HYDRANENCEPHALY

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From time to time various types of encephalodysplasia associated with cyst formation have been described. Under the term 'cystic aplasia', Turnbull (1904) described a case where both parietal lobes were involved. Hamby, Krauss and Beswick (1950) have reported seven infants with a related dysplasia under the term 'hydranencephaly'. In these infants the cerebral hemispheres were completely or almost completely lacking, the space normally occupied by them being replaced with cerebrospinal fluid. The meninges were in their normal situation. At birth the skull was normal in size in most of the infants but showed progressive enlargement after two to 12 weeks. The condition has also been described by Bettinger (1940).

The case reported here is a further example of bilateral cystic agenesis involving the prosencephalon and is described because of its extreme rarity.

Case Report

N.M., a Bantu male child aged 7 days, was admitted to this hospital with a history of having had diarrhoea and vomiting for three days. The child was born at home and no history was obtainable from the mother regarding siblings and other members of the family. The child had been breast fed for one day after birth but since then had taken only sips of water.

On examination the child appeared cold and cyanosed and was gasping for breath. It did not appear to make any spontaneous movements but it cried normally. There was an icteric tinge to the conjunctivae. The cardiovascular and respiratory systems appeared normal. The abdomen was soft and no masses were palpable. There was no neck rigidity and Kernig's sign was negative. Externally there were no signs of any congenital defect.

Whilst in hospital the child took fluid with great difficulty and died a few hours after admission.

Necropsy Report

The body was of a normal-looking Bantu male child and weighed 6 lb. 8½ oz. The conjunctivae and subcutaneous tissues were jaundiced. The skull circumferences were as follows (normal values for the newborn European child are given in parenthesis).

Occipito-frontal circumference	35·13 cm.	(35·19 cm.)
Sub-occipito-bregmatic ,,	31·9 cm.	(32·9 cm.)
Occipito-mental ,,	32·0 cm.	(32·53 cm.)

Skull and Vertebrae. Externally the cranial bones were normal in structure and the sutures were in the normal anatomical planes. The diploë were normal. The foramen magnum was intact and there were no defects in the vertebrae.

Brain and Meninges. On the skull being opened both cerebral hemispheres appeared to be completely lacking. The cranial bones were lined with meninges which appeared to be thicker than normal. There was a failure of development of the falx cerebri which was represented by a short fold of dura mater about ½ in. in length. The tentorium cerebelli was intact except, however, that the normal free anterior crescentic border was ill-defined and the dura appeared to blend with a thinner membrane covering the incisura tentorii. This membrane consisted of pia-arachnoid and was continuous with the pia-arachnoid layer lining the dura of the cranial vault. The dura of the tentorium was continuous anteriorly with that covering the lesser wing of the sphenoid in the usual way. The superior sagittal sinus and the transverse sinuses were intact. In view of the failure of development of the falx cerebri, however, there was no sign of the straight sinus.

The cavity above the level of the tentorium contained 19 oz. of amber coloured fluid and floating free in the fluid was a small, round, greyish-brown object, which consisted of a fibrous outer covering enclosing a red-brown pultaceous mass. There was a small dimple on the outer surface suggesting that it had been attached to the surrounding meninges but no definite evidence of such connexion could be found.

Below the tentorium there were a normal looking cerebellum, pons and medulla. All the cranial nerves were present on both sides, with the exception of the first. The upper level of the brain tissue appeared to be about the level of the third ventricle which was open superiorly. The opening of the aqueduct was visible. The structures forming the floor of the third ventricle were intact and can be seen in Fig. 1. These include the optic chiasma, infundibulum, tuber cinereum, corpora mamillaria and posterior perforated substance. The inferior surface of both lobes of the cerebellum was flattened. The falx cerebelli was intact.

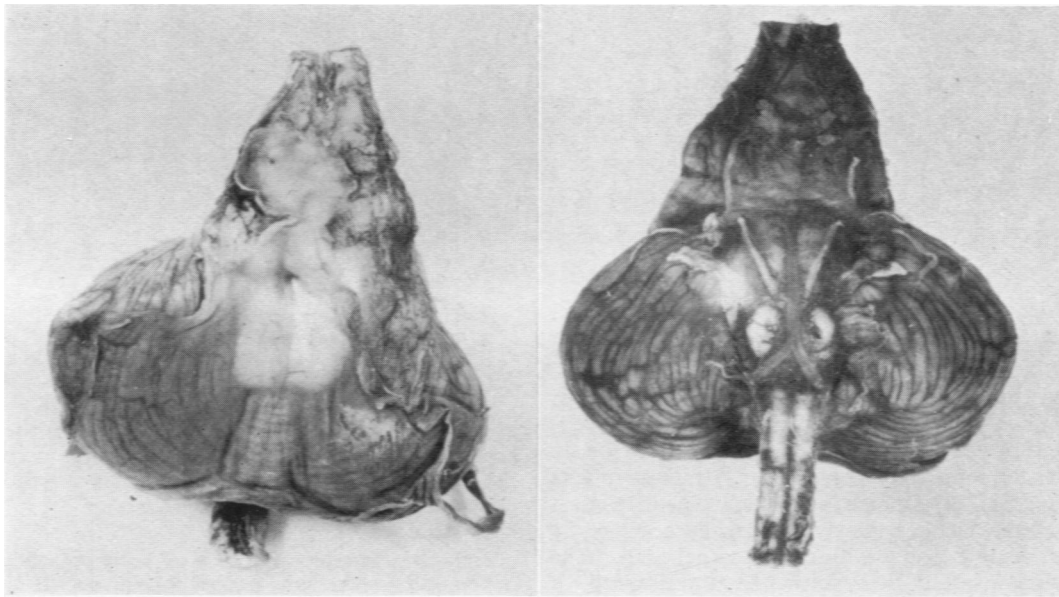

FIG. 1A. FIG. 1B.

FIGS. 1A AND 1B.—The cerebellum and the structures forming the floor of the third ventricle.

Sagittal section through the cerebellum and mid-brain in the midline revealed a normal aqueduct and fourth ventricle. The spinal cord was normal at all levels.

The fluid from the cranial cavity had the following composition: protein, 2·92 g.%; chlorides, 660 mg. %; sugar, 34 mg. %; total bilirubin, 2·5 mg. %; polymorphonuclear leucocytes, 4 per c.mm.; lymphocytes, 10 per c.mm.

Apart from the infundibular stalk there was no evidence of any pituitary tissue derived from Rathke's pouch. The sella turcica appeared shallower than normal.

No abnormalities were detected in the remainder of the skeleton.

Heart. There was a large patent ductus arteriosus and a patent foramen ovale. Both, however, may have been incidental findings since the foramen ovale may remain patent up to 1 year of life and the ductus arteriosus up to 3 months.

Kidneys. The left kidney was of normal size. The right kidney was about one-quarter the size of the left and projecting from the surface were several small cystic swellings.

Adrenals. Both were small and hypoplastic, unlike the large adrenal which is present in the normal infant of 1 week.

Thyroid and Thymus. Both appeared normal.

Histology

Paraffin-embedded tissues were sectioned in the usual way and stained with routine haematoxylin and eosin.

The spherical object found loose in the cranial cavity consisted of blood clot with a fibrous tissue capsule. There were areas of calcification.

The meninges from the cranial vault were thickened and there was an incomplete layer of brain tissue on the inner surface. The right kidney showed many hyalinized glomeruli and numerous small cysts, but no cartilage, as is often found in malformed kidneys. The hepatic bile capillaries were distended, and there was much pigment in the Kupffer cells. Also the collecting tubules of the normal kidney were stained with bile.

The pancreas, thyroid and thymus appeared normal.

Discussion

When the skull was first opened in this case it was felt that this might be an example of anencephaly associated with normal development of the skull. Such cases are rare but an example is quoted by Wilson (1940). However, the presence of brain tissue attached to the meninges of the cranial vault, in a very thin incomplete layer, ruled this out since anencephaly is essentially an agenesis of the prosencephalon and is almost always complete. In the present case the optic nerves and the structures of the floor of the third ventricle were intact, all structures derived from the prosencephalon.

The aetiology of cystic aplasia of brain tissue is

unknown. Examples of hypoplasia usually involving one cerebral hemisphere have been attributed to toxi-infective lesions during intra-uterine life. Turnbull (1904) attributed his case of cystic aplasia to arrested development. In the present instance there was no evidence of any inflammatory process and the aetiology seemed to be an endogenous failure of development of part of the prosencephalon due to abnormality of the primitive germ plasm.

Potter (1952) in her monograph on the pathology of the foetus and newborn points out that in hydranencephaly the fluid is cerebrospinal fluid. However, in our case the composition of the fluid was quite different. The high chloride level suggests that there may have been an admixture of blood and C.S.F. The presence of the mass containing old blood clot suggests that there had been previous bleeding and this may have been responsible for the high protein content of the fluid.

Hypoplasia of the adrenals is an invariable accompanying feature in anencephaly, and was also found in this infant. It has been suggested that this hypoplasia is secondary to failure of pituitary development. Potter (1952), however, has pointed out that in the normal foetus the adrenals may be bigger than those found in anencephalics before the pituitary could produce any hormonal effects. The cause of the adrenal hypoplasia is still obscure.

The condition of hydranencephaly is not incompatible with life and cases have been reported in children more than three years old (Potter, 1952). The mental age remains at the newborn level. These infants, just as in some instances of anencephaly, may feed and cry normally. In addition spontaneous movements of arms and legs are common.

Diagnosis of the condition may be made during life by transillumination of the skull.

Summary

A case of hydranencephaly is described. There was almost complete absence of both cerebral hemispheres the space being occupied by fluid. There was only a thin, incomplete layer of brain tissue in the region of the vault. The parts of the brain derived from the rhombencephalon and the mesencephalon were intact.

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