CRANIA BIFIDA*

by

A. M. VARE

Crania bifida, like its spinal analogue could be defined as a condition where there is a defect in formation and closure of the cranial end of the neural tube together with a concommitant defect in the formation of various elements of the skull, resulting into an overgrowth of brain tissue outside the defective skull or an extrusion of a normal intracranial brain due to raised intracranial pressure or some such eventualities.

Obviously, therefore, it manifests itself in various degrees ranging from a tiny defect in the skull giving passage to a very small sac of meninges with a few millilitres of cerebrospinal fluid in it, to a very large defect pushing practically the whole bulk of brain matter outside the skull cavity. Such herniation of the brain or meninges is called meningocele or encephalocele as the case may be. When it is coexistent with a defect in the upper cervical spinal column, it is accompanied by herniation of a portion of the spinal cord too, thus producing an encephalomyelocele.

It is the purpose of this paper to present a rather unusual case of this kind.

A foetus was sent from department of obstetrics and gynaecology for dissection. Unfortunately, no track of the birth history was obtainable. The foetus was embalmed through the umbilical cord and dissected a few days later. It was in a condition of decomposition and the dissection was done as early as possible. The only information that could be obtained was that the foetus had passed the 7th month of gestation.

External appearances

Nothing remarkable was seen below the neck. The neck was extremely short. A large discoloured mass was hanging on the back from a large defect in the upper and posterior part of the skull.

The skull was deficient in the parietal, occipital and temporal regions. The frontal bones were well formed but there was no fontanelle. The parietals and squamous part of the temporals were completely missing. The occipital bone behind the foramen magnum was absent. The foramen magnum had no posterior margin. This defect was continuous with the defect in the cervical and upper thoracic column.

There was a triangular gap of a size sufficient to admit a finger tip in the lower cervical and upper thoracic spines. It was covered by a very thin membrane (? meninges). On removing this the spinal cord could be seen.

Brain

Whole of the brain was protruding out of the defect in the skull and hanging on the back by its own weight. It was quite large as compared with the size of the skull, brown in colour, deformed and covered only by a thin membrane. No sulci and gyri were seen even after a careful search, except for a well defined lateral

*From the Dept. of Anatomy, Medical College, Aurangabad.

Received for publication on 19-3-1968.

sulcus. The ventricles were dilated showing an internal hydrocephalus and the whole of the cerebral hemisphere was reduced to a mere shell bilaterally. The brain stem and spinal cord were normal. It appeared that the spina bifida in the cervical and thoracic region had involved only the vertebrae and the membranes. The pituitary was present and was of a perfectly normal size and shape.

Thorax

The thyroid could not be found at all. Part of the thymus was above the level of the clavicles. Most of it was lying in the right half of the anterior mediastinum. There were a number of constrictions in both the cervical and thoracic parts. The heart was toward the right side. Otherwise it was quite normal.

The right cavity of the thorax contained a bilobed right lung. Most of the small intestines were lying lateral to this lung and were covered with a thin membrane presumably the peritoneum. There was a large defect in the bodies of upper thoracic vertebrae immediately below the base of the skull through which a loop of intestine protruded into the vertebral canal. This loop was covered with a membranous sac and was displacing the cord. It was connected with the cord by a small fibrous band. The left cavity of the thorax contained the left lung, stomach, spleen and left lobe of liver.

Diaphragm

It presented three openings, one large opening through which the left lobe of liver was herniating into the thoracic cavity, another opening through which the stomach and spleen were entering which opening was rather a questionable one and a third opening which gave passage to the small intestines.

Abdomen

The appendix and caecum with a mesentery were found on left side. Both the kidneys were abnormally high and had a tendency to herniate through the diaphragm but had a covering of a membranous diaphragm on their upper surface.

Discussion

Cases of defects in the posterior part of vertebral column or skull are by no means rare. According to Willis it is very common to find a cystic swelling on the head of a new born with a little fluid in it projecting through a defect in the skull. The size of these defects vary from a tiny fenestration in any of the bones of the cranial vault to a very large opening producing extrusion of the main bulk of brain tissue.

Such cases can be of two types. In one there is a simple extrusion of the brain with a serious defect in the formation of the skull. The bony defect is more pronounced. These cases are akin to anencephaly in that there is a non-closure of cerebral part of the neural tube but differs from it in that there is an eversion of the forebrain so that the ventricular part of brain is exposed and projects. A second group is encephaloceles which are due to failure of closure of a far lesser part of cephalic neural tube and a defect in the skull. The defect in the skull is never so extensive and is present either in the frontal or the occipital pole. The extruded part is essentially larger than the defect. This is a strong evidence that the defect in the skull is secondary to developmental errors of the brain. Thirdly, the extruded tissue is over and above the normal brain tissue which is still intracranial. The extrusion is, therefore, never a simple herniation of a previously intracranial structure but an abnormal extracranial outgrowth, and abnormal folding of the neural tube.

It will be clear that the case under

report cannot be called an encephalocele because of a very pronounced defect and secondly because the intracranial brain has secondarily become extracranial. The whole brain being deformed the details of eversion could not be well made out. But the complete absence of the cranial vault with a total absence of the brain in the cranial cavity are against an encephalocele. It is, therefore, preferable to call this case an extroversion of the cerebral hemispheres.

Associated Anomalies

Most of the abdominal contents were herniating through the diaphragm. Whether this was due to the primary defect in the diaphragm or the maldevelopment of the diaphragm was due to misplacement of the contents of the abdomen is a problem. Most of the diaphragm was well formed except for these defects. The kidneys which had a tendency to herniate had also a membranous covering of diaphragm. When one considers the cord connecting the spinal cord with the gut derivatives, one is rather forced to admit that the cord which probably is the remains of embroyonic neurenteric canal and should have disappeared, persisted and exerted a traction on the developing gut. This traction drew the gut derivatives (stomach, duodenum, spleen, liver, intestines) into the thoracic cavity and hence the defects in the diaphragm. The kidneys having no such connection with the proximal gut remained covered with a membranous covering, presumably the diaphragm. Even here the traction has manifested itself in the form of high position.

One may, therefore, expect numerous gut anomalies and they are manifest in this case in the form of abnormal position of caecum and appendix, failure of absorption of mesenteries and reverse rotation.

In addition to these there is rightward displacement of the heart, evidently because of extensive diaphragmatic hernia.

It is unfortunate that we could not get any information about the prenatal and intrapartum events about this foetus. This case has been presented for whatever innate interest it may have.

Acknowledgement

I am extremely indebted to Prof. P. C. Bansal, M.S., F.R.C.S., for his valued guidance.

I am also thankful to Dr. G. K. Karandikar, Dean, Medical College, Aurangabad, for his kind permission to publish this paper.

My thanks are also due to the Department of Obstetrics and Gynaecology for the supply of the specimen.

References

1. Potter, E. I.: In Pathology of Foetus and New Born ed. 2, Chicago, 1961, Year Book Medical Publishers. Inc., p. 508.
2. Stowens, D.: In Paediatric Pathology, ed. 1, Baltimore, 1954, Williams and Wilkins Co., p. 299.
3. Willis, R. A.: In Borderland of Embryology and Pathology ed. 1, London, 1958, Butterworth & Co., Publishers Ltd., p. 150, 161.

Figs. on Art Paper VI