazil.

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Angiostrongylus cantonensis abscess in the brain

Angiostrongylus cantonensis is a rat lung worm with a geographical distribution from Madagascar to Hawaii. Humans are a paratenic intermediate host and angiostrongyliasis can present with protean clinical manifestations. Diffuse eosinophilic meningoencephalitis is the commonest presentation caused by the young adult worms in the subarachnoid space. We report an angiostrongylus abscess presenting with focal seizures which to our knowledge is the first report of its kind.

A 45 year old man presented with headache and right focal tonic clonic seizures, which he had had for a few months, starting in the right eyelid and face, with subsequent generalisation. On examination he had right spastic hemiparesis grade IV. There were no signs of raised intracranial pressure or any systemic disease. Plain and contrast CT scan of the head showed a left parietal enhancing disc shaped lesion 2 cm in diameter (fig 1). He was treated with anti-tuberculous and anticonvulsant drugs after a presumptive diagnosis of tuberculoma. Two months later he presented with abnormal tonic movements in the right upper limb. On examination he had spastic right hemiparesis grade III-IV with atrophy of the right upper and lower limb muscles. There were no features of raised ICP. Routine haematological and biochemical tests were normal. ESR, eosinophil count, EEG,

Table Clinical findings of NC and idiopathic group of chronic epileptics

	Neurocysticercosis	Idiopathic
Total number	61	114
Sex (M/F)	24/37	60/54
Sex ratio	1.0:1.5	1.1:1.0
Present age, mean (SD)	35.6 (14.2)	32.8 (12.4)
Type of epilepsy		
1) Generalised	27 (44.2%)	53 (46.5%)
2) Partial		
Simple	3 (4.9%)	4 (3.5%)
Complex	6 (9.8%)	7 (6.1%)
3) 1 + 2	4 (6.5%)	10 (8.7%)
4) Secondarily generalised	18 (29.5%)	36 (31.5%)
5) Unclassifiable	3 (4.9%)	1 (0.9%)
Normal EEG	43 (70.5%)	62 (54.4%)*
Abnormal EEG	18 (29.5%)	52 (45.6%)
Focal	13 (72.7%)	30 (57.7%)
Generalised	5 (27.7%)	22 (42.3%)
Therapy		
Monotherapy	51 (83.6%)	82 (72.0%)
Control of seizures		
Free of seizures	36 (59.0%)	72 (63.1%)
Decrease of frequency	21 (34.4%)	32 (28.0)
Unchanged	4 (6.5%)	10 (8.7%)

*—Chi square test, p < 0.05.

Figure 1 CT scan of head showing a left parietal enhancing disc shaped lesion.

chest and lower limb radiographs, abdominal ultrasound and barium meal follow up were normal.

A left parietal craniotomy was carried out and a circumscribed firm mass 2 cms in diameter was palpated under a widened sulcus. When opened the lesion was purulent and contained a 2.5 cm long motile thread-like worm. The mass and worm were excised. Postoperatively the patient gradually recovered from the hemiparesis and is free from seizures and dystonia at two years follow up.

Sections studied showed microabscesses with necrotic material, neutrophils, eosinophils and lymphomononuclears (fig 2). The parasite was identified as that of *A. cantonensis* (fig 3).

Angiostrongylus cantonensis is a metastrongylid nematode with the rat as a definitive host, and slugs and snails as intermediate hosts. Human infection is an example of aberrant parasitism and is caused by ingesting

raw or insufficiently cooked food containing the third stage larvae.

Cerebral angiostrongyliasis usually has an incubation period of two weeks with headache, lowgrade fever and meningeal signs as the common presenting features.¹ Watts reported five cases of eosinophilic meningitis with epileptic seizures in one case.² Schmutzhard *et al* reported five patients with *A. cantonensis* meningoencephalitis with one case having bilateral abducens nerve palsy and unilateral papilloedema, with no spinal or intracerebral lesions.³ Prommiindaraj *et al* reported ocular angiostrongyliasis.⁴ Escobar and Nieto described pathological findings of *A. cantonensis* infection in the human brain and proposed that as the larva dies the inflammatory reaction changes from diffuse eosinophilic meningoencephalitis to a more focal and granulomatous response.⁵ This case is unique in that the patient presented with focal neurological manifestations without diffuse meningoencephalitis. CT scan revealed a single lesion and histopathology showed an abscess with a live worm.

In India, an enhancing ring or disc lesion in the cerebrum with perilesional oedema is usually considered to be a tuberculoma.⁶ It is our usual practice to treat these patients with anti-tuberculous drugs, with follow up scans to monitor the response. Our patient was initially given anti-tuberculous drugs. His condition deteriorated, however, and at follow up eight weeks later a CT scan showed that the lesion still persisted. The diagnosis was therefore revised and excision of the lesion confirmed our diagnosis.

AK PUROHIT
I DINAKAR
C SUNDARAM
KS RATNAKAR

Departments of Neurosurgery and Pathology,
Nizam's Institute of Medical Sciences,
Panjagutta, Hyderabad 500 482 (AP),
India

Correspondence to: Dr Purohit

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Posterior fossa dermoid cysts and the Klippel-Feil syndrome

Although the Klippel-Feil syndrome as originally described comprises a triad of short neck, painless restriction of neck movements and low posterior hairline due to the congenital fusion of two or more cervical vertebrae, more serious anomalies of the cardiovascular,

renal and central nervous systems may co-exist.¹ The following case report illustrates another important association which we feel has not received sufficient emphasis.

A 14 year old girl presented with a 12 month history of bitemporal headache and intermittent vertigo with nausea for two months. From the age of three months she had developed intermittently a soft discharging swelling over her occiput, which had been incised on three occasions in her local casualty department. She had been known to have Klippel-Feil syndrome since the age of four when she presented with a Sprengel's deformity of the right shoulder.

She had a short webbed neck with a low hairline which extended as far as the C7 vertebral prominence. No occipital swelling was present, but a small punctum was present just above the inion. Neurological examination revealed vertical nystagmus, dysidiadochokinesis, and mirror movements of the hands.

Radiographs of the cervical spine (fig a) showed occipitalisation of the posterior arch of C1 with the anterior element apparently fused to C2, posterior fusion of C2 and C3, and anterior and posterior fusion of C4-6. A chest radiograph showed fusion of the anterior ends of the first and second ribs on the right, a left hemivertebra at D3 and a scoliosis concave to the right.

CT scan of the skull (fig b) showed a large midline hypodense mass lesion in the posterior cranial fossa with peripheral flecks of calcification, and associated compression of the inferior pons and medulla and anterior displacement of the fourth ventricle. There was moderate dilatation of the upper part of the fourth, third and both lateral ventricles, and a small midline defect in the inner table of the occipital bone with adjacent thickening. Appearances following the injection of intravenous contrast were unchanged.

At operation she was seen to have a tract with thick walls which ran inferiorly through the occipital bone into a large dermoid cyst which filled the cisterna magna, causing upward displacement of the cerebellar hemispheres. The posterior arch of C1 was absent. The cyst was opened and decompressed. A portion of the cyst capsule was adherent to the upper spinal cord and medulla and was not removed. Histologically the cyst capsule was lined with stratified squamous epithelium. The cyst contained hair and scattered foci of calcification. The patient had no complications from surgery and remains well.

Congenital fusion of the cervical vertebrae is due to failure of normal segmentation of the cervical somites during the third to eighth week of gestation. Similarly, dermoid cysts originate during the third to fifth week, when cleavage of the neuroectoderm from the epithelial ectoderm along the mid-dorsal aspect of the embryo may be incomplete. The resultant persistent cutaneous defect may extend from the skin into the substance or central canal of the central nervous system, and expand into a cyst containing sebaceous material, hair and epithelial debris.² Posterior fossa dermoids may present with symptoms and signs of a mass lesion or with staphylococcal meningitis.³ The pathway of infection is via the dermal sinus which may be seen as a small punctum or dimple over the occiput.

We are aware of four previously reported cases of posterior cranial fossa dermoid cysts associated with cervical fusion anomalies.²⁻⁴ Given the rarity of both Klippel-Feil syndrome⁷ and intracranial dermoid cysts,⁸ the association is probably significant, but is not

Figure 2 Abscess with neutrophils and eosinophils around the cut section of parasite. H & E $\times 40$.

Figure 3 *Angiostrongylus cantonensis*. H & E $\times 10$.