THE LOCALIZING VALUE OF DOWNBEAT NYSTAGMUS

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ABSTRACT

Downbeat nystagmus (DBN) is a primary position nystagmus with the fast phase in a downward direction. It is a rare but distinctive disorder of ocular motility and usually localizes the lesion at the posterior fossa.

Four patients with DBN were seen in the department. One had a medullary glioma, and another congenital basilar invagination. The other two were initially diagnosed as demyelinating disease. One was subsequently found to have Arnold Chiari Malformation on magnetic resonance imaging (MRI).

Review of the literature showed that cerebellar ectopia (Arnold Chiari Malformation) is the commonest cause of DBN. However ⅓ of reported cases have no obvious cause.

DBN is of such high localizing value that we recommend MRI of cervicomedullary junction for all patients with DBN to exclude cerebellar ectopia or medullary lesion.

Key Words: Downbeat nystagmus, Cerebellar ectopia, Cervico-medullary junction, Magnetic resonance imaging, Posterior fossa lesion

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INTRODUCTION

Downbeat nystagmus (DBN) is a specific neurologic entity of primary position vertical jerk nystagmus, usually exacerbated by lateral gaze. It differs from gaze-evoked DBN in that the latter is produced only by downward gaze and not in primary position. DBN is usually maximum when the eyes are deviated laterally and slightly below the horizon and not in downward gaze. DBN is an easily elicited clinical sign and has high localizing value.

DBN was first highlighted by Cogan and Barrow in 1954 (1) when they described nine cases of platybasia and Arnold Chiari Malformation (cerebellar ectopia) with DBN. Since then DBN has been described in various conditions with lesions at the cervico-medullary junction (2, 3). More recently it has been reported with alcoholic (3, 4, 5), anoxic (3) para neoplastic cerebellar degeneration (3, 6), spinocerebellar degeneration (2, 3, 7, 8), encephalitis (9), magnesium depletion (10), amiodarone toxicity (11), lithium intoxication (3, 12), Vitamin B12 deficiency (13), anticonvulsant therapy (14, 15, 16) and even in a congenital form (17).

One third of all the patients with DBN will have Arnold Chiari Malformation (ACM) and up to forty percent of cases the cause remains undiagnosed (3).

We describe here four cases of DBN.

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CASE I

This 35 year old Chinese man had a 2-year history of unsteady gait and presented himself to Alexandra Hospital in March 87 in a comatose state from spontaneous hypopnea. He was intubated in the emergency room and the cyanosis cleared off before he regained full consciouness. He was then transferred to Department of Neurology, Tan Tock Seng Hospital for further management. Clinical examination revealed a conscious man on a mechanical ventilator with no cranial nerve palsy. DBN was observed in the primary position and maximum on looking laterally. There was no pyramidal signs. However he had gross truncal ataxia with dysmetria and dysdiadochokinesia of left extremities. Blood investigations were essentially normal and CT scan of the cervico-medullary junction (Fig 1) was reported as normal.

He was managed conservatively and weaned off the respirator. He was breathing spontaneously and the rest of the clinical signs remained unchanged. A bilateral vertebral angiogram showed no abnormality. However later that day, he developed spontaneous apnea and was put back on mechanical ventilation. The neuro-ophthalmic examination remained unchanged but EKG showed non-Q anteroseptal myocardial infarction. He was managed conservatively and remained well. He was extubated again without any problem three weeks later.

CT scan brain with intrathecal contrast was done and it finally revealed a mass in the right anterior lateral aspect of the medulla extending down to the CI spinal cord (Fig. 2). In view of his recent myocardial infarction, surgical resection was postponed.

Three months after first presentation, he was found to be cyanotic and unresponsive in bed at 8.00 am. After resuscitation, he regained consciousness but neurologic examination revealed a right Abducen Nerve palsy, right facial nerve palsy and inability to look to the right both voluntarily and on oculo-cephalic reflex. He also had dense left hemiplegia. Repeat CT scan two weeks later showed an additional low radiolucency area in the right internal capsule.

He was managed on mechanical ventilation for the rest of his two-month stay in hospital. Four days before he died, he was noticed to be drowsy and neuroophthalmic examination revealed unequal pupil size, right 3mm and left 2mm, which reacted only sluggishly to light. Urgent CT scan (Fig 3) showed bilateral dilatation of ventricles with extension of the tumour into right inferior aspect of the thalamus. He died in August 1987.

Re'sume': A 35 year old man presented with spontaneous hypopnea highly suggestive of a lesion in the medulla oblongata with involvement of the respiratory centers. The localization was further ascertained with signs of DBN and cerebellar dysfunction. However initial CT scan and vertebral angiogram did not disclose any abnormality. It was only with intrathecal contrast medium that the CT scan revealed the medullary tumour. Unfortunately he suffered a myocardial infarction during one of the apneic spells and finally died from complications of the tumour.

CASE 2

This 29 year old Chinese man was seen in a medical department in June 1982 for complaints of weakness of both lower limbs. Clinical examination then revealed mild weakness of right lower limb with hyperreflexic knee jerk. There was no nystagmus and the rest of the neurological findings were normal. CT scan of the brain revealed no abnormality. His lower limb weakness became progressively worse and he was admitted on after a fall. Neurological examination showed full extraocular movement with downbeat nystagmus; hyperreflexia of all 4 limbs with spasticity, bilateral ankle clonus and Babinski signs; left hemianesthesia and left cerebellar dysfunction. A cervico-medullary junction lesion was suspected and CT scan with intrathecal contrast (Fig 4) revealed that the atlas and odontoid peg were inside the posterior fossa and the subarachnoid space between the medulla and the odontoid process was obliterated. Subsequently he had a transoral resection of odontoid peg. Post operatively there was no change in his clinical state except that the nystagmus was no longer detectable.

Re'sume': A 29 year old man with basilar invagination presented with progressive weakness with spastic gait for 5 years. The final diagnosis of basilar invagination was only made when he was investigated for downbeat nystagmus.

CASE 3

This 20 year old Indian lady presented with difficulty in swallowing with regurgitation of fluid in Oct 1983. Neurological examination was normal except for mild left ptosis with weak orbicularis oculi bilaterally. EMG showed decremental response in the left orbicular oculi and she was treated as for myasthenia gravis with thymectomy and pyridostigmine. Post-operatively, she continued to have mild regurgitation of fluid despite medication. During follow up she developed left lateral rectus weakness with horizontal nystagmus on lateral gaze in Aug 86. There was no evidence of any cerebellar dysfunction. CT scan brain revealed no abnormality in Aug 86. Subsequently, her external ocular movement became full but the gaze-evoked nystagmus persisted.

In Jan 87 she developed mild left dysmetria with dysdiaodochokinesia while in India and CT scan brain done there revealed no structural abnormality too. By March 87, DBN on vertical gaze was noticed for the first time besides horizontal nystagmus. Bilateral vertebral angiogram was done and detected no abnormality. She was tentatively treated as for demyelinating disease without much improvement. When MRI facility was made available in Singapore in July 87, it revealed an Arnold Chiari Malformation with tonsillar herniation (Fig 5). A sub-occipital atlas-axis decompression was done in September 1987. Six months after the operation, her cerebellar dysfunction was no longer present but she still has gaze-evoked nystagmus.

Re'sume': A young lady presented with signs and symptoms of myathesia gravis 5 years ago and started to have brain-stem signs 2 years ago. However all investigations then were negative and she was treated as for demyelinating disease. The final diagnosis of ACM was only made with the help of MRI after she developed DBN.

CASE 4

This 34 year old lady had sudden onset of oscillopsia for one week before admission in June 1988. Neuro-ophthalmic examination revealed gross DBN in all direction maximum on lateral gaze. The extraocular movements were full and there was no pyramidal signs. However there were mild dysmetria and dysdiaodochokinesia on the right side. Both CT scan brain and MRI were normal and excluded an Arnold Chiari Malformation. Cerebro-spinal fluid showed a cell count of 95 with 30 mg% protein. A diagnosis of brainstem encephalitis was made. However she developed bilateral gross cerebellar dysfunction with ataxia over the next 10 days. Despite a course of high dose steriod, she remained grossly ataxic with marked cerebellar dysfunction though the amplitude of DBN became smaller. Repeat CSF examination was completely normal and three months later, she still had the same clinical findings.

Re'sume': A young lady presented with DBN and developed gross cerebellar dysfunction over the next few days. Cervico-medullary junction lesion was excluded with CT scan and MRI. Three months later, she still had the neuro-ophthalmological deficit. She most probably had post viral demyelination.

DISCUSSION

All four patients in our series did not indicate a uniform etiology of DBN. They were basilar invagination, medullary glioma, Arnold Chiari Malformation and demyelinating disease. Zee et al (18) have shown that DBN develops in all flocculectomized monkeys, and medullary midline section extending from the obex to Abducen nucleus produces DBN. It is clear that the vestibulo-cerebellum is the anatomic site of the major lesion in DBN but the pathogenesis remains uncertain. Baloh and Spooner (7) postulated an imbalance in the central vestibular system as the mechanism for DBN. Zee et al (19) proposed that DBN originated from a deficit in the downward pursuit sustem, whereas Halmagyi (3) and Gresty (20) suggested that a loss of vestibulo-cerebellar inhibition of upward otolith ocular reflexes as the mechanism.

Review of three large series (table 1) showed that the sites of lesion are either in the lower medulla or cerebellum, and none of them had any lesion caudal to the Foramen Magnum. As pointed out by Cogan and Barrow (1), the neuro-ophthalmologic symptoms in cerebellar ectopia are due to the impingement of the odontoid

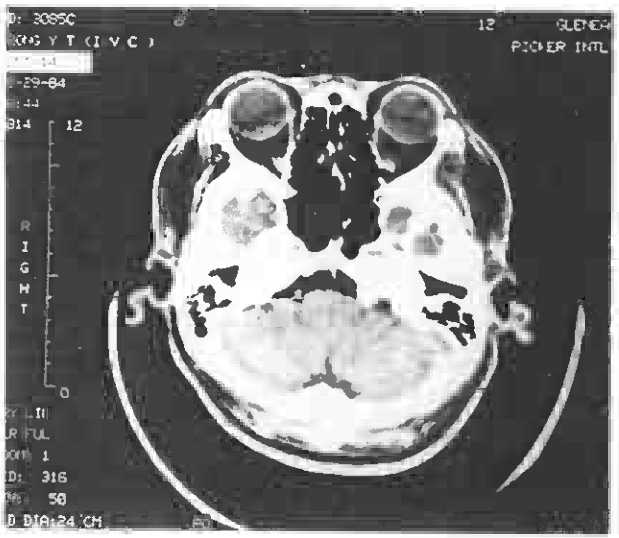

Fig 1

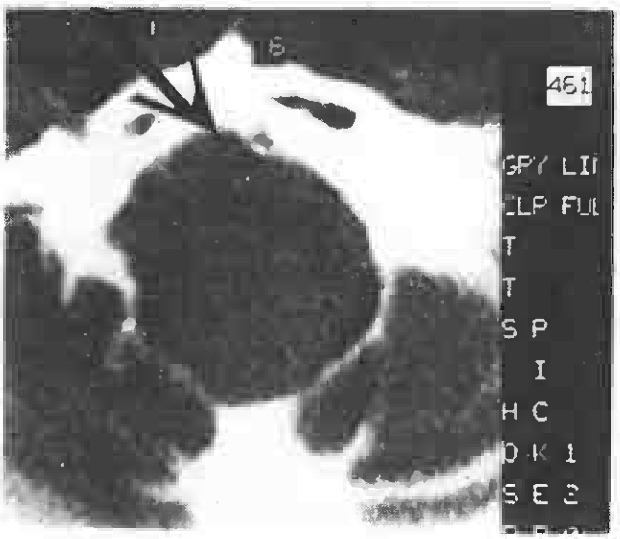

Fig 2

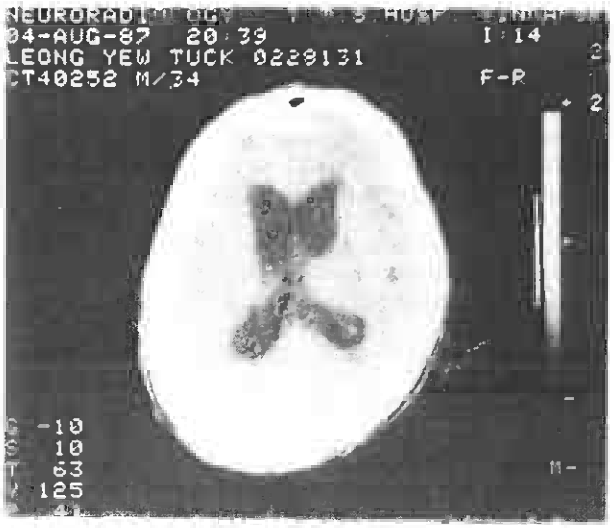

Fig 3

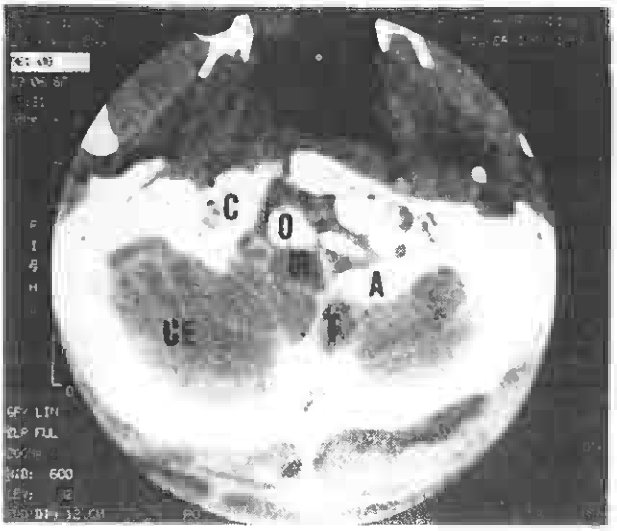

Fig 4

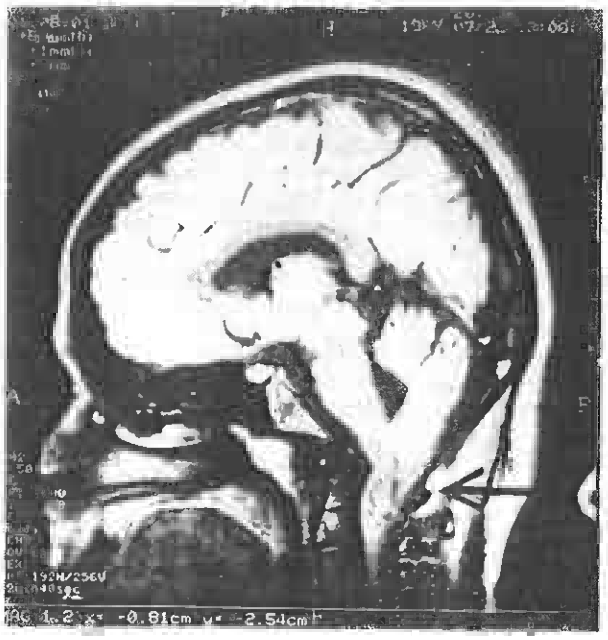

Fig. 5

Fig 1 (Case 1): Normal CT scan done on admission.

Fig 2 (Case 1): CT scan with intrathecal contrast showing a mass (arrowed) anterior to the medulla.

Fig 3 (Case 1): CT scan showing dilated ventricles.

Fig 4 (Case 2): CT scan of posterior fossa showing the atlas and odontoid peg within the posterior fossa and the closeness between the odontoid peg and the medulla.
M , medulla
C , clivus of the occipital bone
O , odontoid peg;
T , tonsil of the cerebellum;
A , part of the atlas;
CE, cerebellum

Fig. 5 (Case 3): MRI showing tonsillar herniation.

process on the brain stem and cerebellum. Fisher (21) also postulated that DBN is mainly a dysfunction of the medulla and cerebellum as lesions limited to the cervical spinal cord do not cause DBN. This was shown by Yasuoka et al (22) who analysed 57 cases of foramen magnum tumours and did not notice any DBN in their patients. Howe and Taren (23) also noted the absence of DBN in cases of foramen magnum tumours.

We conclude that DBN indicates, with rare exception, a lesion of the posterior fossa involving the medulla or the flocculus of the cerebellum rather than the cervico-medullary junction.

DBN occurs in a number of disorders. It may occur as an isolated congenital phenomenon in children without any evidence of intracranial disease (17). DBN has been seen in patients with various metabolic disorders viz lithium toxicity, amiodarone toxicity, anti-convulsant therapy especially dilantin and carbamazepine, vitamin B12 and thiamine deficiency and magnesium depletion. DBN due to these causes tends to be reversible with withdrawal of the offending drugs or correction of the deficiency. If DBN persists after medical treatment, a diligent search should always be made for a surgically correctable lesion in the posterior fossa near the cervico-medullary junction. The neurological investigations should include magnetic resonance imaging as this is much more informative than a metrizamide computed tomographic scan. As seen in this report, the clinical recognition of DBN led to a suspicion of cerebellar ectopia and subsequently ACM was confirmed on MRI. With the introduction of MRI, we believe that more and more cases of 'undiagnosed' DBN will be shown to have cerebellar ectopia.

Table 1.
CAUSES OF DOWNBEAT NYSTAGMUS

	Cogan 1968	Baloh 1981	Halmagyi 1983
Cerebellar ectopia	9	9	17
Cerebellar degeneration	4	2	18
		6 Hereditary	
		5 Sporadic	
		1 alcoholic	
		4 para-neoplastic	
		2 anoxic	
Posterior fossa tumour	3	2	1
Brain stem infarction	3	2	
Multiple sclerosis	3		
Lithium carbonate			1
Others	1		1
Undiagnosed	4	2	27
Total	27	17	65

Addendum:
On follow-up, a breast lump was found in Case 4 and histology confirmed the presence of carcinoma. The final diagnosis should be Ca Breast Downbeat nystagmus.

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