IN THE NAME OF





MULTIPLE SCLEROSIS AND SYRINGOMYELIA

<u>H.Pakdaman</u> MD Professor of neurology <u>K.Gharagozli</u> MD Professor of neurology <u>Y. Kholghi</u> MD



Department of Neurology,Loghman Hospital,Shahid Beheshti University of Medicine,Tehran,Iran

INTRODUCTION

>ASSOCIATION OF MS WITH OTHER DISEASES.

>ASSOCIATION OF MS WITH SYRINGOMYELIA.



(SEIZURE, SYRINGOMYELIA?)

> ONE DISEASE AS CAUSATIVE FACTOR FOR OTHER.

(CIDP,AUTOIMMUNE DISEASES)

(STROKE,TUMORS,ANOMALIES) > POSSIBLY HAVE COMMON ORIGIN.

> COMPLETELY COINCIDENTAL.

ASSOCIATION OF MS WITH OTHER DISEASES

ASSOCIATION OF MS WITH SYRINGOMYELIA

IT HAS BEEN RARELY ASSOCIATED WITH MS. (Br j Radiol 1985;58:1206-8,Neurology

19940;40:718-21, Neurology 1992;42:464-5)

IT HAS BEEN REPORTED TO OCCUR MORE FREQUENTLY IN THE ASIAN MS PATIENTS. (Ann Neurol 1996;40:569-574)

SYRINGOMYELIA

- > Prevalence : 8.4 cases per 100,000 (No geographic or race difference)
- Familial cases have been described.
- > Occurs more frequently in men than in women.

The disease usually appears in the third or fourth decade of life, with a mean age of onset of 30 years.

SYRINGOMYELIA PATHOLOGY

A chronic process leading to cavitations and gliosis usually in cervical or thoracic spinal cord.

SYRINGOMYELIA PATHOPHYSIOLOGY > This theory proposes that syringomyelia results from a "water hammer"-like transmission of pulsatile **CSF** pressure via a communication between the fourth ventricle and the central canal of the spinal cord through the obex. A blockage of the foramen of Magendie initiates this process.

SYRINGOMYELIA PATHOPHYSIOLOGY

William's theory

- This theory proposes that syrinx development, <u>particularly in patients with Chiari malformation</u>, <u>follows a differential between intracranial pressure and spinal pressure</u> caused by a valvelike action at the foramen magnum. The increase in subarachnoid fluid pressure from increased venous pressure during coughing or Valsalva maneuvers is localized to the intracranial compartment.
 - The hindbrain malformation prevents the increased CSF pressure from dissipating caudally. During Valsalva, a progressive increase in cisterna magna pressure occurs simultaneously with a decrease in spinal subarachnoid pressure. This craniospinal pressure gradient draws CSF caudally into the syrinx.



SYRINGOMYELIA PATHOPHYSIOLOGY Oldfield's theory

Downward movement of the cerebellar tonsils during systole can be visualized with dynamic MRI. This oscillation creates a piston effect in the spinal subarachnoid space that acts on the surface of the spinal cord and forces CSF through the perivascular and interstitial spaces into the syrinx raising intramedullary pressure.

SYRINGOMYELIA PATHOPHYSIOLOGY

- SPINAL CORD <u>NECROSIS</u> LEADING TO CAVITATION(Ischemia,Tumor,Radiation,...).
- EX-VACUO PHENOMENON DUE TO LARGE MYELIN LOSS AND GLIOTIC PROCESS.

MORE LIKELY IN MS; DEPEND ON AN <u>ALTERNATION OF CSF CIRCULATION BY EDEMA</u> <u>AROUND LESIONS(PLAQUE) LEADING TO</u> <u>ENLARGEMENT OF SPINAL CANAL AND CAVITY</u> FORMATION.

SYRINGOMYELIA

COMMUNICATING SYRINGOMYELIA(CS) OR HYDROMYELIA.

> NONCOMMUNICATING SYRINGOMYELIA(NCS).



CS OR HYDROMYELIA

Syringomyelia with fourth ventricle communication

Here, syrinx develop due to dilatation of the central canal by cerebrospinal fluid (CSF) because of:

Small posterior fossa, Platybasia and basilar invagination, Assimilation of the atlas, Chiari malformation, Arachnoid cysts, Spinal dysraphism.

NCS

Syringomyelia without fourth ventricular communication (cavity may be separate from the central canal)

Here, the syrinx or cyst develops in a segment of the spinal cord damaged by one of these conditions :

Arachnoiditis, Spinal trauma, Radiation necrosis, Hemorrhage, Tumor, Infection (Abscess, HIV), Transverse myelitis, Ischemic injury, Degenerative disease.



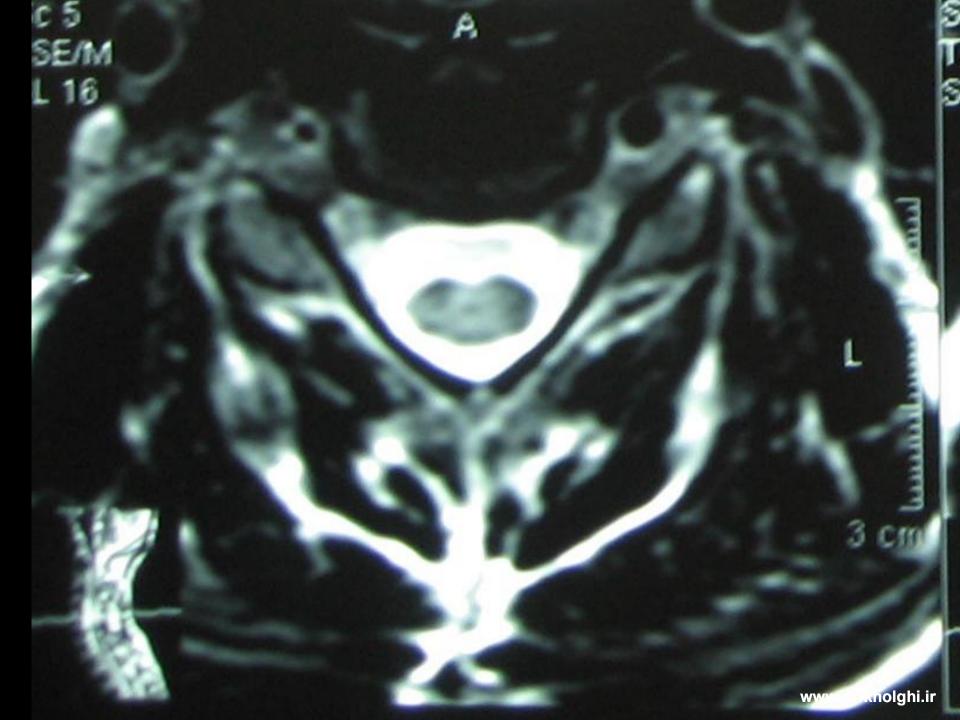
DEFINITE DIFFERENTIATION BETWEEN CS AND NCS

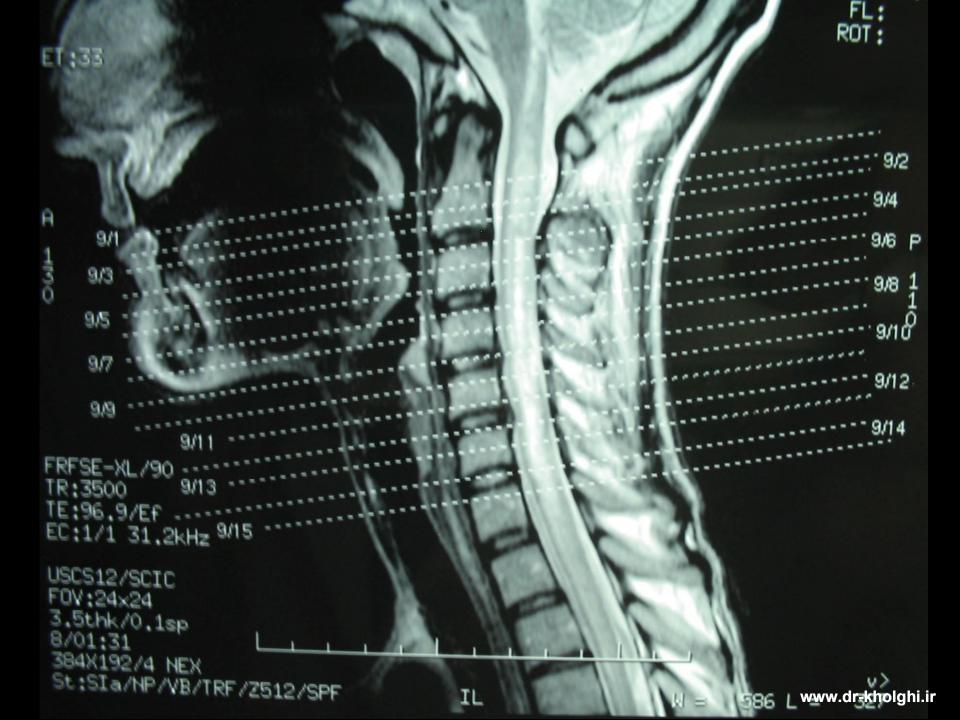
- IS POSSIBLE ONLY WITH PATHOLOGIC EVALUATION.
- > EPANDYMAL LINING OF CAVITY IN CS OR HYDROMYELIA.

> BECAUSE OF THIS; NCS OFTEN NAMED SYRINGOHYDROMYELIA MISNAMELY.









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REPORT OF A CASE REPRESENTING **MULTIPLE SCLEROSIS AS A CAUSATIVE** FACTOR

- A 31-years old female admitted with difficulty in walking and urinary retention since 3-days PTA.
- N/E showed spastic quadriparesia with more spasticity and weakness in lower limbs, abnormal deep sensation, left side sensory loss without definite sensory level.
- No atrophy in upper limbs or any decreasing in dermatomal sensation or asymmetry in biceps, triceps and brachioradialis reflex.

- > she was healthy until 9 years ago but have history of paraparesia, quadriparesia, blurred vision in both eye vertigo, difficulty in walking and left side sensory loss since 9 years PTA.
- > Altogether, she had 4 attacks of lower limbs weakness prior to recent attack.
- In one attack, 3 months PTA, she had quadriparesia with preference of weakness in lower limbs and pain and weakness in left upper arm

PARACLINIC

- Positive OCB were seen in CSF study.
- > Abnormal VEP in both eyes.
- Her routine lab, collagen vascular and vasculitis tests were negative.
- > EMG-NCV study was unremarkable for denervation in muscle.

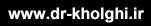


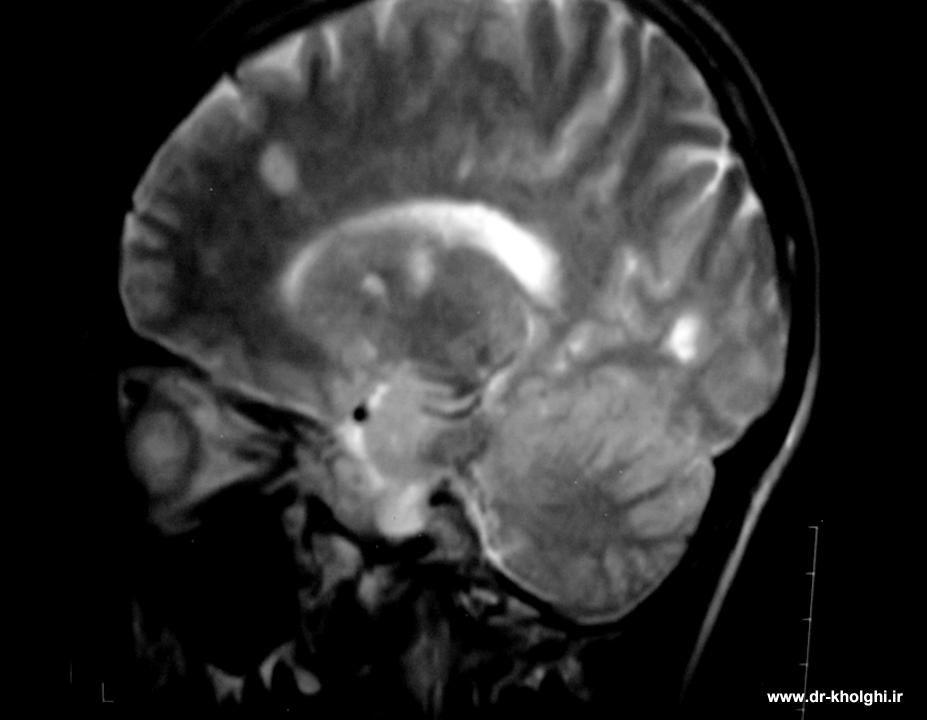
> There is multiple T2W hypersignal lesions in brain.

In cervical MRI and in T2W image there is a hypersignal area in C4 to C6, that is hyposignal in T1W, as a longitudinal cystic cavity showing syringomyelia.

In T1W MRI, there is no enhancement nearby cavity after contrast injection.

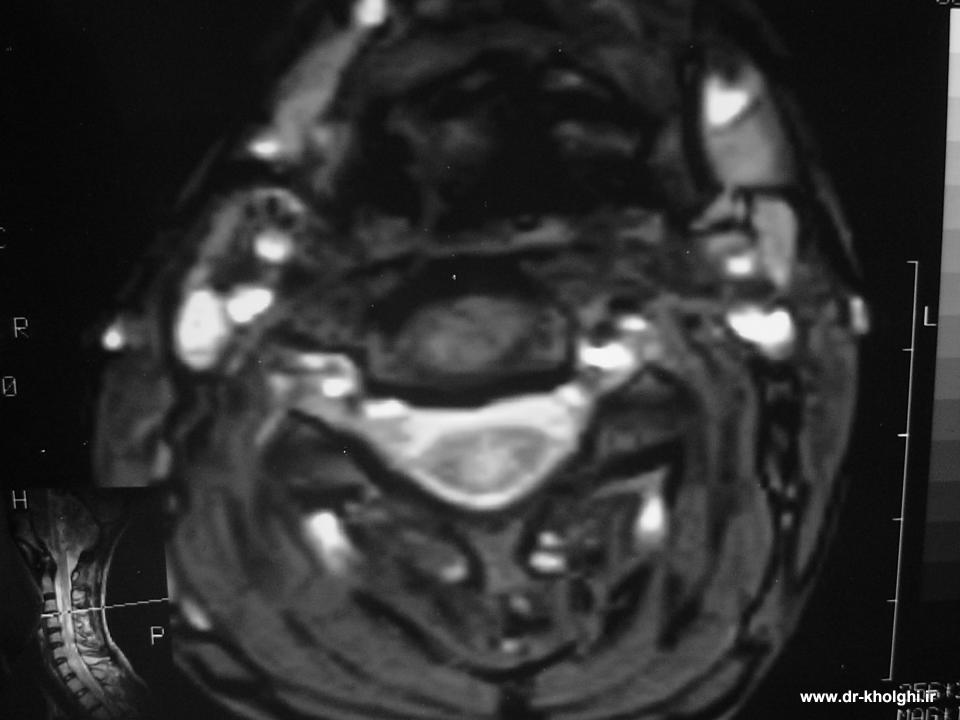












This is a NCS and not a hydromyelia or CS and not separated from central canal.

Upper limit of downward displacement of cerebellar tonsil:

Decade of lifemmFirst6Second or third5Fourth to eighth4Ninth3

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DISCUSSION

- She is known case of definite MS that has predilection of attacks in her cervical cord.
- > She has syringomyelia in her cervical cord.
- She has no involvement in spinal cord gray matter (normal EMG-NCV study).
- She has no Chiari anomaly or history of any other predisposing factors.



SYRINGOMYELIA PATHOPHYSIOLOGY Spinal cord injury theory

> SPINAL CORD <u>NECROSIS</u> LEADING TO CAVITATION(Ischemia,Tumor,Radiation,...).

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MORE LIKELY IN MS; DEPEND ON AN ALTERNATION OF CSF CIRCULATION BY EDEMA AROUND LESIONS(PLAQUE) LEADING TO ENLARGEMENT OF SPINAL CHANNEL AND CAVITY FORMATION.

CONCLUSION

> Very likely, at least in our patient that

CONCLUSION

> Our patient show us; not only NCS