CNS - Pathology

Malformations during embryonic development of CNS are rare BUT serious

Sometimes isolated but more in Multiple

Earlier malformation (early in embryonic life) → more severe insults

Etiology: insults that happen during embryonal life
- Gene mutations
- Chemicals (ex: certain drugs)
- Some infections (ex: rubella)

Not all insults can cause a gross/microscopic malformation, some result in functional problems, ex: disability

Types of malformations:

① Neural tube defects - most common
- A → brain
- P → Spinal cord

defects are more common (in born babies)

Causes:
- Genetic (recurrence 4-5%)
- Folate deficiency (folate taken early in pregnancy ↓ risk by 70%)

Spina bifida occulta → some herniation of spinal cord covered by meninges + bone defects

no risk for patients

neural tissue — covered by meninges — cyst (going outside body)

More severe → myelomeningocele
→ Spinal cord goes out of the vertebral canal (bone defect)
→ can cause problems depending on its site
↓
most common → lumbosacral region

Defects in ant. part of NT → problem in brain:
- less common: ① Anencephaly absence of brain + skull covering it
- ② Encephalocele → herniation of brain through defect in cranium

② Forebrain malformations
[A] → related to volume of brain / number of neurons
- large → megalencaphaly
- small → microencephaly → more common

Microcephaly → small ~~of~~ head circumference
Microencephaly → due to ↓ generation of neurons of cortex (↓ number of neurons) → NOT all are retarded but will have certain problems

[B] → related to problems with neuronal differentiation
problems with gyri → problem in structure of cortex
- Agyria
- Patchy gyri
- Poly microgyri (small but large in number)

③ Posterior fossa abnormalities (cerebellum)

severe ← a) Arnold-Chiari malformation (Chiari type 2)
b) Chiari type 1 → milder form of " "
c) Dandy-Walker → large posterior fossa ⊕ no vermis

④ Spinal cord abnormalities:

Most common 2 problems:

- Hydromyelia → expansion of central canal of spinal cord → mild + subtle
- Syringomyelia → same but with fluid-filled clefts

Brain edema:

↑ fluid in extracellular space of brain

- from cells → cytotoxic
  - initial problem within neurons or glial cells
  - due to toxins, hypoxia
- from blood vessels → vasogenic
  - initial problem in BBB + vessels of brain
  - generalized vs. localized → more dangerous

(Usually they coexist)

Shiny smooth surface + ↑ brain weight

Hydrocephalus:

↑ CSF in ventricles

due to ↑ production or ↓ resorption

- ↑ production: rare (tumors in choroid plexus)
- ↓ resorption: more common → due to obstruction

Many

↓ resorption of CSF

depends where the obstruction is ← localized → non-communicating → Enlarged ventricles

generalized → communicated → ↑ CSF in all parts of ventricular system

Herniation:

Brain → in skull (protection) → but any ↑ in size will ↑ pressure (intracranial)

⇒ expansion of brain tissue

⇒ 3 sites where this can happen:

falx

subfalcine

cingulate

can complex anterior cerebral artery → area supplied ⇒ ischemic

tentorium

transtentorial

uncinate

medial aspect of temporal lobe

can compress CN III → affect movement of eye + pupils

ischemia ← Post. cerebral artery

foramen magnum

tonsillar

cerebellar tonsils

will affect Cardio + resp. centers in medulla

most dangerous