



Congenital Neurosurgical Diseases

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Learning Objectives:

- Introduction to Neurosurgery
- Approaching neurological symptoms
- Congenital diseases
- Hydrocephalus - Neural Tube Defect-Chiari Malformation**
- Dandy-Walker Malformation
- Craniosynostosis and Arachnoid Cyst
- Doctor said you have to know the details about red color and the others just definition

Color Index:

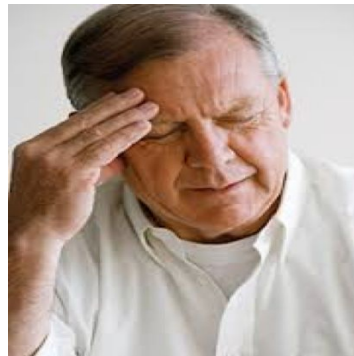
- Slides -Important -Doctor's Notes -Davidson's Notes -Surgery Recall
- Extra

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Approaching Neurological Symptoms

Headache or facial pain



In taking history not every pain in head considered headache, it could be facial or dental problems. So make sure is it headache or something else?!

Headache

Primary headaches

Are benign, recurrent headaches not caused by underlying disease or structural problems

*Examples: **Migraines**, **tension-type headaches**, **cluster headaches**

Secondary headaches

-serious “**brain tumor, bleeding**”

-Are caused by an underlying disease.

-International Headache Society (IHS) classification

→**Important:**

(**SSNOOP**) is a mnemonic to remember the **red flags** for identifying a secondary headache

- **S**ystemic symptoms (fever or weight loss)
- **S**ystemic disease (HIV infection, malignancy)
- **N**eurologic symptoms or signs
- **O**nset sudden (thunderclap headache)
- **O**nset after age 40 years
- **P**revious headache history (first, worst, or different headache)

In general People complaining of their "first" or "worst" headache Progressively worsening

*→Certain "**red flags**" indicate a secondary headache. When someone known to have migraine but she feels it change in the last week **for example:** it was in the right side but now she feel it everywhere, she feel numbness in the left hand this is **red flag**

Differential Diagnosis of headache :

- Vascular
- Inflammatory/Infectious
- Neoplastic
- Degenerative/Deficiency/Drugs
- Idiopathic/Intoxication/Iatrogenic
- Congenital
- Autoimmune/Allergic/Anatomic
- Traumatic
- Endocrine/Environmental
- Metabolic

The most common type of headache is tension headache.

Is the headache serious?

The American College for Emergency Physicians published criteria for **low-risk headaches**.

- Age younger than 30 years
- Features typical of primary headache
- History of similar headache
- No abnormal findings on neurologic exam
- No concerning change in normal headache pattern
- No high-risk comorbid conditions (for example, HIV)
- No new concerning history or physical examination findings

Differential Diagnosis of CNS space-occupying:

Neoplastic, Vascular, Congenital, Inflammatory and Infectious.

what to ask the patient about?

1- Local compression

(functional areas)

- motor cortex → weakness
- sensory → numbness or seizure
- cerebellum → tremor, dysarthria, ataxia or even truncal ataxia
- pituitary adenoma → visual loss...

2- Mass effect & Herniation: when it starts to increase in size, the brain tissue will shift to the other side most common and serious.

Q: if there is a left side mass increasing in size causing herniation what other 2 major symptoms you are going to see in the exam? Uncal herniation

- 1-contralateral weakness
- 2-ipsilateral fixed dilated pupil (afferent in cranial nerve 2 and efferent in cranial nerve 3 which is usually compressed by the herniation)

3- High ICP

- Headache.
- Nausea.
- Vomiting.
- High BP
- impaired level of consciousness
- papilloedema

Differential diagnosis of 906 patients who presented to a general neurology clinic with headache or facial pain as the major or only symptom

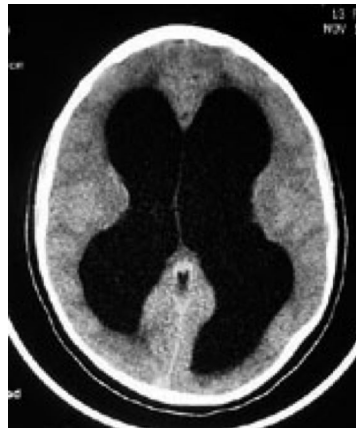
Diagnosis	Number	%
Tension headache	296	32
Migraine	241	27
Headache ? Cause	139	15
Post-traumatic	71	8
Facial pain ? cause	38	4
Depression	29	3
Trigeminal neuralgia	29	3
Cluster headache	19	2
Malignant IC Tumour	14	1.5
Benign IC Tumour	9	
Temporal arteritis	6	
Post-herpetic neuralgia	5	
Benign IC hypertension	4	
Cough headache	3	
Subdural haematoma	2	
Sinus infection	1	

CT scan: left brain lesion



Hydrocephalus

- Hydrocephalus: is an increase in the CSF volume, associated with increased ventricular size
- In old patient, they loss some of volume of brain that cause shrinking of the brain and losing tissue →we call it* ventriculomegaly or hydrocephalus ex vacuo). and they present with dementia like symptoms
- Not all ventriculomegaly is hydrocephalus
- Very important to know this enlargement of ventricles resulted from ventricle under pressure or hydrocephalus



Normal

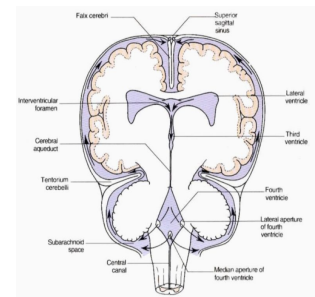
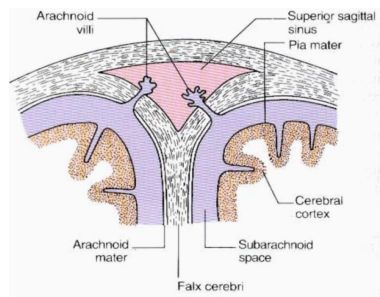
Hydrocephalus

Causes of hydrocephalus: (based on pathology)

1. Overproduction of CSF: such as : b/c of Choroid plexus papillomas in children (very rare)
2. Under absorption of CSF: example **post meningitis because of scarring**
3. Obstruction of CSF flow : **most common cause**

Physiology of CSF:

- Total volume of CSF in the ventricles varies from 5-15 ml in neonates to 150 ml in adults.
- Produced mainly by choroid plexus.
- Rate of production is 0.3-0.4 ml/minute.

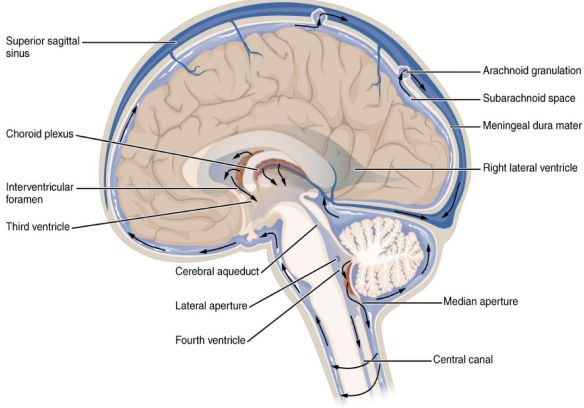


Where is the absorption of CSF? in Superior sagittal sinus and Arachnoid villi.

very thin layer, the CSF and blood vessels is under it . the sinus is separation between the dural (venous system) and inside it are the arachnoid villi → here where the absorption happen

Types of hydrocephalus

based on site of obstruction

Communicating (non obstructive)		Non-Communicating (obstructive)
<p>unimpaired connection of CSF pathway from lateral ventricle to subarachnoid space</p> <p>Cause:</p> <p>Overproduction or under absorption of CSF</p> <ul style="list-style-type: none"> -All ventricles are dilated -No obstruction in the pathway of CSF within the ventricles (the ventricles can communicate with each other) 		<p>complete or incomplete obstruction of CSF within or at the exit of the ventricular system</p> <p>Cause:</p> <p>blockage of the flow of CSF (obstruction) within ventricles or the pathway of CSF (obstruction to the CSF flow at the foramen of Monro, the third ventricle, the aqueduct of Sylvius, the fourth ventricle, or the foramina of Magendie or Luschka.)</p> <ul style="list-style-type: none"> ➡ Congenital, since birth ➡ Acquired, develops after birth -Partial dilatation

Etiology

Q's will ask: which one is a cause of hydrocephalus?

Congenital (happen during pregnancy)

Acquired

Q: all of the following can cause congenital hydrocephalus except?

with red are important, will ask about them in exam

- **Aqueductal anomalies: (most common cause):** Infants come after 2 week of natal with vomiting, sleepy, crying and increased head circumference very rapidly and CT show **obstruction (stenotic)** in the level of the aqueduct. → non communicating

-in the MRI and CT scan we see the lateral ventricle and the 3rd dilated but not the 4th. (abnormal communication between the 3rd and 4th)

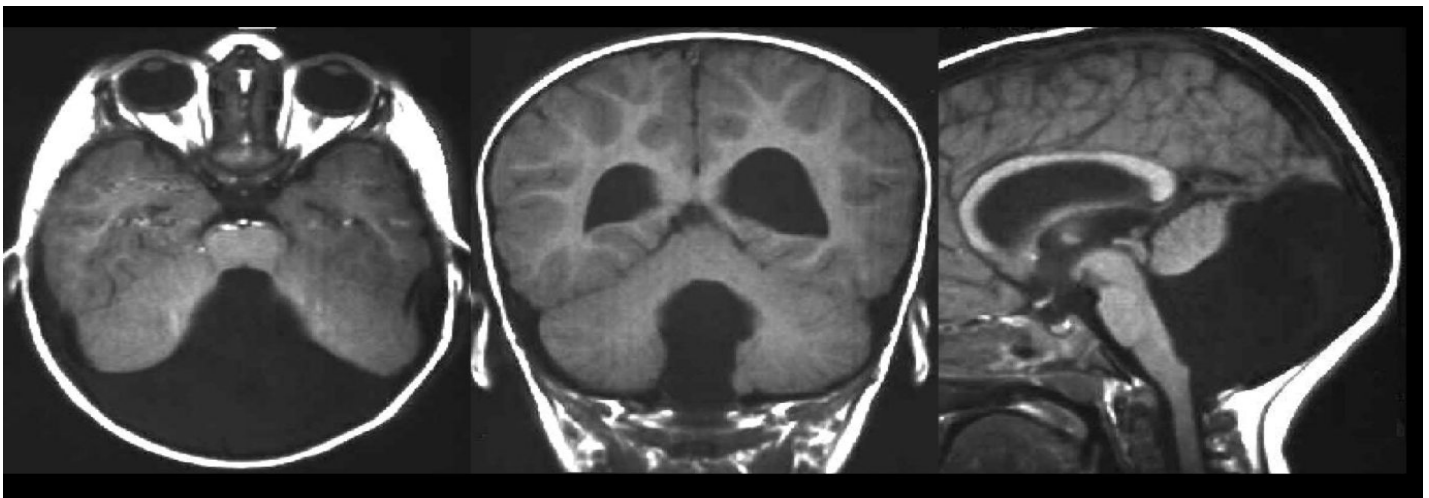
- **Dandy Walker malformation** (next title)
- **Chiari II malformation** (explain later)
- **Myelomeningocele** (explain later)
- Intrauterine viral infection (CMV, mumps, rubella, varicella)
- Toxoplasmosis
- Congenital tumors
- **Vein of Galen aneurysms** (next title)
- Chromosomal anomalies (trisomy 13 and 18)
- Congenital or primary hydrocephalus.

- **Germinal plate hemorrhage:** in premature babies <1500 gm (30%-40%) (immature blood vessel wall)
- **Post-meningitis**
- **Tumors**
- **SAH (subarachnoid hemorrhage due to trauma)**
- **Severe TBI (traumatic brain injury)**
- **Dural venous sinus thrombosis or Superior sagittal sinus thrombosis**

Dandy Walker malformation: (Congenital)

clinical features: Important

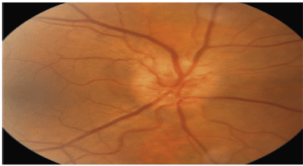
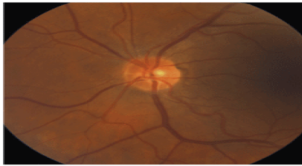
it is congenital disease cause increase in ICP b/c of hydrocephalus (noncommunicating or obstructive). the pathology is cystic formation in posterior fossa with cerebellar agenesis (cerebellar vermis hypoplasia), ataxia and mental retardation . in CT → to dilatation of 3rd ventricles + most of the time we can not see the 4th ventricles due to compression of the cyst



Pic note :in the middle is **Vein of Galen aneurysms** it is vascular congenital disease. so it cause obstruction at the level of 3rd ventricle
 Anything take contrast should be vascular or tumor



Clinical features: (Acquired) important

Infants & young children	Juvenile & adult: (↑ ICP)
<ol style="list-style-type: none"> 1. Increasing head circumference. 2. Irritability, lethargy, poor feeding, and vomiting. 3. Bulging anterior fontanelle. 4. Widened cranial sutures. <i>by examination not history</i> 5. McEwen's (cracked-pot) sign with cranial percussion. زي الحجب تخبط فيه تسمع صوت مويه 6. Scalp vein dilation (collateral venous drainage). 7. Sunset sign (downward deviation of the eyes). increase in ICP leads to the patient can't look up 8. Episodic bradycardia and apnea (If hydrocephalus is left untreated – the increased intracranial pressure will press on the brainstem – where the respiratory centers are located- which will lead to this) 	<ol style="list-style-type: none"> 1-Headaches 2-Nausea 3-Vomiting 4-Decreased level of consciousness 5-Focal neurological deficit (rare) 6-Papilledema some one have meningitis ,severe headache,vomiting and nausea. before doing LP you have to make sure there is no high ICP ,how? doing fundoscopy and brain imaging . 7-Drowsiness 8-Cranial nerve 6 palsy <div style="display: flex; justify-content: space-around; align-items: flex-end;"> <div style="text-align: center;">  <p>1. Optic nerve edema, O.D.</p> </div> <div style="text-align: center;">  <p>2. Normal optic nerve with 0.2 cupping, O.S.</p> </div> </div>

Investigations

- CT or MRI:
- The pattern of ventricular enlargement can help delineate the cause:
- Lateral & 3rd ventricle dilatation
 - normal 4th ventricle: **suggests aqueduct stenosis**
 - deviated or absent 4th ventricle: **suggests posterior fossa tumor**
- Generalized dilatation: **suggests a communicating hydrocephalus**



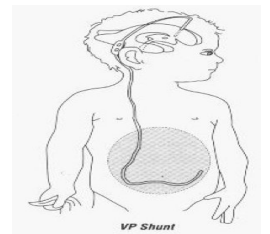
obstruction in 4th level obstructive

obstructive at 3rd

Non

Treatment

- Communicating hydrocephalus : **Medical** → **Carbonic anhydrase inhibitors** : {**Acetazolamide (diamox)** } or **surgical (AV shunt)**
- Obstructive hydrocephalus : **SURGICAL TREATMENT ⇒ VP Shunt¹**
- Possible complications of VP shunt infection with skin commensal organism such as *staph.epidermidis* and shunt could be blocked or malfunction



¹ Ventriculoperitoneal Shunt

Neural Tube Defect(NTD): groups of disease

Spinal Dysraphism is **failure of closure of posterior neural arch**

Most common site is lumbar or lumbosacral. Two major types: Open and Closed

If there is no closure at 3-4 week you may have any type of Neural Tube defect

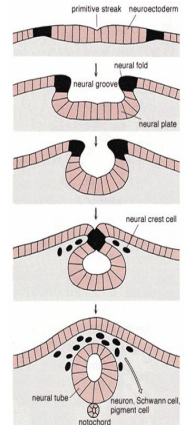
Open :direct communication with CNS structures and CSF leaking outside

Close: Communication with skin some time bone structure.

Closed neural tube defect is a malformation of the fat, bone or membrane. In some persons it cause few or no symptoms.



Close neural tube defect



•Incidence

2/1000 birth. Risk increase to 5% if a sibling is affected .Teratogens . How to prevent?

By giving folic acid supplement during pregnancy

What are the risk factor of neural tube defect or spina bifida?

Folic acid deficiency , upland pregnancy , family history (not that much risk), if she pregnant and take an anti-epileptic drug (teratogenic) prevent it by plan pregnancy with doctor

Types of Myelodysplasia (spinal Dysraphism)

Spina bifida occulta (Closed) Failure of closure the vertebral arch without herniation of intraspinal content	Meningocele (Closed) Failure of closure of vertebral arch with dura and arachnoid (CSF) herniation forming lumbosacral cyst	Myelomeningocele (open) Failure of closure of vertebral arch with herniation of both meninges and spinal cord
<p>5-10% of population, not clinically significant, tuft of hair, dimple sinus or port wine stain , high incidence of underlying defect and no treatment required</p> <p>Small gap in posterior arch in lumbar area it is asymptomatic and cause nothing</p> <p>The common scenario to discover it : Someone carry heavy weight then have back pain we do to him x-ray we find Spina bifida occulta. we do nothing. and the pain came from heavy weight not from spina bifida</p>	<p>-Cystic CSF-filled cavity lined by meninges</p> <p>-No neural tissues (Spinal cord)</p> <p>-Communicates with spinal canal</p> <p>-Look for other congenital Anomalies</p> <p>-Seldom any neurological deficit</p> <p>-Diagnosis by U/S or MRI</p> <p>Tx: Excision; urgent in case of CSF leak (rupture).</p>	<p>-Spinal cord and roots protrude through the bony defect</p> <p>-Lie within cystic cavity</p> <p>Lower motor lesion signs and numbness</p> <p>-Observe limb movements (degree & level of neurological damage)</p> <p>-Note dilated bladder (neuropathic bladder) & patulous annual sphincter</p> <p>-Paralysis and loss of deep tendon reflexes and sensation in lower extremities.</p> <p>-Gross hydrocephalus, multiple serious congenital anomalies</p> <p>-Diagnosis by U/S or MRI</p>

Case: if you have baby has pulse in back (may Meningocele or Meningomyelocele) what you going to do ?

Do Complete history and exam

Examine CNS for lower limb TONE, nature of moving . Babies will not tell you I am feeling or not but lower limb tone will Usually associated with other stuff like **Chiari Malformation, hydrocephalus** and other. better to do full CNS imaging

Case: if you have patient with thoracolumbar Meningomyelocele what do you expect having later?

weakness ,sensory disturbance ,hydronephrosis ,urinary retention(sacral nerve supply bladder S1,2,3 is lost) due to cord equina(compression to lumbosacral area all root in cord equina get compress S1,2,3)

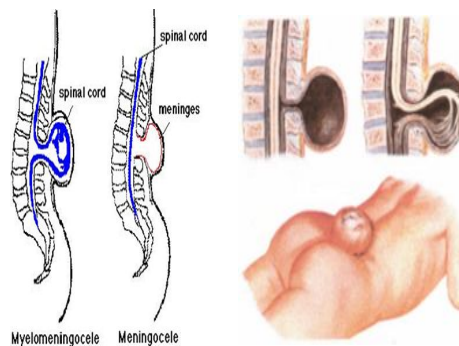
Treatment for both : close it.

We call it Emergency if :it open (risk of infection)/ Less emergency if: it close .

If the lesion in L1-2(sacral area: patient will not walk)

Risk factors make it more serious : 1/If it higher it become worse. 2/If it come with Infection

Meningomyelocele: common in lumbosacral area and it associated symptom are :hydrocephalus and chiari malformation



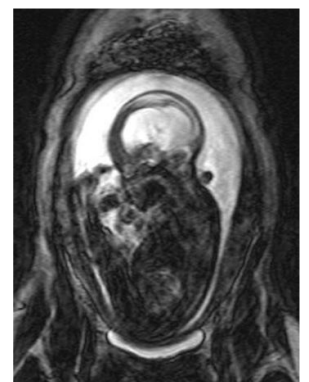
Antenatal diagnosis:

- Maternal U/S,MRI
- Maternal serum/amniotic fluid level for alpha-fetoprotein and acetylcholinesterase
- Contrast enhancing amniography
- Possibility of therapeutic abortion

We can see it during pregnancy by doing ultrasound if there is question mark we do MRI
MRI image we see : high Meningomyelocele . hydrocephalus(we see CSF in T2 (appears as Wight))

Should we do abortion ?

Depend on age, religion ,philosophy , parents opinion , if pregnancy 30 week do we do it ?



Other congenital anomalies:

1/Encephalocele: Herniation of brain meninges

Usually occipital .may contain occipital lobe or cerebellum. Often associated with hydrocephalus. Required immediate treatment if ruptured, outcome depends upon contents

it is high from brain can be: frontal or occipital . It is more serious : associated with dysfunction ,low IQ and rarely they survive. Risk factor no need to know it: Open Vs close. Big Vs small. If there is other brain anomalies (bad risk factor)



2/Chiari Malformation:

When part of the cerebellum is located below the foramen magnum, it is called a Chiari malformation. Very important usually we find it late (common in Females)

Case : 20 y/o female with headache in the back radiate to front of head with n/v and when she cough or go to toilet, she feels like electrical shock. The pain increases when turn her head down when she eats, she choke up(due to compression to brainstem).

-If there is cystic formation in posterior fossa is not *Type II* it is **dandy walker**

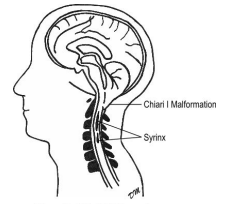


Figure 2. Chiari I Malformation



Type I	-Extension of the cerebellar tonsils into the foramen magnum , without involving the brainstem . can be congenital or acquired (MCQs)	Type 1 we try not to touch always in young 20-30 complain nothings only headache if it is severe we do decompress surgery and we remove part of posterior fossa to give space Associated with: Syringomyelia and obstructive Hydrocephalus
Type II	-Extension of both cerebellar (cerebellar vermis) and brainstem tissue into the foramen magnum congenital (MCQs).	-Type 2 (common)we should close Myelomeningocele and if there is ventricle dilated we do VP shunt -Patient has dysphagia ,Syringomyelia ,loss of pain and temperature sensation because of Syringomyelia (C8-T1) myelomeningocele and hydrocephalus
type 3,4	severe form you don't need to know them	

case :4 y/o No past medical history ,Worsening headache, occipital area 7 weeks,His headache worsened last night,Dizziness,loss of balance,N/V twice over the last 3 weeks:

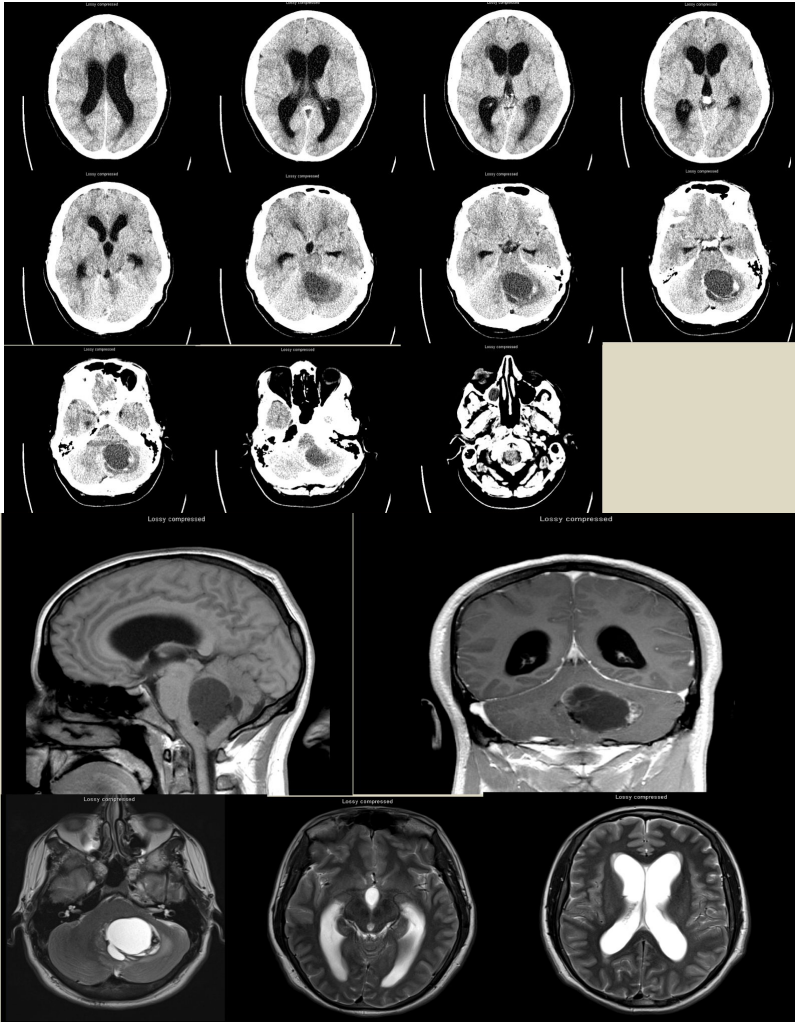
red flag: New headache ,Localize

worsened last night is a sign of high ICP specifically hydrocephalus

N/V also sign of high ICP

Ddx: Hydrocephalus caused by :vascular(bleeding), congenital, neoplastic infections(meningitis)it is not go on

-infection(meningitis) due: no fever ,duration 7 weeks



Ct scan shows:

Mass in posterior fossa causing obstructive (noncommunicating) at level of fourth ventricle because lateral ventricle and third are dilated but not the fourth (bone is wight)

MRI: the Difference in MRI that you can see details like :

small nodule inside cyst and also you can see aqueduct (bone is grey)

Cerebellar sign

-Gait ataxia

-Truncal ataxia

-Limb ataxia :Finger-nose and heel-knee-shin intention> tremor , dysmetria (past pointing) & dysrhythmia



-Cerebellar dysarthria

-Hypotonia

-Rapid alternating movements (dysdiadochokinesia)

-Tremor

-Nystagmus gaze-evoked, horizontal drift followed by a fast correction

Surgical Recall

1-What is the prognosis if untreated?

50% mortality; survivors show decreased IQ (mean 69); neurologic sequelae: ataxia, paraparesis, visual de cits.

2-What are the possible complications of treatment?

1. Blockage/shunt malfunction
2. Infection

3-What is hydrocephalus ex vacuo?

Increased volume of CSF spaces from brain atrophy, not from any pathology in the amount of CSF absorbed or produced

4-what are the three most common anatomic site of neural tube defect ?

- 1-lumbar region
- 2-lower thoracic region
- 3-upper sacral region

5- what is the prognosis of NTD?

95% survival for first 2 years in patient undergoing surgical procedure
25% survival for first 2 years in patient not undergoing surgical procedure

6-which vitamin thought to lower rate of NTD in utero?

folic acid

MCQs.

1/The investigation of choice in increased ICP is:

- a. Skull x-ray
- b. MRI
- c. Lumbar puncture
- d. CT scan

2/Obstructive hydrocephalus is best treated by:

- a. Surgery
- b. Drainage
- c. Craniotomy
- d. Endoscopic third ventriculostomy

3/Which of the following can cause communicable hydrocephalus?

- a. Tumor Cousin Obstruction
- b. Post Meningitis
- c. Germinal Plate Hemorrhage
- d. Aqueduct Anomalies

4/Which of the following is NOT ASSOCIATED with Chiari type II?

- a. Hydrocephalus
- b. Brain stem herniation into the foramen magnum
- c. Meningocele
- d. Myelomeningocele

Ans:1-D /2-D /3-B&C /4-C