Bahrain Medical Bulletin, Volume 17, Number 3, September 1995

HYPESTHETIC ATAXIC HEMIPARESIS

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A case of Hypesthetic Ataxic Hemiparesis (AH) in a 64 year old Bahraini female is reported. She was not hypertensive or diabetic. CT scan revealed a lacunar infarct in the opposite posterior limb of the internal capsule and adjacent lateral part of thalamus. The possible sites of lesion in the Ataxic Hemiparesis syndrome and the clinical features of supratentorial and brainstem AH are highlighted. Bahrain Med Bull 1995;17:

Ataxic hemiparesis (AH) a clinical syndrome characterized by ipsilateral pyramidal weakness and cerebellar ataxia was first described by Fisher and Cole1. Originally persistent sensory disturbances were not regarded as part of the AH syndrome2. Later as more cases were reported, the spectrum of AH was widened to include cases with hemi sensory deficits ipsilateral to the ataxia and weakness3,4. This has been termed "hypesthetic ataxic hemiparesis" (HAH). We report a case in a 64 year old normotensive Bahraini woman with sickle cell trait.

THE CASE

A 64 years old woman woke up with weakness of the right upper and lower limbs, inability to stand or walk and numbness of the right half of the body. She could move her limbs but had difficulty to hold objects with her right hand. There was no history of headache, vomiting, vertigo or diplopia. She was not a known hypertensive or diabetic. She gave no past history suggestive of transient ischemic attacks. She used to smoke heavily but gave it up 20 years ago.

On examination she was not anaemica or cyanosed. Pulse rate was 80/mt and regular and BP was 160/90 mmHg. There was an ejection systolic murmur at the left sternal edge and aortic area. Carotid pulses were normal and there was no bruit. All peripheral pulses were well felt.

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Neurologically she was alert and oriented. Speech was normal. Fundi could not be visualized due to old healed corneal ulcers. Mild upper motor neuron type of facial weakness was present on the right side. All other cranial nerves were normal. She had right hemiparesis with a motor power of grade 4/5 in the right upper and lower limbs.

Power was normal on the left side. Deep tendon reflexes were brisk on the right side. Right plantar reflex was extensor and the left was flexor. Marked finger - nose and heel- knee incoordination was observed on the right side. Coordination was normal on the left side. Sensory examination revealed diminished pinprick, touch, vibration and position sense on the right side of the body. She could not walk without support.

Routine hematological and biochemical investigations were within normal range. Blood VDRL was nonreactive. She had a positive sickling test and hemoglobin

electrophoresis revealed sickle cell trait. CT scan of the brain (Fig 1) revealed a hypodense lacunar infarct measuring 1.2 cm x 1.2 cm x 1.5 cm in the posterior limb of the internal capsule and adjacent lateral part of thalamus on the left side. Echocardiogram showed calcific aortic sclerosis without significant stenosis. There were no clots in the left ventricle (LV) and LV function was good.

She was started on soluble Aspirin 100 mg daily and Oxpentifylline 400 mg twice daily, and commenced on physiotherapy. On follow up, six weeks after her discharge from hospital, she had shown good improvement. She could walk without support. Power in the right upper and lower limbs had improved. But she still had finger-nose incoordination and sensory abnormalities on the right side.

DISCUSSION

Fisher and Cole[1] attributed the causative lesion of AH to an infarct in the posterior limb of the internal capsule. Later in 1978 Fisher[5] reported pathological confirmation in 3 patients, in whom infarct was found in the basis pontis opposite to the side of the neurological deficit. He suggested internal capsule, corona radiate and midbrain as well as basis pontis as possible sites of lesion causing this syndrome. Ipsilateral pyramidal and cerebellar deficits can occur in both supra and infratentorial lacunar infarcts. Subsequently lesions have been demonstrated in all these sites resulting in AH. However, on the basis of the clinical features, it may be possible to differentiate AH due to a supratentorial lesion from AH due to an infratentorial or brainstem lesion. Hemi sensory impairment is seen only in supratentorial lesion, whereas dysarthria, nystagmus and lower cranial nerve palsies occur in brainstem lacunar infarcts.

The syndrome of hypesthetic ataxic hemiparesis (HAH) comprises of ipsilateral motor, sensory and cerebellar dysfunction on one side of the body due to a contralateral lesion in the region of the posterior limb of the internal capsule and adjacent lateral part of thalamus or in the paraventricular region. Helgason and Wilbur[4] reported 23 patients with HAH, all of them had infarcts of lacunar size or slightly larger on CT or MRI studies of brain in the contralateral posterior limb of the internal capsule. The infarct was of lacunar size (up to 1.5 cm) in only 8 of these patients, and in only one of them was the infarct confined to the posterior limb of the internal capsule and adjacent lateral part of the thalamus. The patient we are reporting had HAH on the right side and CT scan of brain revealed a lacunar infarct in the posterior limb of the internal capsule and adjacent lateral part of the thalamus on the left side.

Hypesthetic ataxic hemiparesis has been reported not only due to lacunar infarctions of the thalamus[6] but also due to thalamic haemorrhage[7]. In these cases pyramidal signs were minimal and evanescent whereas the sensory deficits and ataxia dominated the clinical picture.

The anatomical basis of the lesions causing AH has been fairly well explained. It is likely that interruption of the cortico-ponto-cerebellar fibers along with the pyramidal fibers in the basis pontis is responsible for pontine ataxic hemiparesis. It has been suggested that the ataxia in capsular AH is due to the involvement of ascending dentato-rubro-thalamo-cortical pathway rather than to the descending cortico-ponto-cerebellar fibers.

The exact vascular supply of the region involved in HAH is uncertain. Recent reports have highlighted the possibility of an anterior choroidal artery infarct responsible for this syndrome[4,6]. HAH was also attributed by Ghika et al[8] to lateral lenticular striate territory infarction.

Majority of the cases of AH reported in the literature had systemic hypertension. In one review 22 of the 26 patients had hypertension[3]. But it is well known that lacunar infarctions can also occur in normotensive individuals[9-11]. The patient under discussion is neither hypertensive nor diabetic. Her BP remained normal throughout her stay in the hospital. It has been suggested that in such normotensive individuals, large vessel disease with small vessel embolization may be responsible for AH. Perman and Racy[12] have demonstrated bilateral vertebral and carotid arterial stenosis on angiographic studies in a case with homolateral ataxia and crural paresis.

Although our patient was detected to have sickle cell trait, review of literature and to the best of our knowledge no association of HAH and sickle cell trait has been reported.

CONCLUSION

Hypesthetic Ataxic Hemiparesis is uncommon clinical syndrome, and in the absence of definite risk factors, one should look for disorders like hemoglobinopathies as a possible risk factor especially in areas where sickle cell disease is common.

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