University Journal of Medicine and Medical Specialities

ISSN 2455- 2852 2018, Vol.4(1)

AN INTERESTING CASE OF SYRINGOMYELIA ASSOCIATED WITH ARNOLD-CHIARI MALFORMATION TYPE I

SUBBURAJ

Department of General Medicine, STANLEY MEDICAL COLLEGE AND HOSPITAL

Abstract : A young male presented to us with chronic head ache ,neck pain, loss of pain and temperature sensation over left arm. Patient was investigated and found to have syringomyelia with Arnold chiari type I malformation. Patient was transferred to neurosurgery for suboccipital craniectomy procedure. He is on regular follow up.

Keyword : syringomyelia, Arnold chiari malformation type I, dissociative sensory loss, suboccipital craniectomy.

INTRODUCTION: Arnold Chiari malformation is diagnosed increasingly in recent times because of the widespread use of MRI .The syndrome has a wide array of clinical manifestations leading to frequent delay in diagnosis or misdiagnosis. Between 30% to 50% of type I Chiari malformations and 45% to 90% of type II Chiari malformations have an associated syrinx,. The syrinx associated with the Arnold chiari malformation usually is cervical or cervicothoracic .

Case report: A 16 years old male came to our medical opd with the complaints of Headache and neck pain - 4 years
Abnormal mobility of left shoulder joint - 2 years
H/o chronic occipital head ache for 4years, which was dull aching & lasting for 1-2 hrs daily; mostly in morning which is non progressive. Headache was aggravated by coughing, sneezing & playing. It was not associated with visual disturbances, vomiting & aura.
H/O neck pain for 4 years ,more in left side ,Insidious , non progressive, dull aching, continuous and not radiating. It was aggravated by playing. It was not altered by neck movements.
H/o abnormal excessive mobility of left shoulder for 2 years . There was no h/o trauma and swelling.
H/o recurrent subluxation of left shoulder while abducting the left arm after which patient voluntarily reduces.
H/O loss of pain & temperature al mobility of left shoulder joint - 2 years
H/o chronic sensation in left upper limb. he is able to perceive the cloth sensation.(dissociative sensory loss present)
No h/o cranial nerve involvement
No h/o weakness or wasting of muscles
No h/o cerebellar involvement
No h/o bladder and bowel involvement

GENERAL EXAMINATION:
Patient Conscious, oriented ,afebrile ,no pallor. not icteric/ cyanosis/clubbing/lymphadenopathy.
Height : neck ratio was 11
Upper :lower segment ratio-1
Height: arm span ratio –normal.
No neuro cutaneous markes.
No trophic changes in left upper limb .

VITAL SIGNS:

Blood pressure in supine posture-110/80 mmhg , standing- 108/80 mmhg

RR- 12/MIN

PR -78/ MIN

EXAMINATION OF CENTRAL NERVOUS SYSTEM;

- HIGHER MENTAL FUNCTIONS: normal
- CRANIAL NERVES - all cranial nerves normal
- MOTOR SYSTEM
- power all 4 limbs - normal (5/5)
- tone of all 4 limbs- normal

deep tendon reflex - left biceps jerk & Supinator jerk - absent, other reflex were normal.b/l plantar flexor

- No muscle wasting
- SENSORY SYSTEM- Pain and temperature absent in left upper limb,nape of neck in the left side ,angle of mandible and pectoral region up to T 2 dermatome .

posterior column sensation- normal

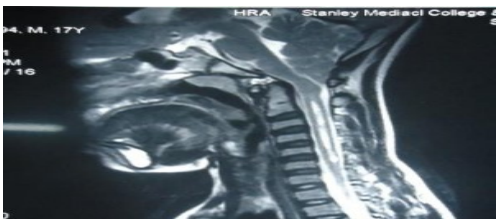
- EXAMINATION OF CEREBELLUM- normal
- GAIT – normal.
- AUTONOMIC FUNCTION TEST- normal
- EXAMINATION OF SPINE AND CRANIUM- normal
- FUNDUS - normal.

OTHER SYSTEM EXAMINATION:
CVS- S1 S2 HEARD . NO MURMUR
RS - NVBS HEARD
P/A- SOFT

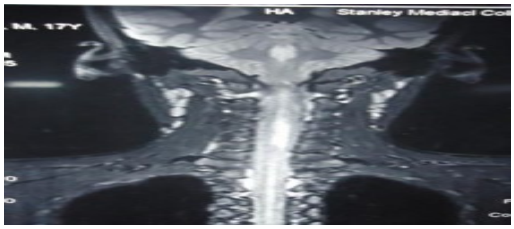
INVESTIGATIONS:

- CBC-Hb-12 gms%, Pcv -40, TC-6000cu mm, P60 L40 E2 ESR -10/20 mm

- RBS-120 mg Urea-24mgs%,Creatinine-0.7mgs%,Na-140 meq , K-4.5med
- X ray chest PA view-normal,
- x ray left shoulder- normal study
- ECG- normal study
- VDRL –NEGATIVE
- HIV I & II –NEGATIVE
- Nerve conduction study – normal
- **Orthopaedician opinion** was obtained for excessive shoulder joint mobility and diagnosed as multi dimensional instability of shoulder and no feature suggestive of charcot shoulder. they adviced shoulder strenthening excercises
- *MRI CERVICAL SPINE* showed linear hyperintense signal extending from C 2 to T 9. Cerebellar herniation of about 17 mm seen below foramen magnum with tight foramen magnum, suggestive of Arnold-chiari malformation type I.



Sagittal T 2W MRI - herniation of cerebellar tonsil with syrinx in cervical cord



Coronal image of Arnold chiari malformation type I

FINAL DIAGNOSIS-SYRINGOMYELIA INVOLVING CERVICAL, THORACIC CORD REGION WITH ARNOLD –CHIARI MALFORMATION TYPE I

DISCUSSION: Our patient presented with chronic occipital head ache , neck pain and unilateral dissociated sensory loss . Unilateral dissociated sensory loss is probably due to dissection of left spinothalamic fibres before crossing due to enlargement of syrinx. Type I chiari malformation most commonly presents with head ache and syringomyelia related symptoms. Chiari malformations types I-IV, refer to a spectrum of congenital hindbrain abnormalities affecting the structural relationships between the cerebellum, brainstem, the upper cervical cord, and the bony cranial base. True incidence of type I malformation is not known. Male: female ratio is 2:3. It is common in both adult & paediatric age group

PATIENT

Classification of Chiari Malformations:
Type I-Elongation of the cerebellar tonsils and the medial parts of the inferior lobes of the cerebellum , as cone-shaped projections accompanying the medulla oblongata into the spinal canal.
Type II -Displacement of the parts of the inferior vermis, pons and medulla oblongata together with elongation of the fourth ventricle (most cases are associated with spina bifida)
Type III -The entire cerebellum herniates into the cervical canal Type IV -Cerebellar hypoplasia
IV -Cerebellar hypoplasia .

SYMPTOMS RELATED TO SYRINGOMYELIA
1 .Disruption of CSF flow through foramen magnum

- Most common symptom-head ache
- Head ache and neck pain in Chiari I are often exacerbated by cough and Valsalva manoeuvre
- syringomyelia and central cord symptoms such as hand weakness and dissociated sensory loss

2 . Compression of medulla and upper spinal cord

- myelopathy
- lower cranial nerve palsies
- nuclear dysfunction
- Compression of cerebellum
- ataxia
- dysmetria
- Nystagmu
- ysequilibrium.
- dysequilibrium.

Symptoms and Signs of Adult Chiari Malformation (excluding those related to syringomyelia)

system	subjective symptoms	*objective signs*
ocular	transient visual obscurations ,photophobia, diplopia	papilledema, absent venous pulsations, decreased visual acuity
otological	dizziness, tinnitus,decreased hearing,vertigo,hyperacusis	nystagmus, sensory neural hearing loss, abnormal vestibular testing
lower brain stem & lower cranial nerves	dysphagia, dysarthria, sleep apnea, throat pain, palpitation, syncope,shortness of breath, hypertension	impaired gag reflex ,vocal cord paralysis, hypoglossal nerve palsy
sensory system	retro orbital & occipital head ache, cervical pain, facial & acral numbness, paraesthesias ,poor position sense , burning dysesthesia	analgesia, impaired proprioception
motor system	weakness	weakness, spasticity, hyper reflexia
cerebellum	unsteady gait, poor coordination, impaired fine motor function, tremor	dysmetria, ataxia
others	Chronic fatigue, altered recent memory,nausea, vomiting, urinary incontinence,impotence, trophic disturbances	

How syrinx is formed in Arnold chiari malformation type I ?
Herniated tonsil at the foramen magnum acts like a valve .It separates the CSF in to cranial & spinal compartments. The increase in subarachnoid fluid pressure from increased venous pressure during coughing or Valsalva manoeuvres is localized to the intracranial compartment. Increase in cisterna magna pressure occurs simultaneously with a decrease in spinal subarachnoid pressure. This craniospinal pressure gradient draws CSF caudally forming the syrinx.

Recommended Therapy:
Treatment is mostly surgical. However not every tonsillar ectopia, Chiari-related syringomyelia, or symptomatic chiari malformations needs to be treated. The following
factors are taken into consideration when addressing the risk/ benefit ratio:
• The severity and nature of the symptoms
• The alteration in the patient's quality of life secondary
to these symptoms
• Associated psychological factors
• The presence of symptomatic syringomyelia
• The surgical complication rate and
• The long-term results of surgical treatment for the particular symptom with most studies is
between 50% to 85%.

The surgical decision is highly individualized, especially because most symptoms are subjective . Usually surgery is reserved for patients with disabling or unbearable symptoms that are likely to be related to the chiari malformation. Some patients with minor symptoms need only reassurance that their symptoms are caused by a real disease and that they are not dangerous or life -threatening.

In ACM associated with syringomyelia, there is concern that the destruction of spinal cord tissue may lead to irreversible neural damage. Some recent evidence has suggested conservative management for asymptomatic cases. eight of nine patients in the Nishizawa et al. series remained asymptomatic after more than 10 years follow-up. Surgery is indicated in cases of symptomatic syringomyelia, especially when there is clinical deterioration or the patient experiences unbearable symptoms. In patients with ACMs whose symptoms are caused by basilar invagination or compression from a ventral pannus management follows that of the ventral pathology.

Surgical Techniques
Preferred technique includes suboccipital craniectomy, C1 laminectomy, dural opening, and duraplasty. Intraarachnoid exploration is reserved for cases in which macroscopic assessment of restoration of the CSF flow is not possible. A C2 laminectomy is performed in cases of tonsillar herniation below the midpoint of the C1-C2 interspace. Other variations include incising the outer leaf of the dura and leaving the inner leaf intact, intra-arachnoid exploration, tonsillar shrinkage, and fourth ventricular shunting. A variety of materials are used for the duraplasty. It includes pericranial , pericardial, fascia lata, dural allografts, bovine pericardium and synthetic
patches.

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