PORENCEPHALY AND CEREBRAL ABSCESS SIMULATING INTERNAL HYDROCEPHALUS.

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COURVILLE (1945) remarks that: "It was almost a medical habit in England on the part of physicians with literary leanings in the first half of the nineteenth century to write an essay on 'water on the brain'." It was not, however, until the experimental work of Dandy (1917) was published that a dynamic and scientific approach to the problem became possible. To this day no generally effective and applicable method of treatment has been devised. This is possibly due in part to the tendency to lump together a variety of conditions of different aetiology and pathology under the one heading. To emphasize this point it is perhaps useful to publish the following case, which clinically seemed to be one of simple hydrocephalus, but post-mortem examination of which showed that the symptoms had quite a different origin.

CASE HISTORY.

Born 19.x.41; illegitimate. Nothing significant in the family history on either side. There is a normal half brother ten years older than the patient. The mother tried to bring about an abortion, otherwise pregnancy and delivery were normal. The mother (a writer who had had several books published) was 33 at the birth of the patient. Enlargement of the head and backwardness were noticed shortly after birth. The child was admitted to hospital aged 14 months, and was then unable to stand, sit, or lift his head. He was blind from optic atrophy. The head circumference was 20½ in., the length of the skull 6¾ in. and the breadth 5¾ in. Otherwise he appeared very well nourished and in good general health. He was classed as an idiot and diagnosed as a case of hydrocephalus. There was no improvement in his condition from his admission to the time of his death on 23.viii.47. He never became able to play or to take any active interest in his surroundings. He had a major epileptic fit in 1946. On several occasions throughout his stay in hospital he had bouts of pyrexia reaching to 105 degrees. His death occurred at the end of one such bout. A lumbar puncture done on the day prior to death showed a normal cerebro-spinal fluid. All the febrile episodes were accompanied by vomiting and by drowsiness increasing in depth to coma at times. No cause for the pyrexia was found and the possibility of derangement of the heat regulating mechanism due to hydrocephalus was considered.

At post-mortem examination there was an incipient broncho-pneumonia. The skull was very much thickened, measuring $\frac{7}{16}$ in. at the thickest part, the vertex, and showing coral markings on the inner surface to the left of the vertex, the hypertrophied area also extended somewhat to the right of the mid-line (see photograph and X-ray films). The weight of the skull vault when removed was 14 oz. (Figs. 1 and 2). The meninges were firmly attached to this abnormal area and showed a ragged appearance when stripped (Fig. 3). The head as a whole was disproportionately large to the body and the skull was globular with a bulge forward over the eyes, which is usually regarded as characteristic of hydrocephalus (*cf.* A. F. Tredgold, 1947, "The peculiar enlargement of the skull makes diagnosis

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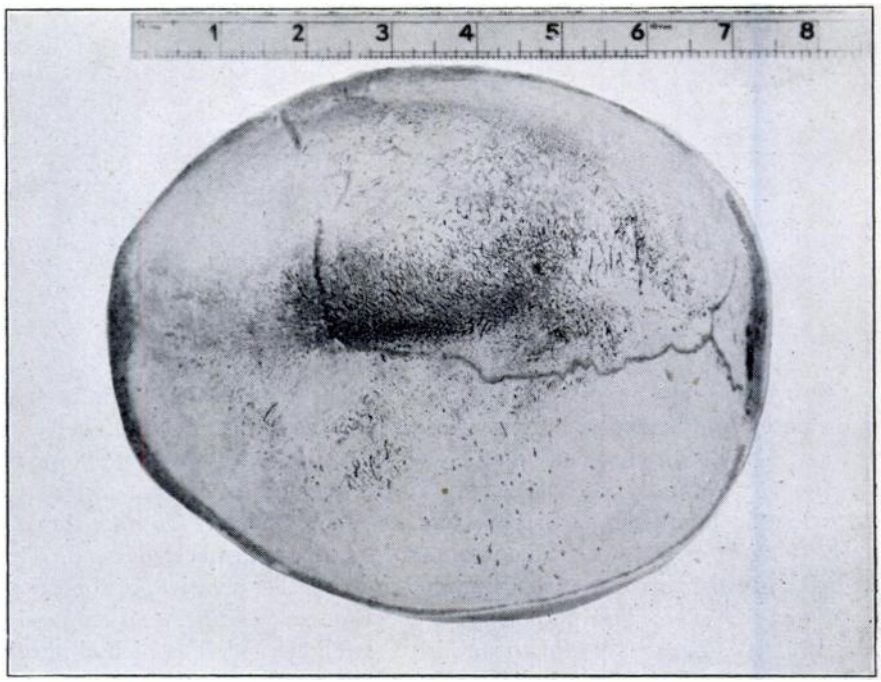


FIG. 1.—Photograph of cranial vault showing hairbrush marking corresponding to site of cerebral abscess. Note partial obliteration of sutures.

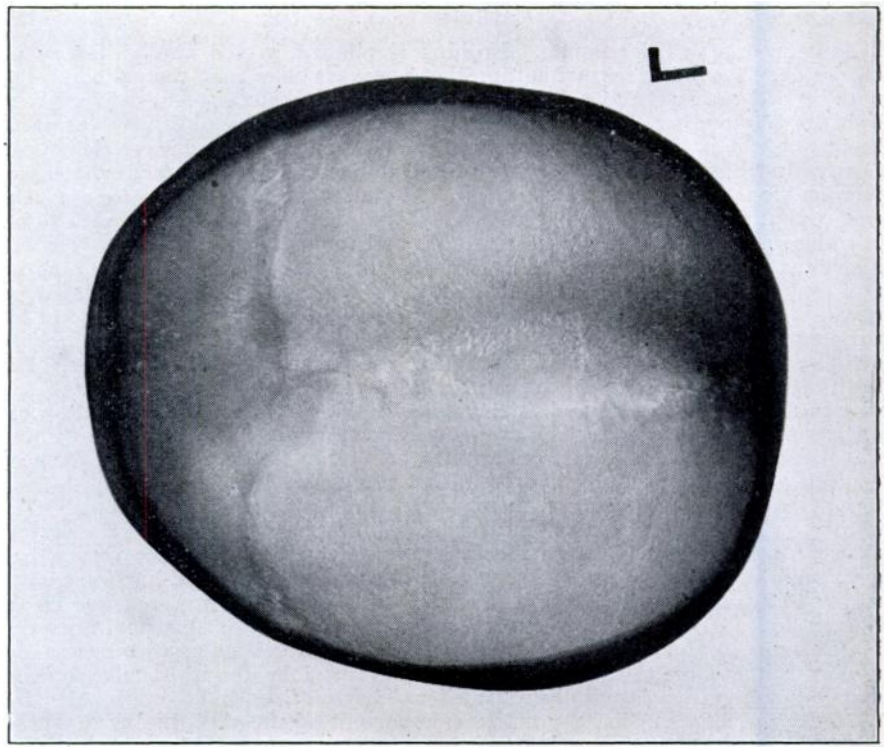


FIG. 2.—X-ray of cranial vault showing disturbance of normal bone architecture corresponding to site of cerebral abscess.

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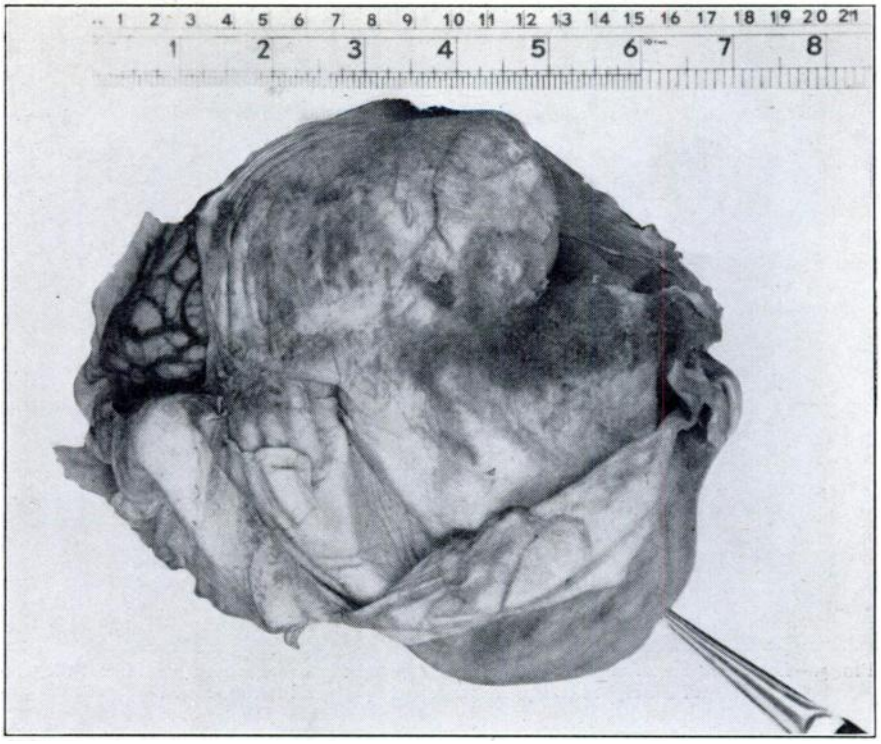


FIG. 3.—Superior view of brain showing thickening of meninges over site of cerebral abscess and ragged appearance where detached from skull.

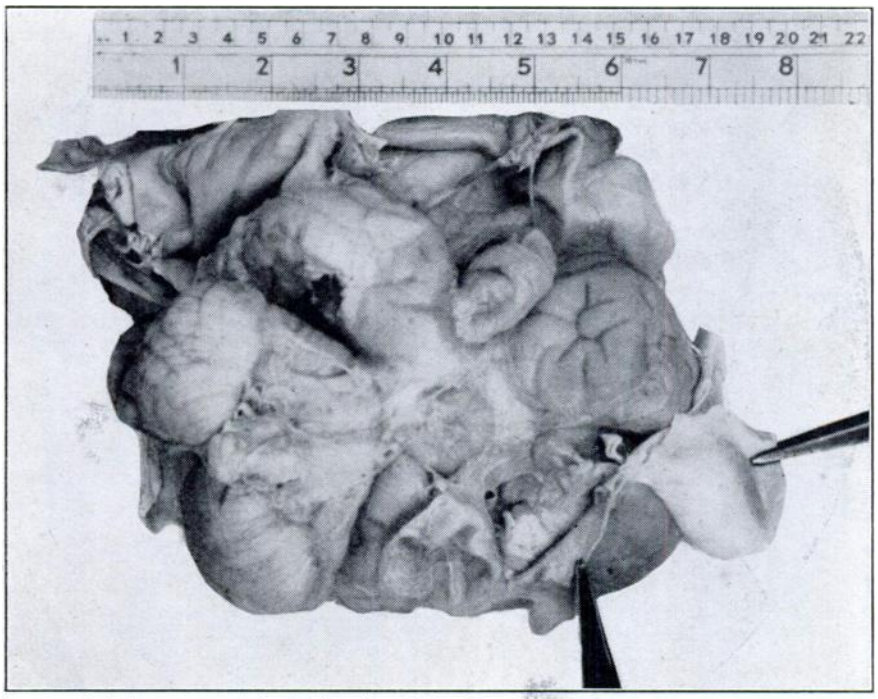


FIG. 4.—Inferior aspect of brain showing thickening of meninges, adhesions and openings to cavities on both sides.

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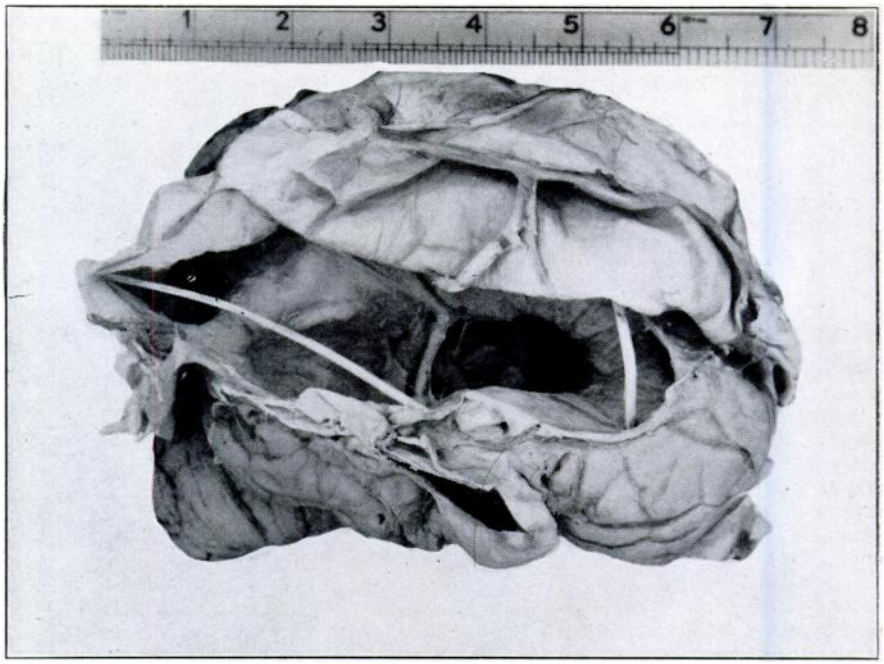


FIG. 5.—Lateral aspect of brain showing large porencephalic cavity occupying almost the whole of the right hemisphere traversed by several fibrous cords.

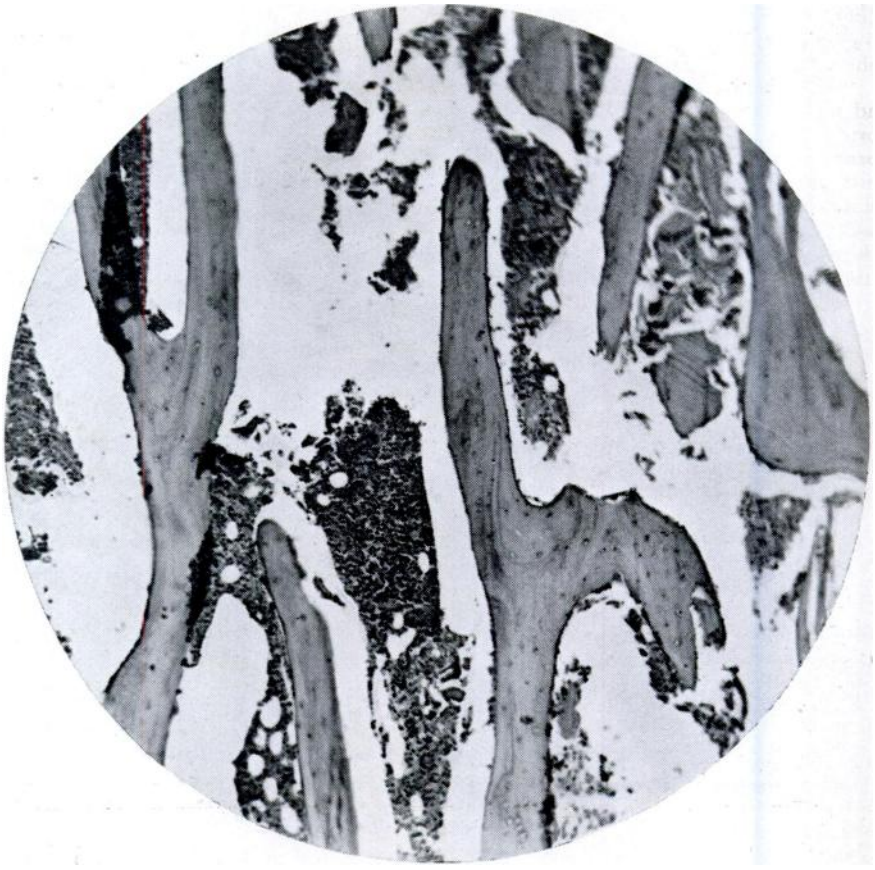


FIG. 6.—Section of bone from skull vault at site corresponding to that of cerebral abscess.

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easy."). The length was 7½ in. and the breadth 6 in. The fontanelles were closed and the sutures had in places entirely disappeared. There were no Wormian bones. In general, the union between the individual bones was so effective that the cranium could be said to consist of one piece.

Both sides of the brain (see Fig. 4) were distended and approximately equal in size. On the right there was little brain substance and the hemisphere which was filled with clear fluid showed a condition of porencephaly. On the left there was more brain substance and the cavity was filled with thick, yellow pus. On microscopy this pus showed degenerate polymorphonuclear leucocytes and diplococci. On culture pneumococci, staphylococci (*aureus*) and *B. Friedlander* were grown. It was assumed that this was a chronic pneumococcal abscess which had been responsible for the obscure pyrexia.

A report from Dr. Turner McLardy, of the Maudsley Research Laboratory, states :

" Brain weight : 1040 gm.

" In the *left hemisphere* the temporal pole was completely replaced by a smooth-walled cyst lined superficially by pia-mater. The whole of the white and grey matter of the frontal lobe except in the most mesial 1 cm. of its substance was replaced by a ragged-walled cavity. This cavity had no apparent connection with the temporal cyst or with the ventricular system. It was lined dorsally by thickened leptomeninges and dura mater, which contained a thick layer of sub-dural haematoma.

" In the *right hemisphere* (see Fig. 5) the temporal lobe showed the same condition as on the left side, but here the superior and middle temporal gyri were also extensively involved and the defect formed part of a large single cavity which replaced also the whole of the parenchyma of the occipital lobe and of the dorsal half of the frontal and parietal lobes. This cavity was continuous with the entire length of the lateral ventricle and was covered dorsally by thickened leptomeninges. It was smooth-walled throughout and traversed by several fibrous cords, one of which corresponded in position to the posterior pillar of the fornix. There was no evidence of corpus striatum or thalamus in this hemisphere. In the left hemisphere the basal ganglia were not accessible to inspection.

" Hypothalamic structures, except for sclerosed remnants of the optic chiasma and nerves, were obscured by dense fibrosis. The arteries of the base appeared normal in consistency. The cranial nerves (other than first and second) appeared normal on the right, but the fifth and sixth could not be identified on the left. Midbrain, pons and medulla appeared normal. The right cerebellum was slightly smaller than normal.

" There is no doubt that this is a widespread bilateral porencephalic process in the main, probably not of the schizencephalic type described by Yakovlev and Wadsworth. The cavity in the left hemisphere had probably been infected from the septic focus in scalp and skull."

Dr. M. K. Beattie, of the Central Pathological Laboratory, West Park, reports on the histology of the skull cap as follows :

" There is considerable dilatation of the Harvesian canals and marrow spaces which appear more variable in size and shape than normal. The lacunae also in parts are larger and more variable. The general pattern is more irregular and the inner compact layer of bone appears to be defective." (See Fig. 6.)

Discussion.

The inclusion of external and internal hydrocephalus under the same general head tends to obscure a proper appreciation of the subject. External hydrocephalus is, of course, a purely static accumulation of fluid in those instances where there is loss or non-development of cerebral substance. Internal hydrocephalus (including both communicating and non-communicating varieties which are clinically identical), on the other hand, is a dynamic and menacing phenomenon which is invariably fatal unless arrested. The case under discussion presented clinically the features of the latter type. Study of the

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cerebral anatomy was rendered more difficult by the great distension of the brain and the adhesions consequent on abscess formation, but if the porencephalic interpretation of the findings is correct then a diagnosis of non-communicating hydrocephalus is out of the question since the essential feature of porencephaly is that it is an inadequacy of the roofing-in process which normally isolates the ventricular system from surface of the brain except for the foramina in the roof of the fourth ventricle.

It should be pointed out, however, that Yakovlev in discussing this problem (1946) draws attention to the persistence of a thin membrane, sufficient in many cases to form a complete roof to the cavity, and to confine a hydrocephalic swelling which he ascribes merely to a disturbance of the hydrostatic balance due to the localized thinning of the cerebral wall. The persistence of a membrane representing the attenuated cortex is also well described by Turnbull (1904).

The state of the skull rules out also the diagnosis of a simple progressive type of internal hydrocephalus for in these cases the skull is excessively thinned and the individual bones widely separated where the lesion has developed before or shortly after birth, that is before union of the sutures. The great thickness of the skull was one of the outstanding features of this case and accounted for a considerable part of the clinical increase in the size of the head.

Porencephaly is usually divided into "true" and "false" (Buzzard and Greenfield, 1921). The validity of this division is, however, open to doubt (Kinnier Wilson, 1940) since it is probable that some at least of the "true" cases are instances of malformations resulting from interference with the circulation of the brain during pregnancy (Marburg, Rezek and Marks, 1945), just as the "false" cases are due to a similar disturbance in later life. As Dandy and Blackfan, referring to hydrocephalus, state, "such sub-divisions do not clarify the pathogenesis, but serve to obscure it."

Yakovlev (1946) separates porencephalies into encephaloclastic types with a normal cerebral "blue-print" and the schizencephalic type in which the "blue-print" was abnormal.

Déjerine (1898), using the terms "porencephaly" and "hydrocephalus" as interchangeable, makes a similar distinction, likewise based on a study of cortico-thalamic connections.

The origin of the cerebral abscess is obscure. The child developed a pyrexia of unknown origin soon after admission to hospital and continued to have similar bouts throughout its stay, that is for a period of nearly five years. It seems highly probable that the abscess had been present for the whole of this time. This would account for the extreme thickening of the skull cap and for the chronic appearance of the meningeal adhesions. It is unlikely that the occipital pressure sores which developed only terminally could have been responsible for the infection, particularly inasmuch as the outer table of the occipital bone showed no abnormality.

Grüneberg (1947) lists a number of types of hydrocephalus in rodents, including four inherited varieties in mice which have a suggestive bearing on the human condition. Evidence of inheritance of recessive factors in humans is, however, not always easy to obtain and in the absence of such definite proof

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undue emphasis should not be placed on the hereditary aspect. On the contrary, the search for pre- and post-natal environmental factors should be intensified. This search will be more successful if the heterogeneous groups which are at present labelled hydrocephalus, porencephaly, etc., are subjected to more critical scrutiny and broken down into their component parts. This will enable the importance of pre-natal infections, of blood incompatibilities, of nutritional factors or even as in the present case, of unsuccessful attempts at abortion (*cf.* Penrose, 1933) to be more accurately assessed.

Summary.

A case of porencephaly with cerebral abscess simulating congenital hydrocephalus is described. There was bilateral porencephaly with a unilateral pneumococcal abscess which had probably been present from the first year of life, as indicated by recurrent bouts of unexplained fever. The division of porencephaly into true and false types is criticized and the importance of study of environmental factors in both porencephaly and hydrocephalus is emphasized.

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