Neurosurgery Compact

A Comprehensively Analyzed Work

First Edition
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Pediatric
Trauma
Vascular
[History, Diagnostic, Therapy]

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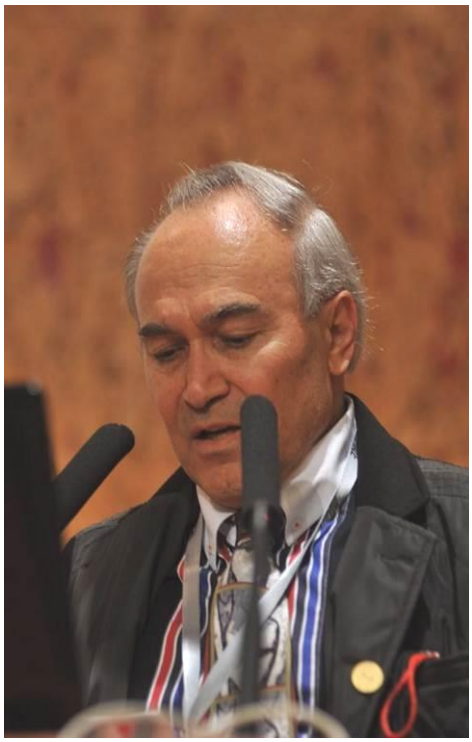

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TO THOSE,
WHO ARE SUFFERING

Preface

Rapid advances in the field of CNS disorders have opened a new route of diagnostic and therapy in this field.

Knowledge of current concepts associated with vascular disorders of CNS around the world is a crucial issue for providing appropriate evaluation, referral and treatment of patients with vascular diseases of CNS.

The main goal of this book is to provide broad based current knowledge of diverse fields of vascular disorders of CNS on a high level.

The book contains a compact text from several neurosurgical sources consisting of many journals and books, which have been published currently in the neurosurgical field.

The topics have been arranged in Top Down starting from cerebral vascular disorders to the spine to provide the readers with sound knowledge base in the fundamentals of vascular anatomy, history, vascular imaging, clinical assessment and also operative and non-operative therapy of vascular disorders of CNS.

The text has been made spare and concise therefore it could be read quickly in the clinic during the patient rounds, daily work and also in the operating room.

We have created about 44 tables by ourselves in the hope to keep the reader s attention to the essential points and to avoid unnecessary details.

In addition we have integrated about 71 suitable original photographs into the book.

The most important clinical and radiological features were usually labeled and highlighted in color.

The intended audience will be wide ranging including from medical student, residents, fellows, of course neurosurgeons and neuroradiology.

The book may also be of interest to physicians and nurses working at the ICU as well as patients with vascular diseases of CNS.

We are especially grateful to Dr. A. Khadem for bringing this book to editorial and technical completion.

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Pediatric Section

Pediatric Diseases of CNS

Developmental Anatomy of Central Nervous System (CNS)

Introduction

An overview of the developmental process of the nervous system is necessary for a better understanding of the anomalies and malformations which cause anatomical defects during the developmental process of the CNS. This process can lead to a range of malformations and clinical abnormalities such as total Dysraphism (prenatal death), cerebral palsy, retardation, as well as epilepsy. That may also occur in the spinal column such as Spina Bifida caused by failure of posterior neural tube to fully close.

History

The development of CNS begins in the third week of gestation. The origin of the NS is in the skin (Ectoderm) and lies on the dorsal of the embryo. This develops neural character and builds the neural plate. The neural plate rounds up and forms the neural tube. The development of the neural tube during the fourth week of gestation proceeds along the antero-posterior and medio-lateral axis and builds a bulge containing the primary 3 vesicles (i.e., neuromeres). This progresses to five secondary vesicles in the fifth week building the five major regions of the brain and the spinal cord.

The neural tube develops as a cylinder composed of a vesicle cavity and walls of variable thickness. The inner surface of the wall contains a cell-dense region that is known as the ventricle zone. The neural tube formation begins in the cervical level of C4 and expands upward to the cranial and downward to the caudal until neuro-pros is built. The opening at both ends of the NS closes at the end of the fourth week of gestation.

The primary Neurulation occurs as follows:

Ectoderm (stem cells) develops to neural character > neural plate > neural groove > neural tube > 3 primary vesicles > five secondary vesicles > five major brain regions plus spinal cord shown in below table and figures

Tab. 1 shows the first 3 above mentioned primary vesicles

- Forebrain: (Prosencephalon) = Telencephalon and diencephalons
- Midbrain (mesencephalon) = Midbrain and aqueduct
- Hindbrain (Rhombencephalon) = Metencephalon- Myelencephalon

Tab. 2 shows the 5 above mentioned secondary vesicles

- Telencephalon = Cerebral hemispheres + lateral ventricles
- Diencephalon = Epi- hypo- and subthalamus + III. Ventricle
- Mesencephalon = Midbrain + aqueduct
- Metencephalon = Pons + cerebellum + upper part of IV. Ventricle
- Myelencephalon = Medulla + lower part of IV. Ventricle

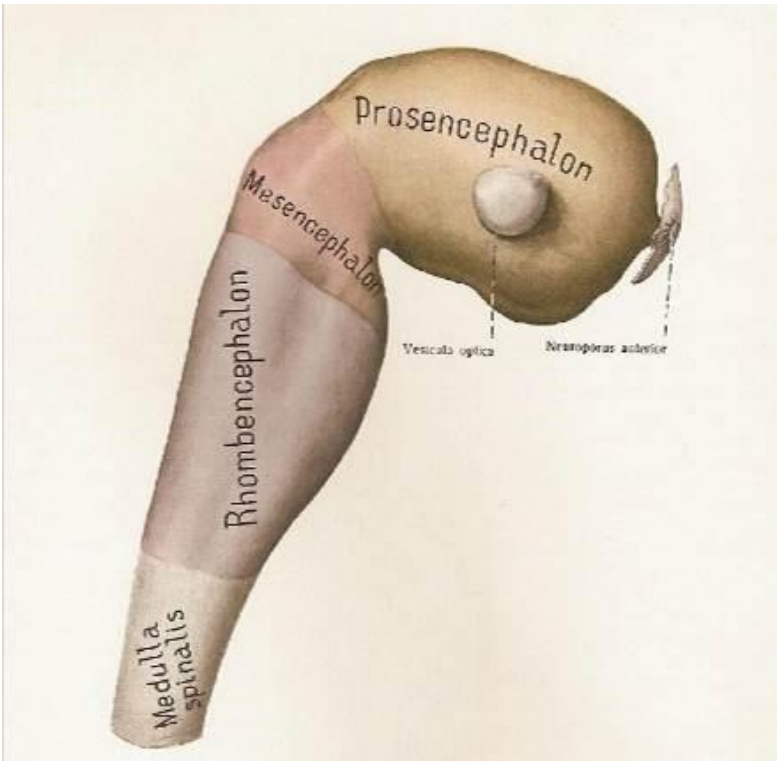

Fig. 1: 3 primary vesicles with Medulla/spinalis (Neuromeres)

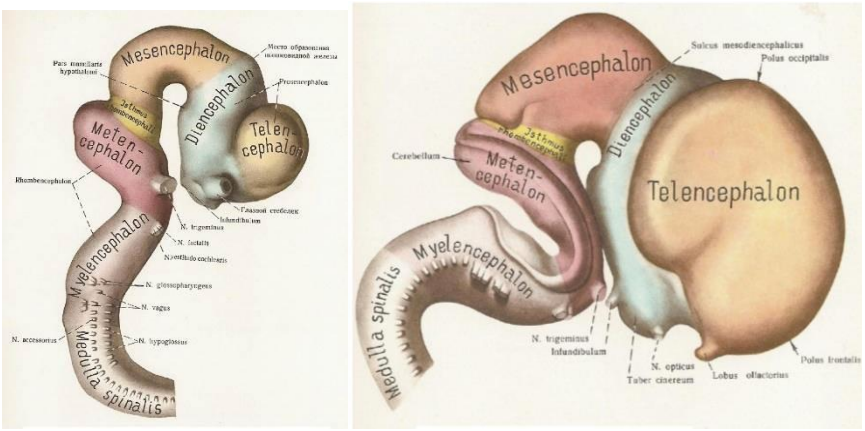

Fig. 2 & 3: 5 secondary vesicles plus Medulla/spinalis

Neural Cell Migration and Differentiation

This occurs in all types of neural cells at the inner surface of the neural tube, where many types of mitotic figures are found. The inner surface and its outside layer compose a cell-dense region called ventricle zone. This is the neurogenic and glia-genic region of the CNS.

Tab. 3 shows three separated layers on the wall of the neural tube

- Matrices zone: Ventricular zone with ependymal cells
- Mantle zone: It develops into gray matter of spinal cord
- Margin zone: It develops into white matter of spinal cord

After migration begins, the differentiation of neurogenic cells building the axon and dendrites simultaneously, this is completed by birth. The axon guidance is a complex multi-component process which has been challenged neurobiologists for years. The final cell division of most neurons occurs during the embryonic stage with the exception of the factory bulb. The last division occurs in the hippocampus area and cerebellum. The failure of any period of the developmental process as mentioned before can lead to malformations, and clinical abnormalities. A set of examples is listed and discussed in this chapter.

Neurolation Defects

Craniorachischisis

In the case of a total dysraphism, the neural groove is unable to close into the neural

tube. Many of them will die as spontaneous abortion.

Anencephaly or Exencephaly

It is caused due to non-fusion of anterior neuropore of either cranial vault or scalp cover. An anencephaly is often accompanied by myeloschisis.

Postneurolation Defects (migration defects)

Cranial Dysraphism

Holoprosencephaly

Introduction

This is the failure of cleavage of the telencephalic vesicle into separate cerebral hemispheres, which occurs during the fifth and sixth week of gestation. There are varying degrees of this, including the single ventricle, semilobar and lobar. In extreme cases, the holoprosencephaly cyclopia can develop (characteristic of only one ventricle and one eye).

Etiopathology /Classification

Trisomy and increased pregnancies by the same couple play a significant role in the development of holo-prosencephaly. Some researchers have found multiple teratogenic and genetic causes (single gene).

De Myer historically and roughly categorized holoprosencephaly into 3 types based on severity shown in below table

Tab. 4 shows different types of Holoprosencephly

- A-lobar holoprosencephaly: This is a mono-ventricle and fused cerebral hemispheres with absence of mid-line forebrain division.
- Semi-lobar holoprosencephaly: This is an incomplete forebrain division with partial separation of cerebral hemispheres mostly posteriorly of brain.
- Lobar holoprosencephaly: This is a complete ventricle separation, but with focal areas of incomplete cortical division or anterior falcine hypoplasia.

Symptoms

Microcephaly is the main sign as a rule, however in case of macrocephaly it will be a sign of hydrocephalus. The main symptom is also mental retardation, as well as seizures, hypotonia and /or hypertonia, dystonia and/or chorea. Hypothalamic and

brainstem dysfunction may lead to autonomic dysfunction and swallowing difficulties, whereas pituitary dysfunction can manifest as endocrine deficiencies. Further signs are usually seen at birth through associated midline facial anomalies including: Proboscis, cyclopia, cleft lip and/or palate, ocular hypotelorism, and solitary median maxillary central incisor.

Tab. 5- All of them have microcephaly or macrocephaly (through brain findings) shown in below table

- Microcephaly: Microcephaly caused by craniosynostosis
- Macro or megalocephaly: Hypertrophy of gray and white matter of brain which causes enlargement of skull
- Hydranencephaly: Loss of major part of cerebral hemispheres replaced by CSF, maximal Hydrocephalus

Diagnosis

MRI is the imaging study of choice 7, 8. The CCT and ultrasonography are the next studies however with limited result in cases of microcephaly with small fontanel.

Therapy

Medical care is necessary for control of problems associated with holoprosencephaly such as seizures, spasticity, dystonia, pituitary dysfunction, and gastro-esophageal reflux.

Surgical intervention is necessary in case of hydrocephalus (shunt procedure). It is usually obvious at birth even if antenatal diagnosis has not been made. In additional, these children also have systemic problems with poor feeding, hypothalamic / pituitary dysfunction and developmental delay 3.

Rhinencephaly

Introduction

Rhinencephaly is a developmental abnormality and is characterized as the part of the brain in front of the prosencephalon. This may consist of the one olfactory lobe or mostly two lobes, of them olfactory nerves arise. That usually insists of the olfactory bulb, tract and the regions of the limbic system of brain and functions as a smell brain. This may also comprise the olfactory lobe, the uncus, the subcallosal and supra-callosal gyri, the fascia dentata hippocampi, the septum pellucidum, the

fornix, and the hippocampus. It is a center in the cerebral hemispheres that governs the sense of smell in lower animals. It seems to mediate complex emotional behavior in the humans well known as phobia of rhinencephalon.

Symptoms

The fear or phobia of rhinencephalon can produce both physical and physiological symptoms. with typical anxiety disorder such as sweating, nervousness, nausea, rapid heartbeat, difficulty breathing, increased blood pressure, and tightness in the chest. Depression syndrome or depression symptoms are additional symptoms in a person suffering from a phobia of olfactory brain.

Therapy

Cognitive behavioral therapy is usually the most effective therapy for a phobia of rhinencephalon. That will be achieved by changing the behavior of the person and the person's brain. Through cognitive behavioral therapy, a patient affected by this phobia will "learn" to not fear rhinencephalon anymore. This is also used to treat certain anxiety- and depression disorders as well as mood disorders without the use of anxiety medi-cation, or depression medication. The patients feel better and will be able to focus their self on their life and overcoming their olfactory brain phobia.

Lissencephaly

Developmental failure of cerebral convolutions at an early fetal age, when brain normally starts to folding. In the case of lissencephaly, the brain does not fold properly. Instead, the brain remains smooth. This can affect baby's neural function and may cause severe symptoms.

Etiopathology

Lissencephaly is caused by malformation in genetic conditions. There have been identified malformations in five genes as contributors to lissencephaly. They are LIS1, 14-3-3ε, DCX, RELN, and ARX. Mutations in these genes can cause varying levels of lissencephaly. Lissencephaly typically occurs when a fetus is 12 to 14 weeks old. During this time nerve cells begin to move to other areas of the brain while brain develops. In the case of a fetus lissencephaly, the nerve cells do not move which affects brain development. This can occur alone, but it is also associated with other anomalies (genetic conditions) such as *Miller-Dieker syndrome* and *Walker-Warburg syndrome***.**

Symptoms

Babies born with lissencephaly may have a small head (micro-lissencephaly. The common symptoms of lissencephaly are such as trouble swallowing, difficulty feeding, psychomotor retardation, malformed fingers, toes, or hands, muscle spasms and seizures**.**

Diagnosis

Weeks 25 to 30 are most common period for performing an ultrasound, because the brain should be developing.

After Birth of a baby experiences symptoms related to brain developing, imaging study such as ultrasound, CCT-scan and MRI are the best studies of choice. There are different mal-developing forms such as Brain smoothness is called (agyria), and brain thickening (pachygyria) and also polygyria shown in below table.

Tab. 6 shows different types of Lissencephaly

- Agyria (smooth brain surface)
- Pachygyria (broad and flat gyris)- Pachygyria is not easy to diagnose by CT or MRI.
- Polymicrogyria (small gyri with shallow sulci)

Therapy

Lissencephaly brain is not reversible. Treatments can be only conservative and palliative to support and comfort affected children. If a child suffers from hydrocephalus, or an excessive accumulation of cerebrospinal fluid, a shunt procedure may be necessary. In the case of present seizures in affected children an anticonvulsive therapy will be advocated.

Prognosis

This would be depended upon severity of malformation. Children with severe lissencephaly have a life expectancy range from 2-10 years, according to The National Institute of Neurological Disorders. Common causes of death can be aspiration of food or fluids, respiratory disease.

Schizencephaly

Introduction

Schizencephaly is a form of porencephaly. This is a developmental anomaly of the brain characterized by abnormal slits or clefts in the cerebral hemispheres. It can be a mild form of unilateral clefts in one hemisphere or bilateral clefts in two hemispheres. Patients with schizencephaly may also have varying degrees of

microcephaly, mental retardation, hemiparesis or quadriparesis. Some patients suffer from having seizures and some may have hydrocephalus.

Therapy

Generally, it consists of physical therapy, treatment for seizures and in the case of persisting hydrocephalus a shunt procedure. Prognosis is depending on the size of the clefts and the degree of neurological deficit.

Tab. 7 shows four types of Schizencephaly

- Cleft communicates directly with the ventricle.
- Cystic line through the cortical gray matter may communicate with ventricle (DD: Are porencephaly caused by ICH, penetrating trauma, or vascular infarct).
- Pia and arachnoid fuse.
- Open-lipped and closed-lipped.

Heterotopia

That is migration of gray matter into areas of white matter usually occurred from subcortical to subependymal of ventricles.

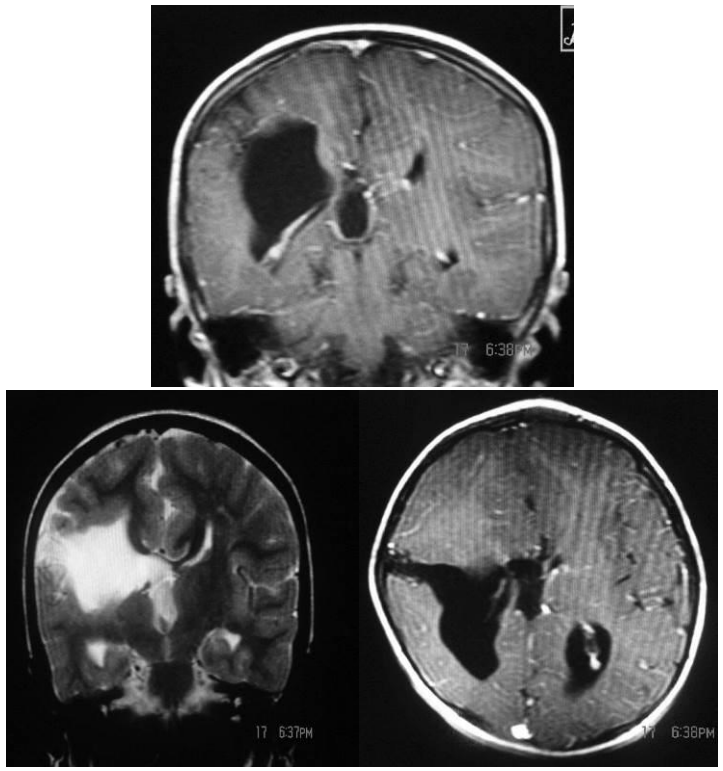

Fig. 4-6: Brain schizencephaly and agenesis of corpus callosum rare case

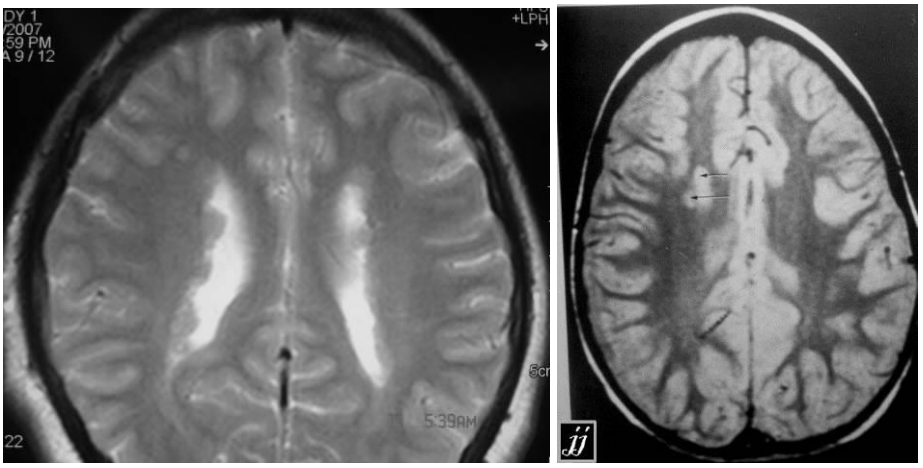

Fig.7 & 8: Axial MRI, T1 and T2 show Migration of gray matter into areas of white matter "Ectopic gray matter"

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