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INTRODUCTION

Cranium bifidum with herniation of the brain and/or its coverings through the bony defect has long been recognized. Neurologists and neurosurgeons are usually aware of this problem but unfortunatly E.N.T. specialists and family physicians are quite often not. It is due to this lack of knowledge that errors in recognition of such deformities may be commited and this subsequently leads to hazardous management.

Although meningoceles and meningoencephaloceles are relatively rare malformations, their occurance will confront the attending nurosurgeon or head and neck surgeon with a challenging situation.

Recent advances in reconstructive craniofacial surgery have much to offer in the treatment of certain types of such deformities.

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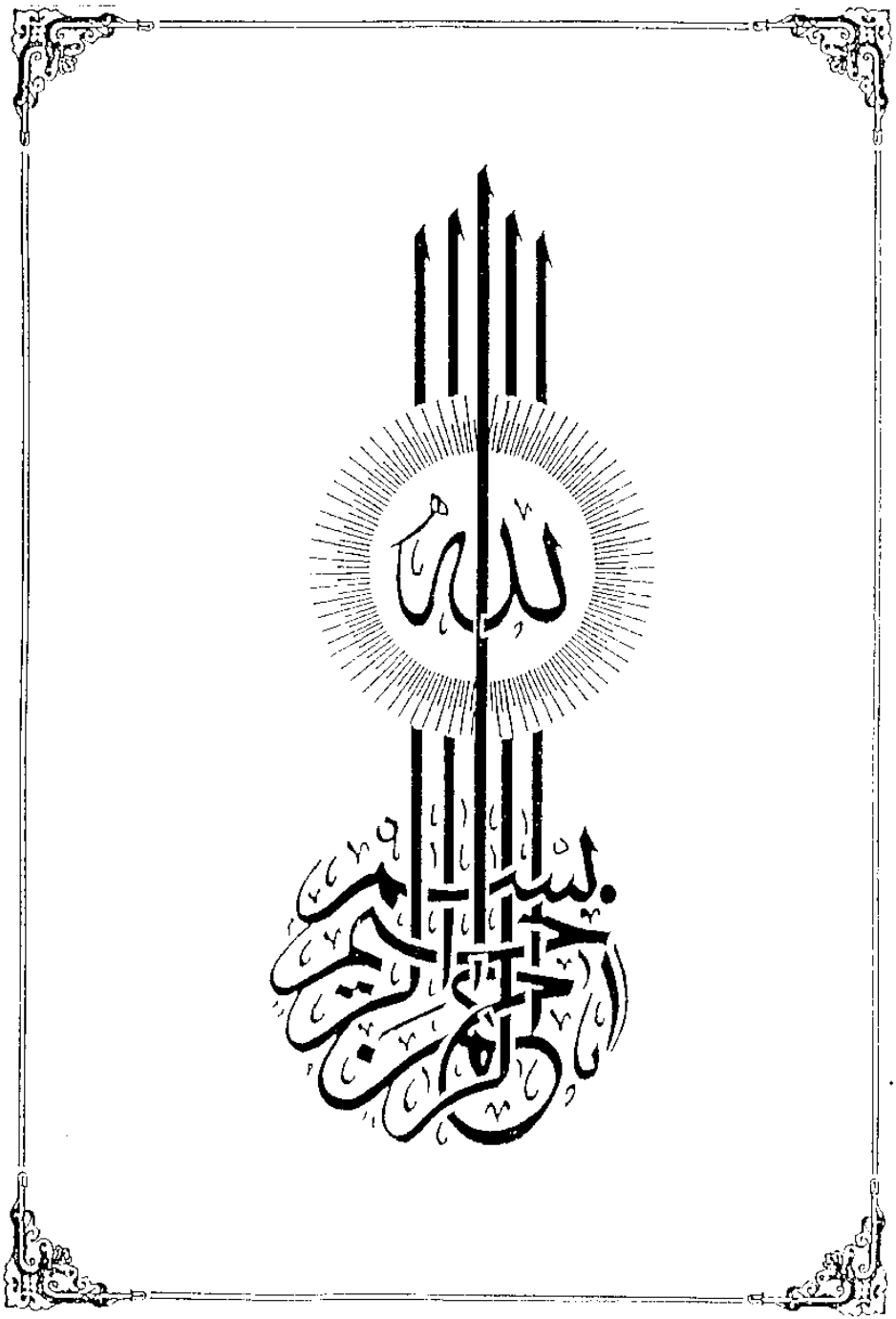

AIM OF THE WORK

Review of the literature as regards the cranial meningoceles and meningoencephaloceles. This will include the following points : Anatomy and Emberyology of the meninges and cranial bones, Aetiology ,Pathogenesis, Classification , Clinical Picture, Differential Diagnosis, Investigations, Prognosis and Management.

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EMBRYOLOGY OF THE MENINGES

The meninges may be devided in development into the pachymeninx [dura mater] and leptomeninges [arachnoid and pia mater] . Experimental work in lower vertebrates indicates that the dura mater is mesodermal in origin while the arachnoid and pia mater are closely associated in their development, being ectodermal in origin and largely derived from neural crest cells. However morphological studies on the spinal meninges of human embryos suggest that all three membranes are derived from the loose mesenchyme of sclerotomic origin surrounding the spinal cord, hence termed the[meninx primitiva]. Neural crest cells mingle with this mesenchyme and appear to be predominantly involved in the formation of the pia mater .

In the head, the mesoderm from which the cranial dura mater develops is closely associated at first with the mesenchyme which is chondrified and ossified to form the skull, and these layers are only clearly differentiated as the venous sinuses develop. [Henry Gray, 1969].

* * *

EMBRYOLOGY OF THE SKULL

The bones of the skull are developed in the mesenchyme which surrounds the cerebral vesicles ; but before the osseous state is reached, the cranium passes through blastemal and cartilagenous stages. However, not all parts pass through the phase of chondrification and hence the skull is formed of a cartilagenous and a mesenchymatous [membranous] parts developing together.

Some of the skull bones are composite structures derived from both sources. The bones of the skull base are mainly cartilagenous in origin, few are membranous.

On the contrary , the skull vault is of an entircly membranous origin.

The blastemal skull begins to appear at the end of the first month as mesenchymal condensation surrounding the cerebral vesicles. The first part to be evident is the occipital plate.

Then, the mesenchymal condensation extends

forwards, dorsal to the pharynx, to reach the primordium of the hypophysis, thus establishing the clivus and the dorsum sellae of the future sphenoid bone. Early in the second month it surrounds the developing stalk of the hypophysis and extends ventrally between the 2 halves of the nasal cavity, where it forms the anlage of the ethmoid bone and of the nasal septum.

Chonrification begins in the 2nd month, the first cartilagenous foci appearing in the occipital plates . The otic capsule, the presphenoid, the bases of the greater and lesser wings of the sphenoid and finally the nasal capsule, in turn become chondrified.

The bones of the cranial base which are thus preformed in cartilage are the occipital [except the upper part of its squama , the petrous part of temporal, the body , the lesser wings and roots of the greater wings of the sphenoid , and the ethmoid.

On the other hand, the frontal bones, the squamous parts of the temporal bones , the lateral parts of the greater wings of the sphenoid, and the upper [interparietal] part of the occipital squama are ossified directly in mesenchyme.

Ossification commences before the chondrocranium has fully developed, and thence, bone overtakes cartilage until little of the chondrocranium remains. However, parts of it still exist at birth, and small regions remain cartilagenous in the adult skull. At birth, the following points are still cartilagenous .

1. The ala and septum of the nose.
2. The sphenoid bone.
3. The spheneoccipital and sphenopetrous junctions .
4. The apex of the petrous bone (foramen lacerum) .
5. The occipital bone.

Henry Gray, [1969]

* * *

ANATOMY OF THE MENINGES

The brain and the spinal cord are enveloped by three membranes [meninges] named from outside inwards :

- The dura mater.
- The arachnoid .
- The Pia mater.

THE DURA MATER

it is a thick , dense inelastic membrane. The part of it which encloses the brain [cerebral dura] differs in several particulars from that which surrounds the spinal cord [spinal dura] ; however the two parts together form one complete membrane and are continuious with each other at the foramen magnum.

THE CERBRAY DURA MATER

It lines the interior of the skull, and serves a double function; acting as an internal periosteum to the bones,and a supportive membrane for the brain.

It is said to be composed of two layers :

1. An Inner or meningeal layer .
2. An outer or endosteal layer.

These 2 layers are closely adherent except at certain lines where they enclose the dural venous sinuses that drain blood from the brain.

The dura mater adheres to the inner surfaces of the cranial bones, and sends blood vessels and fibrous processes into them, the adhesion being most marked at the sutures, at the base of the skull and around the foramen magnum.

The endosteal layer of the dura mater is continuous through the sutures and the foramina of the skull with the pericranium and through the superior orbital fissure with the periosteal lining of the orbital cavity. The meningeal layer provides tubular sheaths for the cranial nerves as they pass through the foramina at the skull base. Outside the skull these sheaths fuse with the epineurium of the nerves.

The meningeal layer of the cerebral dura mater sends inwards four processes or septa which divide the cranial cavity into freely communicating spaces that lodge the subdivisions of the brain.

These septa are :

[1] THE FALX CEREBRI

It is a strong, arched process of dura mater

which descends vertically in the longitudinal fissure between the cerebral hemispheres . it is narrow in front , where it is fixed to the crista galli of ethmoidal bone and broad behind where it blends in median plane with the upper surface of the tentorium cerebelli; the narrow anterior part is thin and frequently perforated by numerous apertures . The upper margin of the falx cerebri is convex and attached to the inner surface of the skull on each side of the median plane as far back as the external occipitol protuberence, enclosing the superior sagittal sinus . The lower margin is free and concave and contains the infsagittal sinus . The straight sinus runs along its attachement to the tentorium cerebelli.

[2] THE TENTORIUM CEREBELLI

Is a crescentic arched lamina of dura which covers the cerebellum and supports the occipital lobes of the cerebrum. Its concave, anterior border is free ,and between it and the dorsum sellae of the sphenoid bone there is a large oval opening ; the tentorial incisure , which is occupied by the midbrain and the anterior part of the superior surface of the cerebellar vermis . Its convex outer margin is athached :

a) Posteriorly : to the lips of transverse

sulci of the occipital bone and postero-inferior angles of parietal bones , where it contains the transverse sinuses .

b) Laterally : to the superior border of petrous parts of temporal bones, enclosing the superior petrosal sinuses.

At the apex of petrous part of temporal bone the free border and attached periphery of the tentorium cross each other ; the anterior limits of the free border are fixed to the ant. clinoid processes of the sphenoid bone, while those of the attached periphery end on the posterior clinoid processes of that bone.

[3] THE FALX CEREBELLI

Is a small sickle shaped process of dura mater situated below the tentorium cerebelli and projecting forwards into the posterior cerbellar notch . Its base, being directed upwards , is attached to the posterior part of the inferior surface of the tentorium cerebelli, in the median plane ; its posterior margin contains the occipital sinus and is fixed to the internal occipital crest ; its apex devides into two small folds which are lost on the sides of the foramen magnum.

[4] THE DIAPHRAGMA SELLAE

Is a small, circular horizontal fold of dura mater, which forms a roof for the sella turcica and almost completely covers the hypophysis ; a small opening in its centre transmits the infundibulum. At or just below this opening, the dura mater, arachnoid and pia mater blend with one another and with the capsule of the hypophysis, so that within the sella turcica it is impossible to differentiate the individual membranes or to recognize the subdural and subarachnoid spaces.

THE VENOUS SINUSES OF THE DURA MATER

These are venous channels which drain the blood from the brain and the bones of the cranium ; they are situated between the two layers of the dura mater and are lined by endothelium continuous with that which lines the veins ; they contain no valves and their walls are devoid of muscular tissue. They may be divided into two groups :

1. Postero superior group : in the upper and posterior parts of the cranial cavity and these are ; superior sagittal sinus , inferior sagittal sinus, straight sinus, two transverse, two petrosquamous , two sigmoid

and occipital sinuses.

2. Antero - inferior group on the base of the skull and these are ; cavernous, intercavernous, inferior petrosal , sphenoparietal, superior petrosal, basilar and the middle meningeal veins.

THE ARACHNOID MATER

It is a delicate membrane enveloping the brain and spinal cord, lying between the pia mater internally and the dura mater externally . It is sperated from the dura by the subdural space . It is seperated from the pia mater by the subarachnoid space, which is filled with cerebrospinal fluid. The cerebral part of the arachnoid invests the brain loosely and does not dip into the sulci between gyri , nor into the fissures.

On the upper surface of the brain it is thin and transparent, at the base it is thicker and slightly opaque towards the central part. It cannot be identified in the hypophyseal fossa .

THE SUBARACHNOID SPACE

It is the interval between the arachnoid and the pia mater. it contains the cerebrospinal fluid and the large blood vessels of the brain, and is traversed by a network of delicate connective tissue trabeculae that connects the pia to the arachnoid.

At certain parts of the base of the brain, the arachnoid is seperated from the pia mater by wide intervals, which communicate freely with each other and are named subarachnoid cisterns ; in these the subarachnoid tissue is scanty and may be abscent.

The subarachnoid space communicates with the general ventricular cavity of the brain by three openings :

- The median aperture ; is in the median plane in the inferior part of the roof of the fourth ventricle .
- The two lateral apertures ; are at the extremities of the lateral recesses of that ventricle behind the upper roots of the glossopharyngeal nerves.

THE ARACHNOID GRANULATIONS AND C.S.F. CIRCULATION

They are macroscopic enlargements, or distensions of minute projections of the arachnoid mater, termed