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HIRAYAMA DISEASE - CERVICAL FLEXION INDUCED MYELOPATHY PROGRESSING TO SPASTIC HEMIPARESIS :-

General Medicine

Dr. Gaurang Patel* 3rd Year Pg Student In Medicine, SBKS MI&RC and Dhiraj Hospital, Piparia,Vadodara.
*Corresponding Author

Dr. P. R. Jha Professor Of Medicine, SBKS MI&RC and Dhiraj Hospital, Piparia,Vadodara.

KEYWORDS

INTRODUCTION:

Hirayama disease (HD) is a rare disease affecting primarily young men in the second to third decades of life[1-2]. Hirayama disease (HD)-cervical flexion induced myelopathy (CFIM) is a lower motor neuron disease conventionally affecting a single upper extremity.[1]It is characterized by the insidious onset of unilateral or asymmetric atrophy of the hand and forearm with sparing of brachioradialis, giving the characteristic appearance of oblique amyotrophy involving the C7, C8 and T1 myotomes.[3]It is thought to be a kind of cervical myelopathy related to flexion movements of the neck.[4] HD differs from classical types of motor neuron diseases (MND) because of its non-progressive course and pathologic findings of chronic microcirculatory changes in the territory of the anterior spinal artery supplying the anterior horns of the lower cervical cord.[5-6]

The pathogenetic mechanism of this disease is attributed to forward displacement of the posterior wall of the lower cervical dural canal when the neck is in flexion, which causes marked, often asymmetric, flattening of the lower cervical cord.[6-7]Since HD differs from MND, or spinal muscular atrophy, this disease entity should be more widely recognized as prognosis in this condition is benign and early detection and effective treatments may be considered.[8]

Initial phase of HD knows as Monomelic amyotrophy (MMA), is diagnosed on clinical and imaging characteristics, with cervical collar as timely intervention for prevention of further progression. [9]

Radiological findings are essential for the diagnosis of Hirayama disease. Neutral and flexion cervical MRI shows asymmetrical atrophy of the spinal cord, forward displacement of dural sac with neck flexion and thus, secondary cord compression against the posterior wall of the vertebral body. Since neck flexion was recognized as a possible cause of the disease, several therapies have proved their utility, shortening the progression period and even improving the patient's strength. For cases in which the disease progresses despite conservative treatment, several surgical options have been used: posterior decompression with dural sac augmentation, with and without fusion, anterior decompression and fusion, and combinations of these methods. Which one is the most effective is still a matter of debate.[10]

History:

A 57 year old chronic bidi smoker farmer by occupation right handed person presented with chest pain and abdominal pain. Chest pain was retrosternal, in epigastric area, not associated with exertion, radiation, gabharaman and palpitation. Abdominal pain was dull aching, in epigastric region and not associated with food, nausea, vomiting, diarrhea, and position of body. Patient also had past history of asymmetrical weakness and loss of muscle mass in right upper limb followed by loosening of clothes on right side, unable to lift heavy objects with his affected limb since last 23 years. Neurological deficits progressed for 3 years and attained a stable course. He also reported that his condition worsened with the cold. There was no family history of neuromuscular disease. On physical examination, oblique atrophy was obvious (atrophy of the thenar and hypothenar eminences and of the cubital border of the forearm, with relative sparing of the brachioradialis muscle), and the patient had severe weakness of the intrinsic musculature of the hand. His wrist flexion and extension and his elbow extension were also affected. He had mild weakness of hip flexion and knee extension in the right lower limb but was still able to walk. The patient's deep tendon reflexes were normal in left upper limb and the left foot, but were brisk in the affected lower limb. He had a positive Babinski sign on the right. No sensory alterations were present, and the cranial nerves were unaffected.

On physical examination. (Muscle nutrition)

Height	175 cm	Right side			Left side		
Weight	44 kg	Upper limb	Forearm girth	17.5 cm	Upper limb	Forearm girth	19 cm
Arm span	185 cm		Arm girth	19.5 cm		Arm girth	20.5 cm
Upper segment	67 cm	Lower limb	Calf girth	25 cm	Lower limb	Calf girth	27 cm
Lower segment	108 cm		Thigh girth	33 cm		Thigh girth	35 cm

Vide figure number A,B,C.

And routine investigation showed as following:-

Hb :- 13.8 gm%		serum sodium :- 145mmol/L
TLC :- 5100 cells/cu.mm		serum potassium :- 4.2 mmol/L
Platelets :- 1,50,000 cells/cu.mm		serum chloride :- 106 mmol/L
random blood sugar :- 89 mg/dL		serum urea :- 30 mg%
serum bilirubin:- 0.4 mg%	Direct :- 0.2 mg%	serum creatinine :- 1.0 mg%
	Indirect :- 0.2 mg%	
SGPT :- 20 IU/L		
SGOT :- 21 IU/L		

Ultrasonography and electrocardiography were with in normal limit. And upper gastro-instestinal endoscopy (UGIscopy) was suggestive of severe duodenitis with H.pylori infection.

MRI brain and cervical cord showed atrophy at C5-6 level of spinal cord but no other abnormality.

NCS/EMG was suggestive of bilateral focal C8-T1 preganglionic (anterior horn cell) involvement with ongoing denervation and changes of chronic renervation.

DISCUSSION :-

My patient is presented with chest pain and abdominal pain since 10 days. Had right side weakness since 23 years. Routine blood investigations, electrocardiogram and ultrasonography of abdomen were with in normal limits. UGI scopy showed severe duodenitis with H.pylori infection. Patient's symptoms were improved with H.pylori regimen. Patient also had history of asymmetrical weakness, loss of muscle mass in right upper limb and not able to perform activity with his affected limb. Neurological deficit was progressive initially, then it remained static. His symptoms are worse with cold and have atrophy of thenar and hypothenar muscle. His reflexes are brisk and positive Babinski on affected side without sensory and cranial nerve involvement. MRI brain and cervical cord showed atrophy at C5-6. Nerve conduction test suggested bilateral focal C8-T1 preganglionic (anterior horn cell) involvement with ongoing denervation and changes of chronic renervation. These clinical, radiological and nerve conduction findings are suggestive of Hirayama disease.

Neck movement has impact on cervical cord[11]. In extension of neck, the dura mater of the cervical spine is slack and thrown into transverse fold. When the neck is in flexion the dura becomes tighter. As the neck moves from extension to flexion, the length of cervical canal is increased. In normal individuals, the slack of the dura can compensate

for increased length in flexion, so spinal canal have no anterior displacement.[11] But in Hirayama disease, there is no slack of dural canal in extension, because of an imbalanced growth of the duramater and vertebra. So tight dural canal is formed, which is not able to compensate for the increased length of posterior wall in flexion. This leads to anterior shifting of the posterior dural wall, with cord compression. Resulting in disturbance of blood supply of anterior 2/3 of spinal cord by anterior spinal artery.[12]

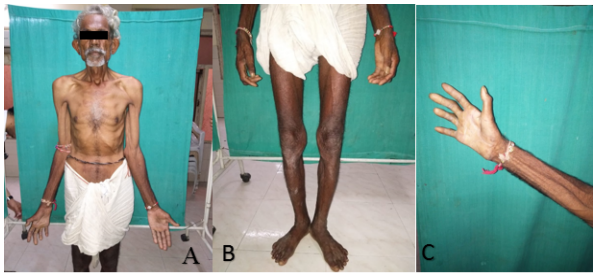
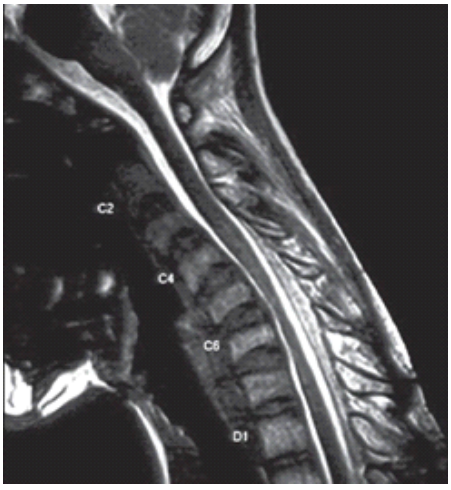


figure A shows:- Right sided muscle bulk is less than left limb with claw hand.
figure B shows:- Right sided muscle bulk is less than left limb claw hand,
figure C shows:- Atrophy of thenar and hypothenar muscle

MRI brain and cervical cord:- [suggestive of atrophy at C 5-6 level of spinal cord (white arrow)]



CONCLUSION:-

Hirayama disease (HD) is a rare disease occurs primarily in young men. It occurs in second to third decades of life. It progresses with flexion movements of the neck. Oblique atrophy is the characteristic of HD. With cervical collar can prevent the progression of disease. Its diagnosis is based on MRI of neck in flexion position.

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